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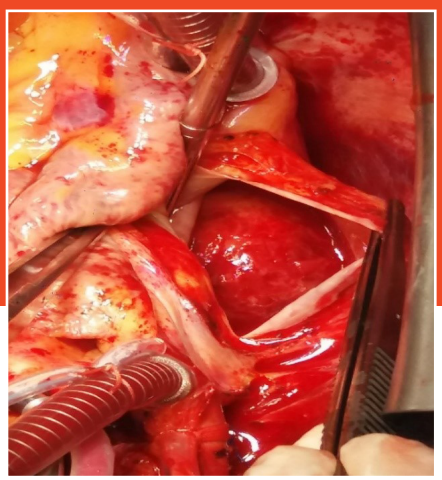
Thoracic aortic rupture associated with multiple aneurysms of medium-sized arteries: case report

Free-floating thrombus in the left atrium

Anomalous origin of the left coronary artery from the pulmonary artery as a life-threatening condition



Intraoperative finding. After incision of the left atrium, a smooth round thrombus was visualised. *(Rečičárová S, Jonák M. Free-floating thrombus in the left atrium)*

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Obsah

Refrakterní zástava oběhu na podkladě vysoce rizikové plicní embolie u mladého pacienta
O. Sirotek, V. Pechman, M. Hromádka, A. Žídková, J. Baxa, R. Rokyta 2

Thoracic aortic rupture associated with multiple aneurysms of medium-sized arteries: case report
K. Rücklová, E. Honsová, P. Tomášek 7

Free-floating thrombus in the left atrium
S. Rečičárová, M. Jonák 12

Anomalous origin of the left coronary artery from the pulmonary artery as a life-threatening condition
N. Seredyuk, O. Babliak, A. Vytrykhovskyi, R. Petrovskyi, A. Matlah, Y. Vandzhura, O. Tsaruk, I. Grabas, O. Skakun, M. Bielinskyi, I. Tverdokhlib, M. Serediuk 15

Vydavatel
Česká kardiologická společnost, z. s.
Netroufalky 6b, 625 00 Brno
Předseda:
prof. MUDr. Aleš Linhart, DrSc., FESC, FCMA

Vedoucí redaktoři
doc. MUDr. Petr Kala, Ph.D., FESC, FSCAI;
prof. MUDr. Petr Ošťádal, Ph.D., FESC

Odpovědná redaktorka
Mgr. Klára Procházková

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Refrakterní zástava oběhu na podkladě vysoce rizikové plicní embolie u mladého pacienta

Ondřej Sirotek[a], Vratislav Pechman[a], Milan Hromádka[a], Alexandra Žídková[b], Jan Baxa[c], Richard Rokyta[a]

[a] *Kardiologická klinika, Centrum vysoce specializované komplexní kardiovaskulární péče, Lékařská fakulta Plzeň, Univerzita Karlova a Fakultní nemocnice Plzeň, Plzeň*
[b] *Klinika anestezie, resuscitace a intenzivní medicíny (KARIM), Fakultní nemocnice Plzeň, Plzeň*
[c] *Klinika zobrazovacích metod (KZM), Fakultní nemocnice Plzeň, Plzeň*

Klíčová slova:
ECMO
Komplikace
Plicní embolie
Refrakterní zástava oběhu
Trombolýza

SOUHRN

Pacienti s plicní embolií vedoucí k zástavě oběhu mají obvykle špatnou prognózu. Adekvátně zvolená léčba je klíčová. Vedle antikoagulační a trombolytické terapie máme k dispozici oběhovou a ventilační podporu, mechanickou podporu oběhu ve formě venoarteriální extrakorporální membránové oxygenace (V-A ECMO) a chirurgickou či katétrovou embolektomii. Využití těchto léčebných modalit může být spojeno se specifickými komplikacemi. Prezentujeme případ 17letého mladého muže s mimonemocniční zástavou oběhu, kde konvenční resuscitační postupy nevedly k obnově účinného oběhu, a proto byl za pokračující kardiopulmonální resuscitace (KPR) transportován do regionálního ECMO centra. Nález při bed-side echokardiografickém vyšetření byl velmi suspektní z plicní embolie. Byl podán nefrakcionovaný heparin a pacient byl napojen na V-A ECMO. Vyšetřením výpočetní tomografií (CT) byla potvrzena rozsáhlá oboustranná plicní embolie s dilatací pravé komory srdeční (PKS). Pro progredující oběhovou nestabilitu byla podána trombolytická léčba. Nastalé závažné komplikace si vyžádaly opakované chirurgické intervence. Tato kazuistika je zároveň příkladem nezbytnosti multidisciplinární spolupráce.

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Keywords:
Complications
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Refractory cardiac arrest
Thrombolysis

ABSTRACT

Patients with pulmonary embolism leading to cardiac arrest usually have poor prognosis. Adequate treatment is essential. Besides anticoagulant and thrombolytic therapy we can employ circulatory and ventilatory support, mechanical circulatory support like venoarterial extracorporeal membrane oxygenation (V-A ECMO) and surgical or catheter embolectomy. The use of these therapeutic modalities could be connected with specific complications. We present a 17-year-old young man with out-of-hospital cardiac arrest where conventional resuscitation methods were not successful so patient was transported during ongoing cardiopulmonary resuscitation (CPR) to a regional ECMO centre. Bed-side echocardiography was very suspicious of pulmonary embolism. Unfractioned heparin was applied and the ECMO support was initiated. Computed tomography verified extensive bilateral pulmonary embolism with right ventricle dilatation. Patient's hemodynamic status was getting worse so thrombolysis was administered. Severe complications have occurred and led to repeated surgical interventions. This case also demonstrates the necessity of multidisciplinary cooperation.

Adresa pro korespondenci: MUDr. Ondřej Sirotek, Kardiologická klinika, Centrum vysoce specializované komplexní kardiovaskulární péče, Lékařská fakulta Plzeň, Univerzita Karlova a Fakultní nemocnice Plzeň, alej Svobody 80, 304 60 Plzeň-Lochotín, e-mail: siroteko@fnplzen.cz

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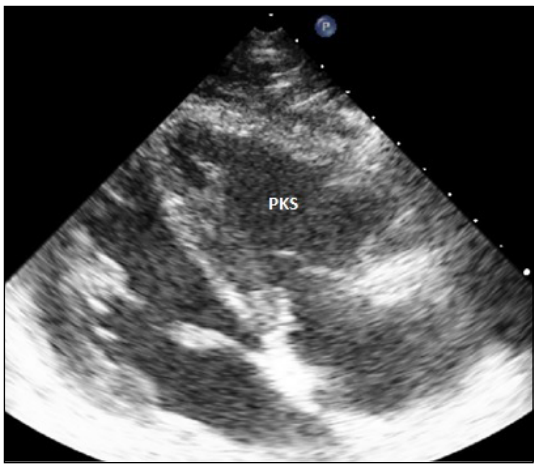
Úvod

U hemodynamicky nestabilních pacientů s vysoce rizikovou plicní embolií je obvykle indikována okamžitá reperfuzní léčba směřující k co nejrychlejšímu zprůchodnění plicnicového řečiště. Nutnost poměrně agresivního přístupu vyplývá z faktu, že u dvou třetin pacientů, kteří zemřou v důsledku plicní embolie, smrt nastává do dvou až tří hodin.[1] Skupina pacientů s nejzávažnější manifestací, kterou je oběhová zástava, může profitovat z iniciální mechanické podpory oběhu ve formě venoarteriální extrakorporální membránové oxygenace (V-A ECMO), kterou můžeme získat čas potřebný k nástupu efektu antitrombotické léčby, eventuálně k rozhodnutí o další léčebné strategii.

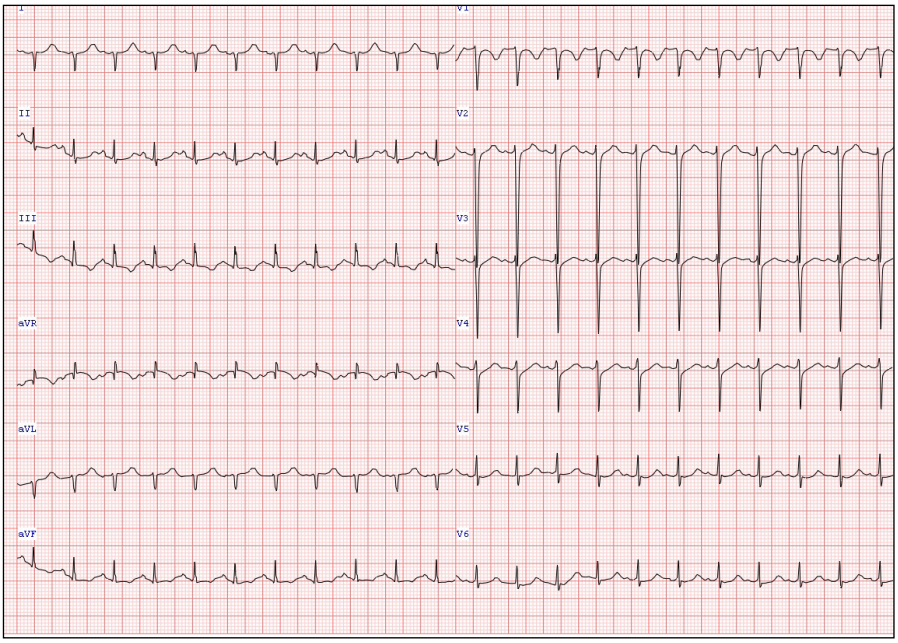
Popis případu

Prezentujeme případ dosud zdravého 17letého mladého muže věnujícího se aktivně lednímu hokeji, který byl v únoru 2019 transportován do FN Plzeň pro refrakterní zástavu oběhu nejasné příčiny. Pacient udával zhruba tři týdny nazpět symptomy infektu dolních cest dýchacích (produktivní kašel, zimnice, pocení), v posledních dnech spojené s progredující dušností, pro kterou byl vyšetřen na plicní ambulanci. Spirometricky byla zjištěna středně těžká obstrukční ventilační porucha, nález byl uzavřen jako spastická bronchitida, byla doporučena léčba bronchodilatancii (ipratropium-bromid + fenoterol) a antileukotrienem. Po návratu z vyšetření došlo doma u pacienta k prudkému zhoršení dušnosti a přechodné poruše vědomí, proto rodina zavolala zdravotnickou záchrannou službu (ZZS). Při jejím příjezdu byl pacient při plném vědomí (Glasgow Coma Scale [GCS] 4-5-6), bez zjevného neurologického deficitu, cyanotický s tachypnoí 25/min, s hraniční saturací krevního hemoglobinu kyslíkem (SpO_2) 92 % na vzduchu, hypertenzí s krevním tlakem (TK) 170/120 mm Hg a se sinusovou tachykardií 140/min na EKG (obr. 1 http://www.cksonline.cz/coretvasa-case-reports/clanky.php?p=detail&id=154&pid=1669&file=1136). Během transportu došlo k rozvoji bradykardie (podaný atropin bez efektu) s následným přechodem do asystolie. Byla bezprostředně zahájena rozšířená KPR, v rámci níž byl pacient zaintubován a napojen na umělou plicní ventilaci. Příčina zástavy oběhu nebyla v daný okamžik zřejmá, pro zvažovaný tenzní pneumotorax byla lékařem ZZS provedena punkce pravé pleurální dutiny. Přes maximální resuscitační úsilí se nepodařilo obnovit účinný oběh a pacient byl za pokračující KPR s použitím mechanizované srdeční masáže transportován do FN Plzeň. V průběhu resuscitace docházelo intermitentně k obnovování oběhu.

Po příjezdu pacienta KPR pro trvající zástavu pokračovala. Bylo provedeno bed-side echokardiografické vyšetření s obrazem přetížení pravé komory srdeční (PKS) (obr. 2 http://www.cksonline.cz/coretvasa-case-reports/clanky.php?p=detail&id=154&pid=1669&file=1135). Pro podezření

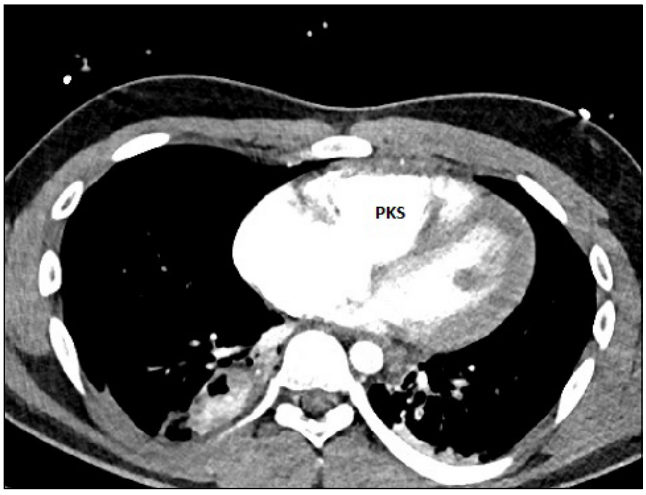


Obr. 2 – Přetížení PKS (modifikovaná SX projekce)

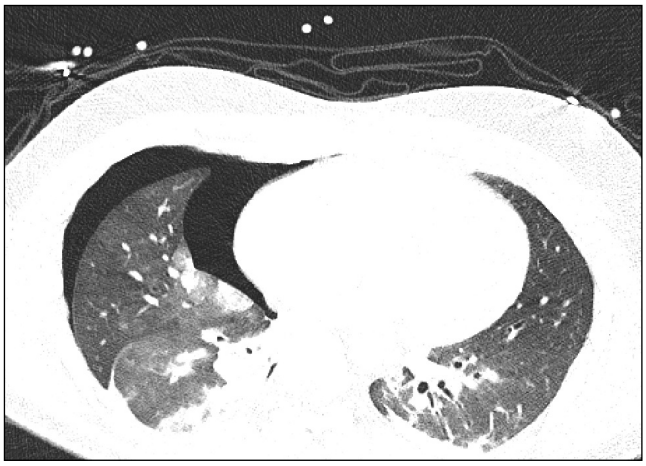


Obr. 1 – EKG se sinusovou tachykardií a vertikálním sklonem srdeční osy

Obr. 3 – Emboly ve větvení plicních tepen. CT dokumentace zveřejněna s laskavostí KZM.



Obr. 4 – Dilatace PKS. CT dokumentace zveřejněna s laskavostí KZM.



Obr. 5 – Pravostranný pneumotorax. CT dokumentace zveřejněna s laskavostí KZM.

na plicní embolii byl podán nefrakcionovaný heparin v dávce 10 000 IU i.v. Vzhledem k přetrvávající neúčinnosti spontánního oběhu a riziku fatálního krvácení po punkci pleury nebyla indikována trombolytická léčba, ale u pacienta jsme indikovali napojení na extrakorporální oběhovou podporu ve formě V-A ECMO. Punkční metodou byla zavedena žilní kanyla (23 Fr) do v. femoralis l. sin. a arteriální kanyla (15 Fr) do a. femoralis l. dx., ECMO bylo spuštěno 12 min od příjezdu do fakultní nemocnice, celkově 84 min od zahájení KPR. Ve vstupní laboratoři byla zjištěna laktátemie > 15 mmol/l, pH 6,8, vysoce senzitivní srdeční troponin T (hs-TnT) 90 ng/l, N-terminální fragment natriuretického propeptidu typu B (NT-proBNP) 8 299 ng/l, C-reaktivní protein (CRP) 20 mg/l, hemoglobin (Hb) 128 g/l, D-dimery > 5 mg/l.

CT vyšetřením byla potvrzena oboustranná plicní embolizace s dilatací PKS (obr. 3 a 4, http://www.cksonline.cz/coretvasa-case-reports/clanky.php?p=detail&id=154&pid=1669&file=1137 a http://www.cksonline.cz/coretvasa-case-reports/clanky.php?p=detail&id=154&pid=1669&file=1138). Dále byl popsán pravostranný pneumotorax (obr. 5 http://www.cksonline.cz/coretvasa-case-reports/clanky.php?p=detail&id=154&pid=1669&file=1139), který byl následně nekomplikovaně zadrenován chirurgem. Ke komplexní poresuscitační péči byl pacient přijat na kardiologickou jednotku intenzivní péče, kde však dále postupně progredovala oběhová nestabilita. K udržení průtoku ECMO byla nutná významná volumexpanze a dávku vazopresorické podpory noradrenalinem bylo nutné navyšovat k 1 µg/kg/min. Echokardiograficky byla patrná dilatace a těžká dysfunkce PKS s ejekční frakcí (EF) 25 %. Při absenci zjevného závažnějšího krvácení bylo přistoupeno k systémovému podání trombolytika alteplázy v redukované dávce 50 mg na 15 min, což vedlo k částečné stabilizaci oběhu, nicméně při echokardiografické kontrole přetrvával neměnný nález těžkého cor pulmonale. Hrudní drenáž odváděla pouze minimální množství hemoragické tekutiny, rovněž třísla byla bez patrného krvácení. Pro opětovnou progresi oběhové nestability byla podána další redukovaná dávka alteplázy 50 mg na 30 min s dobrou hemodynamickou odpovědí. Následně se však stav zkomplikoval výraznými hemoragickými odvody z hrudního drénu s nutností navyšování dávky noradrenalinu až na 3 µg/kg/min a masivní substituce krevními deriváty a koagulačními faktory (erytrocyty, mražená plazma, koncentráty protrombinového komplexu, fibrinogen, rVIIa). Odváděná krev z drénu byla ihned rekuperována. Pacient byl transportován na chirurgický sál, kde byla provedena revize pravé hrudní dutiny s ošetřením zdroje krvácení. Na základě zlepšení systolické funkce PKS (EF 40 %) při perioperačním echokardiografickém vyšetření byla po 25,5 hodinách ukončena podpora ECMO.

V dalším průběhu se stav pacienta komplikoval enteroragií a progredující ischemií pravé dolní končetiny. Na CT byla popsána pneumatóza stěny tenkého střeva a nitkovité zúžení a. femoralis dx., které bylo způsobeno spíše edémem tkání stehna nežli lokálním hematomem. Následně byly při explorativní laparotomii zjištěny pokročilé ischemické změny tenkých kliček s nutností resekce větší části tenkého střeva (ponecháno 120 cm orálního jejuna) a založení terminální jejunostomie. Současně byla provedena fasciotomie pravého stehna a bérce. Nadále trval těžký šok kombinované etiologie s multiorgánovým selháním s anurií a potřebou kontinuální náhrady funkce ledvin (obr. 6 http://www.cksonline.cz/coretvasa-case-reports/clanky.php?p=detail&id=154&pid=1669&file=1140). Vzhledem k vývoji stavu byl pacient po revizi

Obr. 6 – Nemocný v intenzivní péči

prosakující nekrektomie čtvrtý hospitalizační den přeložen na lůžka KARIM.

Na kontrolním CT byla popsána vitalita střeva, tepny dolních končetin se zachovaným tokem a velké žíly bez známek trombózy, ale také edém a bublinky plynu ve svalstvu pravé dolní končetiny. Přes opakované fasciotomie a nekrektomie si sedmý hospitalizační den progrese myonekrózy vyžádala provedení amputace PDK ve stehně. V dalším průběhu se celkový stav pacienta postupně zlepšoval, přetrvávala však potřeba náhrady funkce ledvin. Navzdory protrahované KPR a kritickému stonání se pacient probral k dobrému vědomí a 17. hospitalizační den byl nekomplikovaně extubován. Nejevil známky post-hypoxické encefalopatie, funkce ledvin se zcela zreparovala, velikost a funkce PKS se znormalizovala a známky plicní hypertenze nebyly přítomny. Duplexní sonografie žil ozřejmila starší trombotické hmoty v oblasti levého bérce. Vyšetřením nebyla prokázána trombofilní dispozice. Pacient byl antikoagulován nízkomolekulárním heparinem (LMWH) za kontrol antiXa a později byl převeden na warfarin (přímé perorální antikoagulancium [DOAC] nebylo schváleno vzhledem k nezletilosti). Během hospitalizace na KARIM a následně Chirurgické a Dětské klinice FN Plzeň byly řešeny další komplikace jako neprůchodnost gastrointestinálního traktu, fantomové bolesti (přechodně epidurální analgezie), dehiscence laparotomie, ranné infekce, 47. den hospitalizace byla obnovena kontinuita GIT jejunoascendentoanastomózou. S odstupem pak byla provedena sekundární sutura laparotomie a amputačního pahýlu. K doléčení byl pacient přeložen na Neurorehabilitační oddělení FN Plzeň, kde probíhala komplexní rehabilitace se zhotovenou protézou. Po 113 dnech hospitalizace byl pacient propuštěn do domácí péče. Ve FN Plzeň je nadále dispenzarizován v hematologické a gastroenterologické (syndrom krátkého střeva) ambulanci.

Diskuse

Z uvedené kazuistiky je patrná obtížnost volby optimální léčby. Obecně platí, že v kritických situacích kontraindikace trombolýzy relativizujeme. Guidelines Evropské kardiologické společnosti (ESC) z roku 2019 doporučují při nedosažení iniciální oběhové stability u pacientů s vysoce rizikovou plicní embolií zvážit zavedení V-A ECMO bez předchozí reperfuzní léčby.[2] Pneumotorax u našeho pacienta vznikl jistě punkcí pleurální dutiny lékařem ZZS, nicméně s obdobnou situací se nezřídka setkáme v důsledku fraktury žeber nepřímou srdeční masáží. V případě relativní kontraindikace podání trombolytické terapie pacientům s vysokým rizikem závažného krvácení je akceptovaným postupem podání redukované dávky trombolytika alteplázy 0,6 mg/kg (v max. dávce 50 mg) na 15 min.[1] V našem případě jsme opakovaně podali redukovanou dávku alteplázy pro progredující oběhovou nestabilitu, pro kterou nebyl jiný zjevný důvod.

Dle dostupnosti a zkušeností pracoviště může být alternativou chirurgická či katétrová embolektomie. Data z francouzské práce autorů Meneveau a spol. z roku 2018 srovnávající výsledky jednotlivých léčebných strategií u pacientů s vysoce rizikovou plicní embolií (z celkově 180 pacientů bylo 52 léčeno pomocí ECMO, z toho 13 pro refrakterní kardiogenní šok a 39 pro srdeční zástavu, z nich 13 pro refrakterní srdeční zástavu) ukazují slibné výsledky kombinace ECMO s embolektomií.[3]

Přes použití menšího průměru tepenné kanyly (15 Fr) a relativně krátkou dobu trvání podpory ECMO vedlo těžké ischemicko-reperfuzní poškození ke compartment syndromu pravé dolní končetiny a ani opakované fasciotomie a nekrektomie nezabránily nutnosti vysoké amputace končetiny. Preventivní zajištění distální perfuze dolní končetiny by mělo být zváženo u většiny pacientů. Dle doporučení Extracorporeal Life Support Organization (ELSO) z roku 2021 není zavedení distální perfuze prioritou úvodní léčby, ale mělo by ideálně být provedeno do čtyř hodin.[4,5]

Ischemii tenkého střeva, která vyústila v resekci jeho větší části, dáváme do souvislosti s těžkým šokovým stavem. Podíl trombózy na ischemii střeva (eventuálně i dolní končetiny) nelze vyloučit, nemáme však pro něj důkazy.

Příčinu vzniku tromboembolické příhody u mladého a jinak zdravého pacienta se nepodařilo uspokojivě objasnit. Nebyla zjištěna anamnéza úrazu či imobilizace, užívání anabolik ani jiných rizikových látek, nebyla prokázána trombofilní dispozice.

K prakticky intaktnímu neurologickému výstupu přes kritické stonání nepochybně přispěl nízký věk pacienta.

Závěr

Osud pacientů s plicní embolií a oběhovou zástavou závisí na včasné diagnostice, optimálně zvolené strategii a dostupnosti léčby. Péče o tyto pacienty s sebou může přinášet řadu potenciálně závažných komplikací, které je často nutné řešit multidisciplinárně.

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Odkaz na článek online
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CURRICULUM VITAE

MUDr. Ondřej Sirotek absolvoval studium na Lékařské fakultě Univerzity Karlovy v Plzni v roce 2009. Téhož roku nastoupil na II. interní kliniku Fakultní nemocnice Plzeň, kde v roce 2012 dokončil interní kmen. Od roku 2014 doposud pracuje na Kardiologické klinice FN Plzeň. Jeho hlavním zájmem v rámci oboru je akutní kardiologie.

Thoracic aortic rupture associated with multiple aneurysms of medium-sized arteries: case report

Kristina Rücklová[a,b], Eva Honsová[c], Petr Tomášek[d]

[a] *Department of Pediatrics, 3rd Faculty of Medicine, Charles University and University Hospital Královské Vinohrady, Prague*
[b] *Department of Pediatrics and Inherited Metabolic Disorders, General University Hospital in Prague, Prague*
[c] *AeskuLab Pathology, Prague*
[d] *Institute of Forensic Medicine, 2nd Faculty of Medicine, Charles University and University Hospital Bulovka, Prague*

Klíčová slova:
Aneurysma a disekce hrudní aorty
Koronární a intrakraniální aneurysmata
Příčiny

SOUHRN

Úvod: Aneurysma a disekce hrudní aorty představují vzácnou, ale důležitou příčinu úmrtí v mladém věku. Tato kazuistika popisuje případ mladého muže, který zemřel na tamponádu srdce při ruptuře hrudní aorty. Pitva zároveň odhalila mnohočetná aneurysmata středně velkých tepen. V kazuistice jsou diskutovány možné příčiny těchto mnohočetných cévních anomálií, především dědičná onemocnění pojiva. V případě suspektní dědičné příčiny je v rámci vyšetření post mortem indikována molekulární pitva a kardiogenetické poradenství v rodině.
Popis případu: Devětadvacetiletý muž zemřel náhle během letu na rupturu hrudní aorty. Pitva kromě srdeční tamponády odhalila i mnohočetná aneurysmata bazilárních a koronárních tepen s trombotickým uzávěrem ramus interventricularis anterior. Kromě geneticky podmíněné vaskulopatie mohou být příčiny podobných nálezů různorodé – od aterosklerózy a arteriální hypertenze až po různé typy vaskulitid. Popisujeme klinické a patologické charakteristiky jednotlivých onemocnění, aby se dostaly do povědomí kliniků i patologů.
Závěr: Ruptura hrudní aorty s asociovanými aneurysmaty nebo bez asociovaných aneurysmat středně velkých tepen u mladé osoby zasluhuje zvláštní pozornost, protože může být způsobena potenciálně dědičnou vlohou. Ta se navíc dědí autosomálně dominantním způsobem a může znamenat zvýšené riziko náhlé smrti u prvostupňových příbuzných. V našem případě jsme genetickou příčinu potvrdit nemohli, protože nebyla k dispozici vhodná tkáň pro analýzu DNA. Právě proto tato kazuistika podtrhuje, jak je důležité na dědičná onemocnění pojiva pomýšlet a uchovat vhodnou tkáň pro molekulární pitvu.

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Keywords:
Causes
Coronary aneurysms
Intracranial aneurysms
Thoracic aortic disease

ABSTRACT

Background: Thoracic aortic disease is a rare but important cause of death that may affect young individuals. We present a case of a young man who died of thoracic aortic rupture associated with multiple aneurysms of medium-sized arteries. Possible causes of this multivascular pathology are discussed here with emphasis on potential genetic etiology and its implications for post mortem investigations including molecular autopsy and cardiogenetic counselling in the family.
Case presentation: A 29-year-old man died suddenly due to thoracic aortic rupture during an air flight. His autopsy, in addition, revealed multiple aneurysms in the basilar and coronary arteries with a thrombotic occlusion of an aneurysm in the left anterior descending artery. Apart from genetic etiology the causes of similar findings range from atherosclerosis and hypertension to various inflammatory diseases. The clinical and pathological features of relevant diseases are outlined here to raise awareness of them among clinicians as well as pathologists.
Conclusions: The finding of thoracic aortic rupture in a young person with or without associated medium-sized artery aneurysms deserves special attention due to its potentially heritable etiology with an increased risk of sudden death for first degree relatives. In our case we could not confirm genetic etiology due to lack of suitable material for DNA analysis. We therefore emphasize the importance to consider heritable connective tissue disorders in similar cases and to retain suitable tissue for molecular autopsy.

Address: MUDr. Kristina Rücklová, Ph.D., Department of Pediatrics, University Hospital Královské Vinohrady, Šrobárova 1150/50, 100 34 Prague 10, e-mail: kristina.rucklova@fnkv.cz

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Introduction

Thoracic aortic disease (TAD) with rupture is an important cause of sudden death accounting for approximately 7% of sudden cardiac death cases among people aged 1–35 years.[1] Major risk factors for TAD include ageing, increased mechanical forces acting on aorta such as poorly controlled hypertension or stimulant abuse and an underlying genetic predisposition that disrupts aorta's elastic properties rendering it more susceptible to dilation and dissection.[2] Especially in young persons, a heritable predisposition is predominantly responsible for TAD.[3] Other possible causes of thoracic aortic aneurysm and rupture in young individuals include infectious or non-infectious inflammation, with Takayasu arteritis being the most common vasculitis that affects aorta in patients below 50 years of age.[4]

Patients with heritable or inflammatory TAD may develop aneurysms elsewhere in the arterial tree including coronary and cerebral arteries. However, except for reports on Loeys–Dietz and the vascular type of Ehlers–Danlos syndrome there is a limited number of reports addressing this association.[5–7]

The patient presented here was recruited from a retrospectively collected cohort of sudden cardiac death cases aged 1–40 years autopsied in the Czech Republic. He was the youngest TAD case in our cohort and stands out due to the unusual autopsy finding of TAD associated with multiple aneurysms in the basilar and coronary arteries. The exact etiology of these morphological changes could not be determined as no data regarding his previous medical history were available and genetic testing could not be performed due to lack of suitable tissue. Nevertheless, this case report provides us with an opportunity to discuss the differential diagnosis of such findings and consider practical implications for post mortem investigations in similar cases.

Case presentation

A 29-year-old healthy looking man of Caucasian ethnicity collapsed suddenly during an airplane flight in 2015. Cardiopulmonary resuscitation provided by the airplane personnel was unsuccessful. His ECG showed asystole with no reaction to repeated adrenaline administration.

Autopsy findings

The young man was 186 cm (+0.8 SD above average) tall and obese (BMI 37). His fingers were remarkably long and thin. He had normal facial features, no corneal opacities, no palatal cleft, bifid uvula or chest deformity, nor did he have any other skeletal abnormalities. Apart from two hypopigmented scars on the left shoulder and arm, his skin was unremarkable. It was not particularly thin or translucent and no striae were noticeable. Except for resuscitation induced rib fractures with right lung hematoma and brain and lung edema, no pathological organ involvement was noted. Increased fragility of intestinal wall was not observed and the patient had no liver or kidney cysts.

Thoracic aortic rupture within an aneurysm with cardiac tamponade was revealed as the cause of death. The aneurysm was saccular 3 × 1.5 × 1.5 cm in size and located above the aortic valve. The aortic wall rupture extended 4 cm in length and led to cardiac tamponade. The pericardial sac contained 750 ml of blood. The aorta contained only minor fibrous plaques on macroscopic examination. The aortic valve was tricuspid.

Additional pathological findings included multiple fusiform basilar artery aneurysms along the whole length of the vessel with one aneurysm 1 × 0.5 cm in diameter. Cerebral arteries showed no significant atherosclerosis.

Another fusiform aneurysm was revealed in the left anterior descending (LAD) artery near the branching of the left main coronary artery, where the vessel circumference reached 20 mm and the aneurysm was completely occluded with an old organized thrombus of 3.5 × 2 × 1.3 cm. A pale post-ischemic intramural scar of 2 × 1 cm was noted in the interventricular septum. The scar signified a past myocardial infarction that could have been related to the thrombus in LAD. Circumflex coronary artery was also locally dilated. Minor atherosclerotic plaques were present in the coronary arteries that caused only insignificant stenoses of their lumen.

Abdominal aorta and other arteries did not show aneurysmal dilations and/or atherosclerotic changes.

The heart was concentrically hypertrophic. Its weight of 680 g exceeded the predicted BSA related normal heart weight of 455.3 g (range 370.1–540.6 g)[8] and the left ventricular wall thickness was 16 mm also slightly exceeding two standard deviations above the mean of 12.6 mm.[9] Myocardial disarray pathognomonic for hypertrophic cardiomyopathy was not present.

The patient was tested negative for alcohol.

Histological examination

Aortic and coronary artery specimens were stained with hematoxylin-eosin (HE) and Van Gieson (VG) with modification for additional elastin detection.

Hemorrhage and smooth muscle disorganization was observed in the specimens of the aortic wall as well as elastic fiber fragmentation, focal elastic fiber loss and focal mucoid extracellular matrix accumulation[10] (Fig. 1A and Fig. 1B, http://www.cksonline.cz/coretvasa-case-reports/clanky.php?p=detail&id=155&pid=1669&file=1141a http://www.cksonline.cz/coretvasa-case-reports/clanky.php?p=detail&id=155&pid=1669&file=1142). There were no atherosclerotic changes and no intimal calcifications in the aortic wall.

The specimen of the left anterior descending coronary artery showed an aneurysm with an organized thrombus. Neither aorta nor coronary arteries contained inflammatory infiltrates, granulomas or giant cells that would indicate inflammatory etiology.[11,12]

Discussion

The patient reported here had a cardiac hypertrophy, which may suggest arterial hypertension as one of the

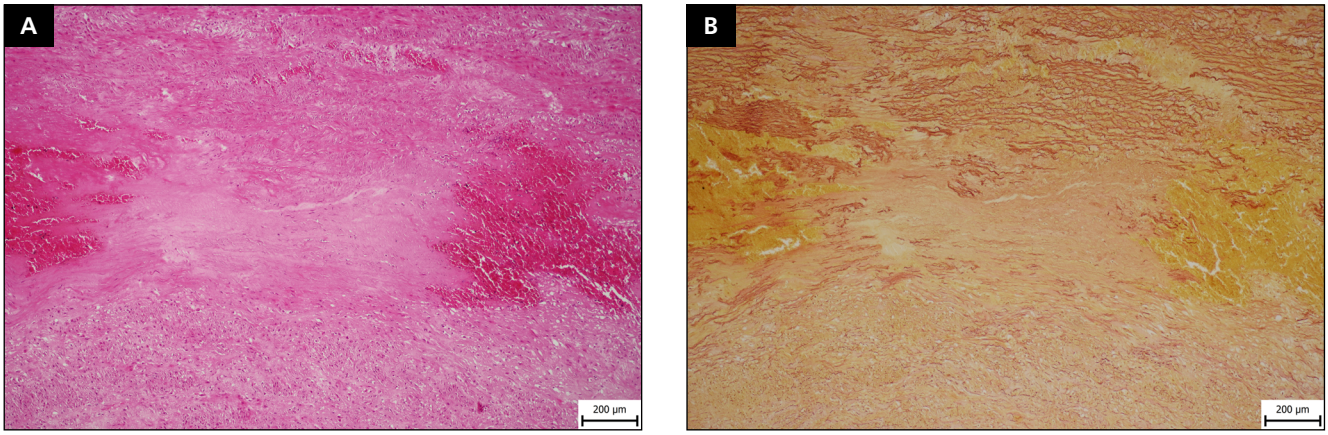

Fig. 1 – (A) Aortic wall with hemorrhage among the laminar layers of the media and smooth muscle disorganization (HE stain). (B) Aortic wall with focal elastic fiber loss and fragmentation creating focal translamellar mucoid extracellular matrix accumulation (VG stain).

major risk factors for thoracic aortic rupture. However, his young age, normal kidneys with smooth surface and additional vascular pathology with multiple basilar and coronary aneurysms make hypertension as a sole cause of these findings rather unlikely.

Atherosclerosis is the most common cause of coronary aneurysms[13] and may be associated with aortic aneurysm and rupture. However, in contrast to abdominal aortic aneurysm, thoracic aortic aneurysm due to atherosclerosis is very rare.[14] In addition, our patient was young and almost no atherosclerotic changes were observed in his aorta.

The young age of the patient suggests rather a genetic or inflammatory etiology of the vascular damage. Considering inflammatory diseases, the most likely culprit could be Kawasaki disease, the second most common cause of coronary aneurysms in adults. However, it has only rarely been associated with thoracic aortic aneurysms[15,16] and thoracic aortic rupture has never been reported. Aneurysms in other medium-sized systemic arteries occur in about 2% of Kawasaki disease patients[17] and their most frequent locations are brachial, axillary and iliac arteries.[18] On the other hand, intracranial aneurysms have been observed only exceptionally. The inflammatory process actually seems to spare intracranial arteries.[12] In our patient, the histological findings together with these facts make the diagnosis of Kawasaki disease very unlikely.

Another vasculitis, which typically affects aorta as well as coronary arteries, is Takayasu arteritis. It may manifest with thoracic aortic rupture and coronary arteries are involved in as many as 53% of these patients. Coronary aneurysms are present in 8% and coronary stenoses in the rest.[19] Intracranial aneurysms including basilar aneurysms have been described in Takayasu arteritis as well.[20] However, Takayasu arteritis typically affects young women of Asian ethnicity. The initial stage of the disease is clinically characterized by symptoms of systemic inflammation or acute aortic insufficiency. Transmural granulomatous inflammation with varying number of giant cells is typically found in histopathological specimens at this stage. The chronic phase is characterized by intimal fibrosis leading to vascular obstruction.[4,11] However, the histopathological findings in our patient exclude the diagnosis of Takayasu arteritis.

Vasculitis involving large as well as medium-sized arteries may also occur in IgG4-related disease.[11,21] However, this disease is usually manifested by tumor-like lympho-plasmocytic infiltrations in various tissues especially pancreas, liver, kidneys, salivary gland or orbital region, which was not observed in our patient. It mostly affects persons more than 50 years old,[22] although much younger patients have also been reported.[23] Autopsy findings and histopathological evaluation rule out this condition in our patient.

Similarly, we can also exclude ANCA-associated vasculitides which may though exceptionally involve large vessels including aorta.[24]

By exclusion and with respect to the young age of our patient we speculate that the most likely underlying etiology of the aortic rupture and multiple medium-sized artery aneurysms might be a pathogenic variant in one of the genes associated with heritable TAD (HTAD). Approximately 10% of patients with HTAD have a concomitant intracranial aneurysm[25] and coronary aneurysms have also been reported[5–7] supporting the possibility of a genetic cause in our patient.

Depending on the presence or absence of involvement of other organ systems, HTAD may be subdivided into syndromic and isolated HTAD. So far 11 genes, whose pathogenic variants confer a very high risk of TAD, have been identified. They encode proteins involved in maintaining elasticity and strength of the medial layer of the aorta, which is composed mainly of smooth muscle cells and elastin. On the cellular level, these proteins are mainly implicated in smooth muscle cell contraction and adhesion to the extracellular matrix (e.g. *FBN1, COL3A1, ACTA2, MYH11, MYLK, PRKG1, MFAP5*), transforming growth factor-β signaling pathway (e.g. *TGFBR1/2, SMAD3/4*) or smooth muscle cell metabolism.[2] Pathogenic variants in these genes may be found in syndromic connective tissue disorders associated with TAD such as Marfan, Loeys–Dietz or vascular type of Ehler–Danlos syndrome as well as in isolated HTAD cases.

Our patient exhibited only arachnodactyly as a characteristic feature of syndromic form of HTAD. He had no other typical musculoskeletal or ocular abnormalities nor did he exhibit less common features such as liver or

kidney cysts associated with Marfan syndrome.[26] His habitus was not asthenic. This latter feature had historically been considered typical for Marfan syndrome but more recently it has been documented that as many as 26% of Marfan syndrome patients are obese.[27] Hence, Marfan syndrome cannot be excluded in our patient as it is the most common syndromic connective tissue disorder associated with TAD. The other above mentioned syndromes may be associated with aneurysms throughout the arterial tree but their characteristic features such as bifid uvula, hypertelorism, translucent and hyperelastic skin, atrophic scars, chest deformities, short or tall stature, joint hypermobility or contractures[3] were absent in our patient.

As no pronounced extravascular anomalies were observed, we consider non-syndromic HTAD to be the most likely diagnosis in our patient. We may hypothesize about a possible role of certain genes as some have been associated with TAD and additional vascular complications. For example, pathogenic variants in *TGFBR2* gene predispose to TAD as well as intracranial and other arterial aneurysms. Another highly suspicious gene could be *ACTA2* as its pathogenic variants lead to TAD together with early-onset coronary artery disease as seen in our patient who had a thrombotic occlusion of the left anterior descending artery.[2] Pathogenic variants in *ACTA2* gene are responsible for 14% of non-syndromic HTAD, thereby being the most frequently identified molecular mechanism in this subgroup.[28]

The greatest limitation of this case report is the fact that genetic testing could not be performed due to lack of tissue for DNA analysis. All paraphin blocks with myocardial samples were used for preparation of histological specimens. Another limitation is absence of medical records.

Conclusions

TAD in a young person should always be suspected of genetic etiology. In fact, approximately 20% of TAD are familial with an autosomal dominant pattern of inheritance thus conferring a 50% risk for first degree relatives.[3] Further deaths in the families may be prevented if individuals at risk are identified and managed in time. Therefore, tissue from the deceased person should be retained for DNA analysis and the relatives should receive an adequate cardio-genetic counselling including a cascade genetic screening.[29,30] In order to implement this approach into routine practice, guidelines for post mortem investigations of sudden cardiac death cases were approved by the Czech Forensic Medicine Society in 2020.

This case report provides forensic pathologists as well as clinicians with an overview of differential diagnosis of multi-vascular aneurysms, which may help them select patients for targeted genetic testing. The report also contributes to increased awareness of heritable cardiovascular diseases, which may prompt general practitioners to indicate cardio-genetic care in families with sudden cardiac death in the young. Lastly, the case presented here demonstrates that TAD may be associated with aneurysms elsewhere in the arterial tree especially in coronary arteries.[31] This fact is not broadly recognized and coronary arteries are not routinely checked during follow up of young patients with TAD.

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Conflicts of interest
None.

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Ethical statement
Ethics committee's approval was not required as all patient's data were strictly de-identified. The study did not involve living subjects nor did it involve any genetic analyses.

Authors' contributions
All authors contributed to the design of the article. Kristina Rücklová was responsible for writing the manuscript. Eva Honsová assessed the histological samples and suggested the differential diagnosis of the histological findings. Petr Tomášek evaluated the autopsy protocol. Both co-authors commented on the first version of the manuscript and all authors approved the final version.

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CURRICULUM VITAE

MUDr. Kristina Rücklová, Ph.D.

Education and qualification: qualification in clinical pediatrics (2nd degree) – Nov 2010; qualification in pediatric cardiology – Jun 2009; qualification in clinical pediatrics (1st degree) – Jun 2005; Ph.D. studies in physiology and pathophysiology, Ph.D. degree, 3rd Faculty of Medicine, Charles University, topic: "Retrospective diagnosis of unrecognized causes of sudden death in infancy" – 2002–2008; 1st Faculty of Medicine, Charles University, Prague, M.D. degree – 1995–2002; Faculty of Medicine, Ludwig Maximilians University, Munich, Germany – 1999–2000; Faculty of Philosophy – English Philology, Charles University, Prague – 1995–1999

Clinical experience: pediatric cardiologist, part-time, Dept. of Pediatrics and Inherited Metabolic Disorders, General University Hospital, Prague – 2018 – now; chief physician at the unit for children up to 3 years of age and pediatric cardiologist, University Hospital Královské Vinohrady, Prague – 2014 – now; pediatric cardiologist, Children's Heart Centre, Faculty Hospital Motol, Prague – 2011–2014; pediatrician and pediatric cardiologist, Dept. of Pediatrics, Faculty Hospital Motol, Prague – 2008–2011; resident in pediatrics, Dept. of Pediatrics, Faculty Hospital Královské Vinohrady, Prague – 2002–2007

Free-floating thrombus in the left atrium

Sandra Rečičárová, Michael Jonák

Department of Cardiovascular Surgery, Institute of Clinical and Experimental Medicine, Prague

Klíčová slova:
Levá síň
Mitrální stenóza
Volně vlající trombus

SOUHRN

Volně vlající trombus v levé síni je vzácný stav, který se často vyskytuje při onemocnění mitrální chlopně. Dovolujeme si prezentovat případ 82leté pacientky s volně vlajícím trombem v levé síni, který byl náhodně odhalen při předoperačním transezofageálním echokardiografickém vyšetření u pacientky s onemocněním mitrální chlopně a perzistentní fibrilací síní. Pacientka užívala 15 let antikoagulační terapii. Závěrem diskutujeme různé možnosti, které vedly k vzniku trombu.

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Keywords:
Free-floating thrombus
Left atrium
Mitral stenosis

ABSTRACT

A free-floating thrombus in the left atrium is a rare condition that usually occurs with mitral valve disease. We present a case of an 82-year-old woman with the free-floating left atrial thrombus which was found incidentally on the preoperative transesophageal echocardiogram in the patient with mitral valve disease and persistent atrial fibrillation. The patient has been on anticoagulation therapy for 15 years. We discuss different possibilities that led to the formation of the thrombus.

Introduction

Free-floating thrombus in the left atrium is a condition that usually occurs with mitral valve disease and atrial fibrillation. It has potentially catastrophic consequences such as obstruction of the stenotic mitral valve orifice causing sudden death or systemic embolization. A transesophageal echocardiogram is the most useful diagnostic tool and once the diagnosis is established, surgical removal of the thrombus with the treatment of the underlying cause is mandatory.

Case report

An 82-year-old woman with a medical history of rheumatic mitral valve disease, persistent atrial fibrillation, and previous cerebral infarction was admitted to our hospital with progressively increasing dyspnea.

The patient was already diagnosed with severe mitral valve stenosis and moderate mitral regurgitation, severe pulmonary artery hypertension, and moderate tricuspid regurgitation. The patient previously suffered a stroke in 1995 from which she made a full recovery and a peripheral thrombo-embolism in the right leg in 1997. Since then she was treated with dicumarol, digitalis, diuretics, calcium channel blockers, and angiotensin II receptor blockers.

Transthoracic echocardiogram has been done as a part of regular follow-up for mitral valve disease every two years since 1998. The transesophageal echocardiogram performed in July 2018 showed progression of the mitral valve stenosis, normal ejection fraction of left ventricle, spontaneous echo contrast in the gigantic left atrium (LAVi 350.8 cm^3) with no mention of thrombus neither in atrium nor in appendage, and moderate tricuspid regurgitation. The patient was subsequently indicated for mitral valve replacement, tricuspid valve annuloplasty, and left atrial appendage closure.

Address: MUDr. Sandra Rečičárová, MBA, Department of Cardiovascular Surgery, Institute of Clinical and Experimental Medicine, Vídeňská 1958/9, 140 00 Prague 4, e-mail: sandra.recicarova@ikem.cz

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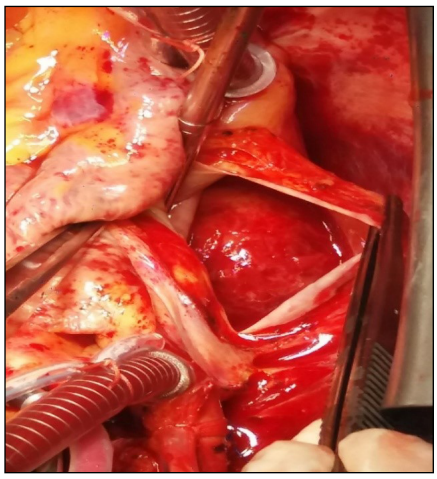

Fig. 1 – Intraoperative finding. After incision of the left atrium, a smooth round thrombus was visualised.

A transesophageal echocardiogram performed at the beginning of surgery in February 2019 showed a round-ball-like mobile thrombus freely floating in the left atrial cavity. During the cardiac cycle, the mass dropped into the mitral orifice, obstructing it temporarily, before bouncing away again towards the posterior left atrial wall (Video 1, 2 http://www.cksonline.cz/coretvasa-case-reports/clanky.php?p=detail&id=156&pid=1669&file=1146, http://www.cksonline.cz/coretvasa-case-reports/clanky.php?p=detail&id=156&pid=1669&file=1145). Fortunately, the size of the mass and the mitral valve stenosis seemed to prevent it from systolic embolization. At surgery, a smooth round thrombus (Fig. 1 http://www.cksonline.cz/coretvasa-case-reports/clanky.php?p=detail&id=156&pid=1669&file=1143). was removed and the patient had a mitral valve replacement with the biological prosthesis, DeVega annuloplasty of the tricuspid valve, and left atrial appendage closure. Anticoagulation was resumed the first postoperative day and the patient was discharged after stabilizing the anticoagulation dosage.

Pathology

The thrombus measured 37 mm and weighed 30 g (Fig. 2 http://www.cksonline.cz/coretvasa-case-reports/clanky.php?p=detail&id=156&pid=1669&file=1144). It was yellow-pinkish colored, soft in consistency with noticeable layering and scarring tendency. Histologically, it was described as a round encapsulated thrombus with layers of thrombotic substances inside and with the signs of organization and seldom epithelization of endothelial cells on the outside. Immunohistochemical verification using antibody calretinin was negative, which ruled out the diagnosis of myxoma.

Fig. 2. – The thrombus outside of the left atrium.

Discussion

We present a case of a free-floating left atrial thrombus that was found incidentally on the preoperative transesophageal echocardiogram in a patient with severe mitral valve stenosis, moderate mitral valve regurgitation, and persistent atrial fibrillation. The patient was anticoagulated with dicumarol since suffering a stroke and a peripheral thrombo-embolism in the right leg 15 years ago. This suggests the previous presence of thrombi in the left atrium and since the mitral valve disease wasn't severe enough at that time, embolic manifestations could have easily occurred. This time, anticoagulant therapy might have been insufficient which caused the formation of the thrombus in the relatively short period of six months between the last transesophageal echocardiogram as a part of regular follow-up without the presence of the thrombus and our preoperative transesophageal echocardiogram where the massive thrombus was freely floating in the left atrial cavity.

Hewitt formulated two criteria for the diagnosis of a free ball thrombus: the thrombus must be larger than the orifice of the valve, and it must have a smooth surface with no signs of attachment to the atrial wall.[1] At the beginning of its formation, the thrombus was either mural micro thrombus, or it was pedunculated and it later lost the attachment to the atrial wall because of the constant motion. The other hypothesis is that it was an initial free-floating thrombus where the stenotic mitral valve didn't allow it to enter its orifice with an accretion phenomenon gradually enlarging the mass like a growing snowball in the background of chronic stasis associated with atrial fibrillation.[2] Its formation usually starts in the left atrial appendage or the left atrial posterior wall. The thrombus comes from fresh clots covered concentrically by a smooth fibrinous layer which together with the incomplete emptying of the left atrium and the blood's tendency for rotary motion prevents it from systemic embolization.[3] Such large thrombi are unlikely to be dissolved with anticoagulation and therefore the first choice of treatment is its surgical removal.

Conflict of interest
None declared.

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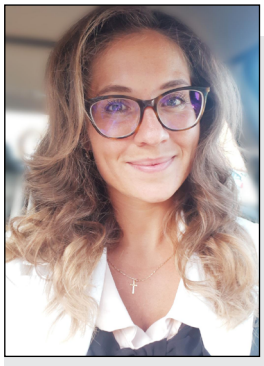

CURRICULUM VITAE

MUDr. Sandra Rečičárová, MBA, graduated from Faculty of Medicine at University of Pavol Jozef Šafárik in 2018. She is currently practicing in the Department of Cardiovascular Surgery at the Institute of Clinical and Experimental Medicine in Prague. She began her Ph.D. studies in experimental surgery at Charles University in 2020.

Anomalous origin of the left coronary artery from the pulmonary artery as a life-threatening condition

Nestor Seredyuk[a,c], Oleksandr Babliak[b], Andrii Vytrykhovskyi[a,c], Roman Petrovskyi[a,c], Andrii Matlah[c], Yaroslava Vandzhura[a,c], Oleksandra Tsaruk[d], Iryna Grabas[c], Oleksii Skakun[a], Mykyta Bielinskyi[a], Ivan Tverdokhlib[a], Marta Serediuk[a]

[a] *National Medical University, Ivano-Frankivsk, Ukraine*
[b] *Medical Center "Dobrobut", Kyiv, Ukraine*
[c] *Regional Clinical Cardiology Center, Ivano-Frankivsk, Ukraine*
[d] *Medical Center «Doctor Tsaruk», Ivano-Frankivsk, Ukraine*

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Sudden cardiac death
Surgical correction

ABSTRACT

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA), also called Bland–White–Garland syndrome, is a rare congenital coronary artery abnormality that often manifests itself by chronic myocardial ischemia, angina during breastfeeding, myocardial infarction, sudden cardiac death. This article reports on the ALCAPA in an adult man. The patient complained of chest discomfort for a long time, a silent myocardial infarction was suspected. When he was 29 years old, he developed out-of-hospital cardiac arrest while exercising (treadmill at the gym) with successful electrical cardioversion and resuscitation. ALCAPA diagnosis was verified using computed tomography coronary angiography (CTCA). Left anterior minimally invasive thoracotomy was performed. Left coronary artery (LCA) trunk was ligated, cut, and sewed, distal LIMA-LAD anastomosis was made. Two weeks after the surgical correction a postpericardiotomy syndrome developed, which was successfully treated using methylprednisolone and ibuprophene.

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Introduction

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA or Bland–White–Garland syndrome) is a rare congenital heart anomaly with an incidence of 1 in 4243 live births or 0.023%.[1] About 10–15% of all cases are diagnosed in adult age.[2] There are two types of this syndrome: an infantile type that is characterized by poor collaterals between the left coronary artery (LCA) and the right coronary artery (RCA), and adult type in which collaterals are well-developed.[3,4] Symptoms of ALCAPA in adults include angina, myocardial infarction, papillary muscle calcification, endocardial fibroelastosis, mitral regurgitation, heart failure.[3,5] Chronic myocardial hypoperfusion often leads to the dilation of the left ventricle and reduced contraction of its walls resulting in a decrease in the left ventricular ejection fraction.[6]

Case report

A 29-year-old patient K. was asymptomatic in childhood. The heart murmur was detected when the patient was 15 years old. ECG and echocardiography were performed and no pathological changes were found. Later left-sided chest discomfort appeared. Repeated ECG and echocardiography showed no pathological changes. The patient finished medical university and became a dentist.

The patient suddenly developed ventricular tachycardia/ventricular fibrillation during exertions on the treadmill at the gym. Prompt resuscitation was started, consequent two defibrillation procedures were performed and sinus rhythm was restored. On a 12-lead ECG performed after resuscitation a sinus rhythm with ventricular extrasystoles is seen, probably signs of previous antero-septal myocardial infarction (qR in I, avL; Qr in V_2), ST elevation

Address: Prof. Nestor Seredyuk, MD, National Medical University and Regional Clinical Cardiology Center, Ivano-Frankivsk, Ukraine, e-mail: seredyuknestor@gmail.com

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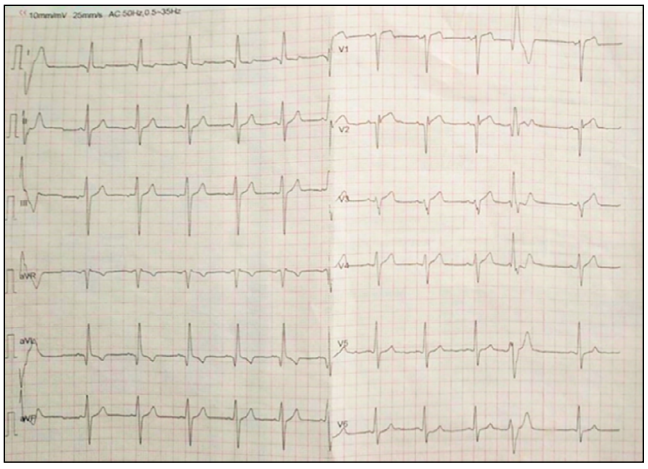

Fig. 1 – ECG recorded from the patient K.: sinus rhythm, ventricular extrasystole, possible anteroseptal myocardial infarction, signs of left ventricular hypertrophy.

in V_1–V_3, signs of left ventricular hypertrophy (Fig. 1 http://www.cksonline.cz/coretvasa-case-reports/clanky.php?p=detail&id=157&pid=1669&file=1147). CT coronary angiography revealed the origin of the LCA from the proximal segment of the pulmonary artery, recessive and convoluted RCA originated from the right Valsalva sinus (Fig. 2 http://www.cksonline.cz/coretvasa-case-reports/clanky.php?p=detail&id=157&pid=1669&file=1148).

A left anterior mini-thoracotomy was performed. The left internal mammary artery was harvested. The left main coronary artery was tied with two ligatures and sewn. Coronary artery bypass grafting LIMA-LAD was performed. There was a normal early postoperative period. But in two weeks the patient developed a fever and tachycardia. Echocardiography revealed hyperechogenic pericardial effusion with a separation of the pericardial layers of 1 cm. PaO_2 decrease to 92–94% was registered. The postpericardiotomy syndrome was diagnosed. Methylprednisolone (32 mg a day with gradual dose reduction by 2.5 mg every two weeks), ibuprophene (600 mg a day), bisoprolol (2.5 mg a day), aspirin (100 mg a day), clopidogrel (75 mg a day), indapamide (1.25 mg a day) were prescribed. There was normal recovery at follow-up at 1, 3, and 6 months, and the patient returned to work.

Discussion

ALCAPA is a severe congenital heart anomaly. Its course may be unpredictable and dramatic, as described in this case. ALCAPA, rather than coronary atherosclerosis, resulted in a dramatic event. Left ventricular myocardium was receiving poorly oxygenated blood with high carbon dioxide content for a long time. For a certain period of time, collaterals between the LCA and the RCA provided myocardial viability, but exertions causing the imbalance between the myocardial oxygen demand and its delivery, has led to the type 2 myocardial infarction (according to the Fourth Universal Definition of Myocardial Infarction, ESC, 2018).[7] Silent anteroseptal myocardial infarction was complicated by ventricular fibrillation and sudden clinical death. The imbalance between the oxygen delivery to the myocardium and its demand occurs in the case of ALCAPA.

In infants, ALCAPA is characterized by intermittent anxiety paroxysms, significant pallor, vomiting, significant sweating during breastfeeding ("breastfeeding angina").[8] Coronary artery aneurysm development can occur in ALCAPA patients.[9] Episodes of chest discomfort in this patient were considered with some skepticism which almost brought him to a fatal outcome.

This case supports an opinion, that ALCAPA is not only a theoretical problem but also a serious problem in real clinical practice. Most cardiac symptoms of ALCAPA are caused by impaired metabolism due to chronic myocardial hypoxia. The imbalance between oxygen delivery and myocardial demand significantly increases upon exertions. It's the background for the development of chronic coronary syndrome, myocardial infarction, sudden cardiac death. Coronary angiography and CT coronary angiography, both are reliable methods for the establishment of the ALCAPA diagnosis.[10,11] Ligation and sewing of the left main coronary artery near its origin from the pulmonary artery and coronary artery bypass grafting LIMA-LAD is an effective method of the surgical correction of the ALCAPA.

Conclusion

ALCAPA is a congenital coronary artery anomaly that can be diagnosed not only in children but in adults as well.

Fig. 2 – CT coronary angiography of the patient K., 29 y. o.

This anomaly may lead to myocardial infarction and sudden cardiac death. So, early diagnostics of this condition is very important. Coronary angiography and CT coronary angiography are reliable methods for confirmation of the ALCAPA diagnosis. Early surgical correction may prevent severe complications. The possibility of the postpericardiotomy syndrome should also be kept in mind, which requires timely pharmacological treatment (methylprednisolone).

Conflict of interest
None.

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Ethical statement
I declare on behalf of all authors that the research was conducted according to the Declaration of Helsinki.

Informed consent
I declare on behalf of all authors that informed consent was obtained from all patients participating in this study.

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Prof. Nestor Seredyuk, MD, head of the Department of Internal Medicine №2 and Nursery, Doctor of Medical Sciences, Professor, Vice-Rector for Research at Ivano-Frankivsk National Medical University (1987–2011 pp.), Head of the Department from 1987 to 2011 and from 2018 to present time, Honored Worker of Science and Technology of Ukraine, member of ASH (the American Society of Hypertension), member of ESC (the European Society of Cardiology).

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