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Figure 1.

Prikaz slučaja

Case report

Neobičan slučaj fistule koronarne arterije

An unusual case of coronary artery fistula

Alexander Kiško, Ján Kmec, Pavol Gazdič, Mária Vereb, Nelli Kishko

J. A. Reiman University Hospital, Prešov, Republika Slovačka

• J. A. Reiman University Hospital, Prešov, Slovak Republic

SAŽETAK: 32-godišnji pacijent za-primljen je radi obrade prve sinkope. Auskultatorno je prisutan kontinuirani šum gore lijevo parasternalno. 12-kanalni EKG je pokazao sinusni ritam 62/min s inkompletnim blokom desne grane i bez značajnih ST-T promjena. Transtorakalnom ehokardiografijom se, uz primjenu obojenog Dopplera, otkrio anomalni mlaz protoka iz lateralne stijenke u plućno arterijsko stablo (PAS). Selektivna koronarna angiografija i multiplanarna rekonstrukcija primjenom višeslojnog CT-a potvrdile su nalaz fistule sa sakularnom aneurizmom koja odgovara proksimalnom dijelu lijeve prednje silazne arterije i PAS. Cikloergometrija i Tc-99m Myoview perfuzijski SPECT nisu ukazali na znakove ishemije miokarda. Kod pacijenta se dijagnosticiralo postojanje fistule koronarne arterije i PAS, bez ishemije miokarda

ABSTRACT: A 32-year-old patient was referred for evaluation of syncope presented him for the first time. A continuous murmur could be heard mainly at the upper left sternum border. Conventional ECG showed a regular sinus rhythm of 62 bpm with incomplete right bundle branch block and no significant ST-T changes. A transthoracic echocardiography revealed an anomalous color flow jet from the lateral wall into the main pulmonary artery (PA). A selective coronary angiography and multi-detector CT scanner using multiplanar reconstruction confirmed fistula with saccular aneurysm corresponding the proximal portion of the left anterior descending artery and the main PA. Cycloergometry and stress/rest Tc-99m Myoview perfusion SPECT revealed no signs of myocardial ischemia. Patient was diagnosed



i bez dokaza o aritmogenoj etiologiji sinkope za koju se zaključilo da je bila vazovagalna. Preporučeno je konzervativno praćenje. Pacijent je ostao bez simptoma, nije više imao sinkope ili drugih komplikacija koje bi bile povezane s fistulom koronarne arterije. Predstavljamo postojeće tehnike za prikaz koronarnih fistula, zajedno s njihovom funkcionalnom ocjenom, završavajući sa sažetkom sadašnjih strategija za njihovo zbrinjavanje.

KLJUČNE RIJEČI: Koronarna fistula, sinkopa, Doppler, Tc-99m Myoview perfuzijski SPECT

Kongenitalne fistule koronarne arterije (CAF), koje je po prvi put opisao Krause godine 1865.¹, karakterizira normalan početak koronarnih arterija u aorti te fistulozna komunikacija s velikim krvnim žilama, ali i svim srčanim šupljinama. Oko 50% CAF-a nastaje iz desne koronarne arterije² i najčešće se dreniraju u desnu pretklijetku ili desnu klijetku, ali povremeno mogu završiti u plućnoj arteriji (PA). Kod ovih abnormalnosti koronarne arterije, krv se drenira u veliku krvnu žilu ili neku drugu strukturu, zaobilazeći kapilarnu mrežu miokarda, što može uzrokovati fenomen krađe. Bolesnici sa CAF su uglavnom bez simptoma, ali neki mogu imati umor, zaduho, anginozne smetnje, aritmije, znakove kongestivnog zatajivanja srca, plućne hipertenzije ili infekcijskog endokarditisa. Sinkopa je vrlo rijetko povezana sa CAF-om. Prikazujemo slučaj fi-

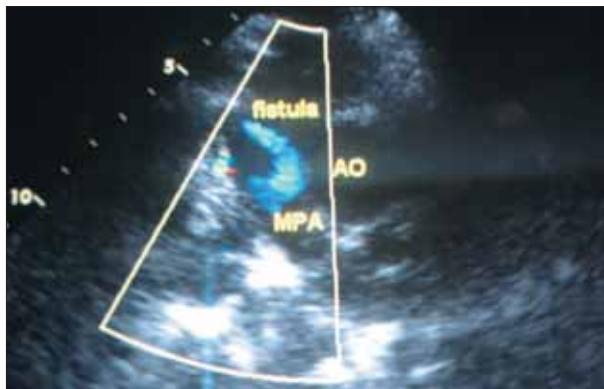


Figure 2. A transthoracic echocardiography parasternal short axis view showing an anomalous color flow jet at color Doppler analysis arising from the lateral wall away from the transducer into the main pulmonary artery.

as having a coronary artery to main PA fistula with no myocardial ischemia and no evidences of arrhythmogenic syncope that was concluded as vasovagal. Conservative follow-up was suggested. Patient remains asymptomatic with no syncope or complications related to coronary artery fistula. We discuss the current techniques available for imaging coronary fistulas, along with their functional assessment, concluding with a summary of current strategies for management.

KEYWORDS: Coronary fistula, syncope, Doppler, Tc-99m Myoview perfusion SPECT.

Congenital coronary artery fistulas (CAF), first described by Krause in 1865¹ are characterized by normal aortic origin of the coronary artery but with a fistulous communication with the great vessels as well as all cardiac chambers. About 50% of the CAF arise from the right coronary artery² and most commonly drain into the right atrium or right ventricle, but occasionally can drain into the pulmonary artery (PA). In these coronary artery abnormalities blood is shunted into a great vessel, or other structure, bypassing the myocardial capillary network and may produce steal phenomenon. The patients with CAF are mostly asymptomatic, but some may present with symptoms of fatigue, dyspnoe, angina, arrhythmias, signs of congestive heart failure, pulmonary hypertension or infective endocarditis. Syncope is very rarely associated with

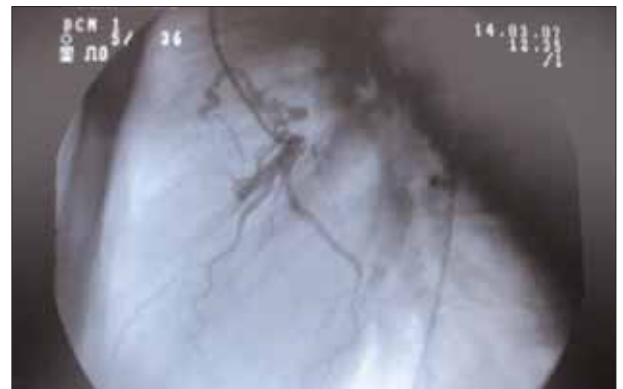


Figure 3. Selective coronary angiography showing a 'serpentine' fistula with aneurysm corresponding the proximal portion of the left anterior descending artery and the main pulmonary artery.

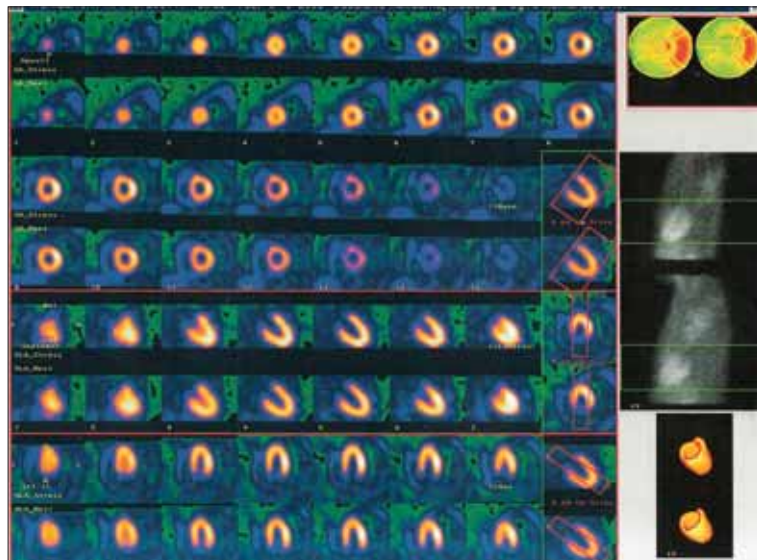


Figure 4. Stress/Rest Tc-99m Myoview perfusion SPECT using a bicycle ergometer shows no perfusion defects due to stress-induced myocardial ischemia.



stule koronarne arterije i plućnog arterijskog stabla (PAS) koja se klinički prikazala sinkopom.

Prikaz slučaja

32-godišnji svećenik upućen je na evaluaciju sinkope koja se pojavila tijekom služenja svete Mise. Pacijent se nije žalio na zaduhu ili bolove u prsnoj koži koji bi bili povezani s fizičkom aktivnošću, a nije imao niti palpitacije ili neke druge kliničke simptome aritmije. Bio je fizički aktivan, a u obiteljskoj anamnezi nije bilo koronarne bolesti srca. Vrijednosti lipida bile su uredne.

Prilikom fizikalnog pregleda pacijent je bio normosteničan, acijanotičan, normotenzivan, imao je normalan auskultatorni nalaz na plućima i pravilnu srčanu frekvenciju od 60/min. Auskultatorno na srcu registriran je kontinuirani šum, stupnja 2/6, koji se čuo najjače u razini drugog interkostalnog prostora lijevo parasternalno. Nije bilo znakova zatajivanja srca. Elektrokardiogram (EKG) je u mirovanju pokazao regularni sinusni ritam od 62/min uz inkompletni blok desne grane i bez značajnih ST-T promjena.

Transtorakalnom ehokardiografijom (TTE) u parasternalnom prikazu u kratkoj osi, primjenom obojenog Dopplera, otkriven je anomalni mlaz protoka koji nastaje iz lateralne stijenke u PAS (**slika 2**). Usprkos različitim projekcijama, točan anatomski tijek moguće fistule se nije mogao jasno prikazati transezofagijском ehokardiografijom (TEE). Selektivnom koronarnom angiografijom utvrđena je fistula u obliku serpentine s aneurizmom koja odgovara proksimalnom dijelu lijeve prednje silazne arterije i PAS, bez značajnih znakova koronarne ateroskleroze (**slika 3**). Složena anatomija fistule je detaljno prikazana višeslojnim CT-om koristeći multiplanarnu rekonstrukciju i tehniku 3D rekonstrukcije. Njime je pokazana sakularna dilatirana fistula koja nastaje iz proksimalne lijeve prednje silazne arterije i drenira se u PAS (**slika 1**, stranica 37).

Kako bi ocijenili hemodinamski značaj otkrivene koronarne fistule izvršena je cikloergometrija do 85% maksimalne teoretske frekvencije srca koja nije registrirala nikakve ST-T promjene (200W; 181/min; 11,0 MET-a) bez simptoma tijekom pregleda. Tc-99m Myoview perfuzijskim SPECT-om u mirovanju i opterećenju primjenjujući cikloergometar, nisu otkriveni perfuzijski poremećaji niti naporom uzrokovana ishemija miokarda (**slika 4**).

Radi ocjene mogućeg uzroka sinkope, pacijent je podvrgnut standardnom postupku ispitivanja. Dva puta je izvršeno 24-satno snimanje EKG-a (Holter) kojim se nije dokazala moguća aritmogena etiologija sinkope. Reakcija pacijenta na head-up tilt test i masažu karotidnog sinusa bila je fiziološka. Transezofagealna atrijska stimulacija nije dokazala abnormalnosti provođenja.

Na temelju učinjene obrade kod pacijenta se dijagnosticira postojanje fistule koronarne arterije i PAS, bez ishemije miokarda i bez dokaza o disritmičkoj etiologiji sinkope za koju je zaključeno da je vazovagalna. Preporučeno je konzervativno praćenje. Tijekom razdoblja praćenja od dvije godine pacijent je ostao bez simptoma, nije više imao sinkope ili drugih komplikacija povezanih s fistulom koronarne arterije. Kontrolna cikloergometrija i Tc-99m Myoview perfuzijski SPECT nije otkrio znakove ishemije miokarda.

CAF. We report the case of coronary to main PA fistula clinically first presented with syncope.

Case presentation

A 32-year-old priest was referred for evaluation of syncope first presented him during servicing the Mass. He reported no history of the episodes of shortness of breath or chest pain associated with physical activity as well as palpitations or other clinical symptoms of arrhythmias. He was physically active with no family history of coronary heart disease with absolutely normal lipid profile.

At physical examination patient was found to be normostenic, acyanotic, normotensive with clear lungs and a regular pulse of 60 bpm with normal dual heart sounds. A grade Levine 2/6 continuous murmur could be heard mainly at the level of the second intercostal space of the left parasternal area. There were no signs of heart failure. An electrocardiogram (ECG) at rest showed a regular sinus rhythm of 62 bpm with incomplete right bundle branch block and no significant ST-T changes.

A transthoracic echocardiography (TTE) in parasternal short-axis view revealed an anomalous color flow jet at color Doppler analysis arising from the lateral wall into the main PA (**Figure 2**). Despite various projections, the exact anatomic course of the suspected fistula could not be clearly shown by the transesophageal echocardiography (TEE). A selective coronary angiography was obtained that confirmed 'serpentine' fistula with aneurysm corresponding the proximal portion of the left anterior descending artery and the main PA with no significant signs of coronary atherosclerosis (**Figure 3**). The complex anatomy of the fistula was demonstrated in detail by a multi-detector CT scanner using multiplanar reconstruction and 3D reconstruction technique. It showed a sacculary dilated fistula that originates from the proximal left anterior descending artery and drainages the main PA (**Figure 1**, Page 37).

To evaluate the hemodynamic significance of detected coronary fistula the cycloergometry was performed up to 85% of the maximum theoretical heart rate which showed no ST-T changes (200W; 181 bpm; 11.0 MET's) with no symptoms during examination. Stress/Rest Tc-99m Myoview perfusion SPECT using a bicycle ergometer revealed no perfusion defects and stress-induced myocardial ischemia as well (**Figure 4**).

In purpose to evaluate a possible cause of syncope the patient underwent a standard exam protocol. Twice performed Holter ECG recording showed no evidences of the possible arrhythmogenic nature of syncopal episode. Patient's response to head-up tilt testing and to the carotid sinus massage was physiological. Transesophageal atrial stimulation revealed no evidence of conductivity abnormalities.

From these results, the patient was diagnosed as having a coronary artery to main PA fistula with no myocardial ischemia and no evidences of dysrhythmic syncope. Last one was concluded as vasovagal. Conservative follow-up with no intervention procedure was suggested. Patient remains asymptomatic with no syncope or other complications related to CAF during follow-up period of 2 years, control cycloergometry and stress/rest Tc-99m Myoview perfusion SPECT revealed no signs of myocardial ischemia.



Diskusija

CAF su rijetke, pretežito kongenitalne anomalije koronarnih arterija koje mogu nastati iz bilo koje tri glavne koronarne arterije i dreniraju se u velike krvne žile kao i sve srčane šupljine. U ranom fetalnom razvoju perzistiranje miokardijalne sinusoide i naknadna povezanost sa endotelijalnim pupoljcima koji su nastali iz arterijskog stabla predstavlja temelj za abnormalnu fistulu koronarne arterije³.

Pojava ove anomalije varira od 0,2% do 0,4% u odabranom nizu bolesnika koji boluju od kongenitalnih srčanih bolesti, a pojava kod cjelokupne populacije se procjenjuje na približno 0,002%⁴. Za vrijeme 18.272 dijagnostičkih srčanih kateterizacija, CAF su bile utvrđene slučajno kod 10 bolesnika (0,05%)⁵. Većina ovih fistula nastaju iz desne koronarne arterije. Fistule lijeve koronarne arterije su rjeđe i često dreniraju u desnu klijetku ili desnu pretklijetku⁶. Kombinacija opisana u sadašnjem slučaju je neobična obzirom da fistule nastaju iz lijeve srčane arterije kod približno 35% slučajeva, a drenaža u PA se dešava kod svega 17%⁷.

Većina bolesnika sa CAF su bez simptoma, ali njihov dugoročni ishod nije u potpunosti poznat. Klinički tijek CAF-a može varirati od spontanog zatvaranja do ozbiljnih komplikacija. Bolesnici s koronarnim fistulama mogu imati zaduhu, kongestivno zatajivanje srca, anginozne smetnje, endokarditis, aritmije ili infarkt miokarda. Sinkopa nije tipičan simptom povezan sa fistulama i vrlo je rijetka klinička manifestacija CAF-a. Može se povezati s nekim drugim kongenitalnim srčanim greškama^{8,9}. Ovdje prikazani pacijent ima izoliranu CAF i nema drugih srčanih abnormalnosti, pa stoga nije bilo potpuno razumljivo da li je epizoda sinkope bila izravno povezana s nalazom fistule.

TTE sa snimanjem protoka obojenim Dopplerom od strane iskusnog ehokardiografičara predstavlja idealan dijagnostički alat prve linije za ocjenu i dijagnozu CAF¹⁰. Dijagnoza koronarne fistule do PAS se obično može utvrditi obojenim Dopplerom kada se vide abnormalni signali protoka s mozaičkim izgledom u području PA. Dijagnostički je korisno vidjeti tijekom shunta sve do izlaza koronarne fistule. Ponekad je teško otkriti fistule koje se dreniraju u PA, a u današnje vrijeme tome može pomoći 3-D TTE¹¹.

TEE može precizno prikazati porijeklo, tijek i mjesto drenaže fistule¹², ali ponekad, kao u prikazanom slučaju, može zatajiti. Tada su za ocjenu konfiguracije fistule korisne selektivna koronarna angiografija i višeslojni CT s multiplanarskom rekonstrukcijom i 3D tehnikom rekonstrukcije. Dodatno ispitivanje može biti potrebno radi određivanja mogućih ishemijskih promjena u srcu. Cikloergometrija i Tc-99m Myoview SPECT u mirovanju i opterećenju mogu biti korisni u ocjenjivanju hemodinamskog značaja koronarnog protoka i ishemije miokarda uzrokovane fistulom.

Kod pacijenata s beznačajnim CAF-om preporuča se konzervativno praćenje, a intervencijski postupci mogu biti nepotrebni¹³. Elektivno zatvaranje fistula perkutanom transkateterskim tehnikama ili kirurški je općenito prihvaćeno kada postoje simptomi, a postoje kontroverze u pogledu zbrinjavanja asimptomatskih bolesnika¹⁴. Na temelju iskustva nema dokaza da su asimptomatske CAF, slučajno dijagnosticirane ehokardiografijom uz primjenu obojenog

Discussion

CAF are rare predominantly congenital coronary artery anomalies that can originate from any of the three major coronary arteries and drain in the great vessels as well as to all cardiac chambers. In early foetal development, persistence of myocardial sinusoids and the subsequent connection with the endothelial buds that originated from the base of truncus arteriosus forms the basis for abnormal coronary artery fistulae³.

The incidence of this anomaly ranges from 0.2% to 0.4% in selected series of congenital heart disease patients, and its incidence in the overall population is estimated to be about 0.002%⁴. During 18,272 diagnostic cardiac catheterizations, CAF were identified incidentally in 10 patients (0.05%)⁵. The majority of these fistulas arise from the right coronary artery. Left CAF are less common, but usually drain into the right ventricle or right atrium⁶. A combination like the one described in the present case is unusual since fistulas originate from the left coronary artery in about 35% of cases and drainage into the PA occurs in only 17%⁷.

The majority of the patients with CAF are clinically asymptomatic but the long-term outcome is not fully known. The clinical course of CAF may vary greatly from spontaneous closure to severe complications. The patients with coronary fistulas may present with dyspnea, congestive heart failure, angina, endocarditis, arrhythmias, or myocardial infarction. Syncope is not a typical symptom related to fistulas and is very rare first clinical manifestation of CAF. It may be associated with some underlying congenital heart defects^{8,9}. Our patient presented with an isolated CAF but no other coexisting heart abnormalities and it was not fully understood whether a syncopal episode, was directly related to the fistula.

TTE with color flow imaging by an experienced echocardiographer is an ideal first-line definitive diagnostic tool in the evaluation and diagnosis of CAF¹⁰. The diagnosis of coronary fistula to the main PA can usually be made by Doppler color flow imaging when abnormal flow signals with mosaic appearance in the pulmonary artery is visualize. It is diagnostically useful to visualize shunt flows originating from the exit of a CAF. However, sometimes fistulas to the PA may be difficult to detect. Nowadays, a live 3-D transthoracic echocardiography can be useful in demonstration of coronary artery to PA fistula¹¹.

TEE is capable of precisely demonstrating the origin, the course and the drainage site of the fistula¹², but sometimes like in presented case it may fail. Then a selective coronary angiography and MDCT scan using multiplanar reconstruction and 3D reconstruction technique are useful to assess the configuration of the fistula. An additional examination may be required to determine the possible ischemic changes in the heart. Cycloergometry and stress/rest Tc-99m Myoview SPECT may be useful for assessing the hemodynamic significance of the coronary flow and the fistula-related myocardial ischemia.

In patients with nonsignificant CAF, conservative follow-up is strongly suggested and intervention procedures may be unnecessary¹³. Elective closure of coronary artery fistulas by percutaneous transcatheter techniques or surgery is generally accepted in the presence of symptoms,



Dopplera, povezane s klinički neželjenim ishodom. Konzervativno zbrinjavanje uz nastavak praćenja ovih pacijenata čini se primjerenim¹⁵. Potencijalno ozbiljne i hemodinamski značajne koronarne fistule zahtijevaju točno prepoznavanje i pravovremenu perkutano i kiruršku korekciju¹⁶.

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E-mail: akisko@unipo.sk

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but controversies exist in the management of asymptomatic patients¹⁴. Based on the experience, there is no evidence that clinically silent CAF diagnosed incidentally by color Doppler echocardiography are associated with adverse clinical outcome. Conservative management with continued follow-up of these patients appears to be appropriate¹⁵. Potentially serious and hemodynamically significant CAF require accurate recognition, and at times, percutaneous or surgical correction¹⁶.