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О. КІШКО, Л. ДЕРНЯРОВА, Я. КМЕЦ, М. БАБЧАК, А. ГУДАКОВА
Пряшівський Університет, Факультет охорони здоровя, Клініка кардіології, Пряшів, Словаччина

КЛІНІЧНИЙ ВИПАДОК ФІСТУЛИ ВІНЦЕВОЇ АРТЕРІЇ У СПОРТСМЕНА З НЕТИПОВОЮ МАНІФЕСТАЦІЄЮ У ВИГЛЯДІ СИНКОПЕ – ДІАГНОСТИЧНИЙ ТА ЛІКУВАЛЬНИЙ АЛГОРИТМ

Представлено унікальний клінічний випадок у 32-річного активного спортсмена-велосипедиста, якого обстежеували у звязку з синкопе підчас передсезонної підготовки. Обєктивним обстеженням виявилено нормостенічну тілобудову, з показниками кровяного тиску в межах норми, ясним легеневим звуком та везикулярним диханням, діяльність серця була ритмічною з ЧСС 60/хв, вислуховувались два серцеві тони та континуальний діастолічний шум 2/6 за Левіним в другому міжребровому просторі, зліва від краю грудинни. Сімейний анамнез не виявив випадків передчасної раптової смерті, показники ліпідів крові були в нормі. На ЕКГ спокою реєструвався регулярний синусовий ритм частотою 62 удари за хвилину, ознаки неповною блокади правої ніжки пучка Гіса, без яких-небудь патологічних ST-T змін. Підчас трансторакальної ЕХОКГ в парастернальній короткої осі спостерігався аномальний діастолчний потік в легеневій артерії від зовнішної її стінки вглиб основного стовбуру, що викликало підозру на наявність коронарної нориці. Однак, ця діагностична підозра не знайшла однозначного підтвердження в процесі черезстравохідної ЕХОКГ. Коронарна ангіографія не виявила стенотичних змін на епікардіальних вінцевих судинах, однак підтвердила можливість існування аномального серпантиноподібного дренажу від лівої передньої низхідної артерії до легеневої артерії. Детальну анатомію вінцевої фістули з мішкоподібними аневризмами, яка бере свій початок від проксимальної частини лівої передньої низхідної вінцевої артерії та дренує основний стовбур легеневої артерії було достеменно підтверджено методом 64-зрізової МДКТ. Перфузійна сцинтиграфія міокарда з Tc-99m не виявила дефектів перфузії в процесі максимальнго фізичного навантаження, що свідчило про відсутність феномену обкрадання і було причиної відмови від інтервенції на фістулі з метою її закриття. Результати довготривалого моніторування ЕКГ, тесту на похилій площині, масажу каротидного синуса і програмованої стимуляція передсердь свідчили про вазовагальний тип синкопе. Протягом трьох років спостереження пацієнт залишається безсимптомним і продовжує виступи в змаганнях, а щойно реалізована контрольна перфузійна сцинтиграфія міокарда не виявила жодних проявів, повязаних з можливою коронарною ішемією. На закінчення, в окремих випадках діагностика коронарної фістули є складною і вимагає використання ряду сучасних методів візуалізації а подальша тактика залежить від наявності чи відсутності ознак міокардіальної ішемії внаслідок феномену обкрадання. У випадку їх відсутності можливою є консервативна тактика з динамічним спостереженням за особою.

Ключові слова: коронарна фістула, синкопе, кольорове допплерівське зображень, перфузійна сцинтиграфія міокарду з Tc-99m

A. KISKO, L. DERNAROVA, J. KMEC, M. BABCAK, A. HUDAKOVA
Presov University in Presov, Faculty of Health Care, Cardiology Clinic, Presov, Slovakia

CLINICAL CASE OF CORONARY ARTERY FISTULA IN ATHLETE ATYPICALLY PRESENTED WITH SYNCOPE – DIAGNOSTIC AND MANAGEMENT ALGORYTHM

Introduction. Congenital coronary artery fistulas (CAF), first described by Krause in 1865 [6] 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 artery [3] and most commonly drain into the right atrium or right ventricle, but occasionally can drain into the pulmonary artery. 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 CAF. We report the case of coronary to main pulmonary artery fistula clinically first presented with syncope.

Case presentation. A 32-year-old active cyclist was referred for the evaluation because of syncope he had performed in preseason. He reported no history of the episodes of shortness of breath or chest pain associated with extreme physical exertion as well as palpitations or other clinical symptoms of cardiac arrhythmias. He was physically absolutely normal with no familial predisposition to sudden cardiac death or ischemic heart disease and normal lipids.

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 diastolic 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 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 in parasternal short-axis view revealed an anomalous colour flow jet in diastola arising from the lateral wall into the main pulmonary artery, identifying the fistula drainage site (fig.1). In contrast, despite various projections, the exact anatomic course of the suspected fistula could not be clearly shown by the transoesophageal echocardiography. A selective coronary angiography confirmed ‘serpentine’ fistula with aneurysms corresponding the proximal portion of the left anterior de-

scending artery and the main pulmonary artery with no significant signs of coronary atherosclerosis (fig.2).

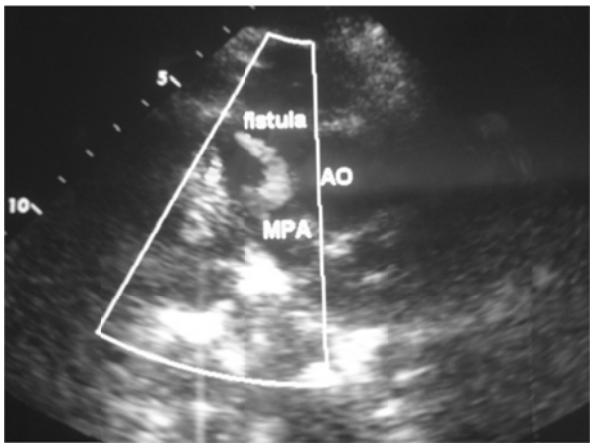

Figure 1. A transthoracic echocardiography in parasternal short-axis view reveals an anomalous colour flow jet in diastola at colour Doppler analysis arising from the lateral wall into the main pulmonary artery.

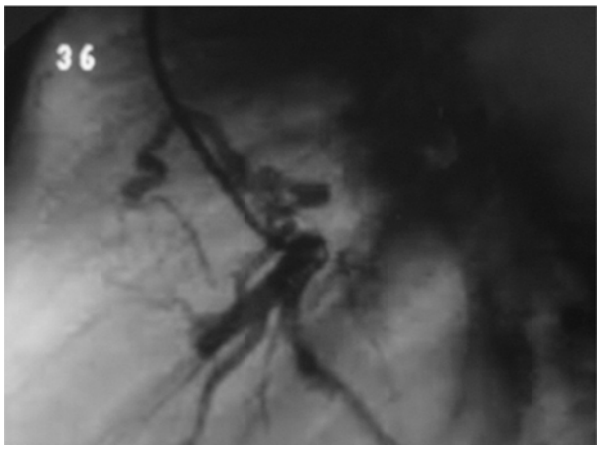

Figure 2. Selective coronary angiography showing a 'serpentine' fistula with aneurysms corresponding the proximal portion of the left anterior descending artery and the main pulmonary artery.

The complex anatomy of the fistula was demonstrated in detail by 64 Slice MDCT scanning using multiplanar and 3D reconstruction techniques. It showed a sacculary dilated fistula that originates from the proximal left anterior descending artery and drainages the main pulmonary artery (fig.3).

Figure 3. MDCT scanning using multiplanar and 3D reconstruction technique shows a sacculary dilated fistula that originates from the proximal left anterior descending artery and drainages the main pulmonary artery.

To evaluate the hemodynamic significance of detected coronary fistula the stress testing with the bicycle ergometer was performed in 3-minute steps up to the maximum theoretical heart rate which showed no ST-T change at the heart rate of 181 beats/min (200W, 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 (fig.4).

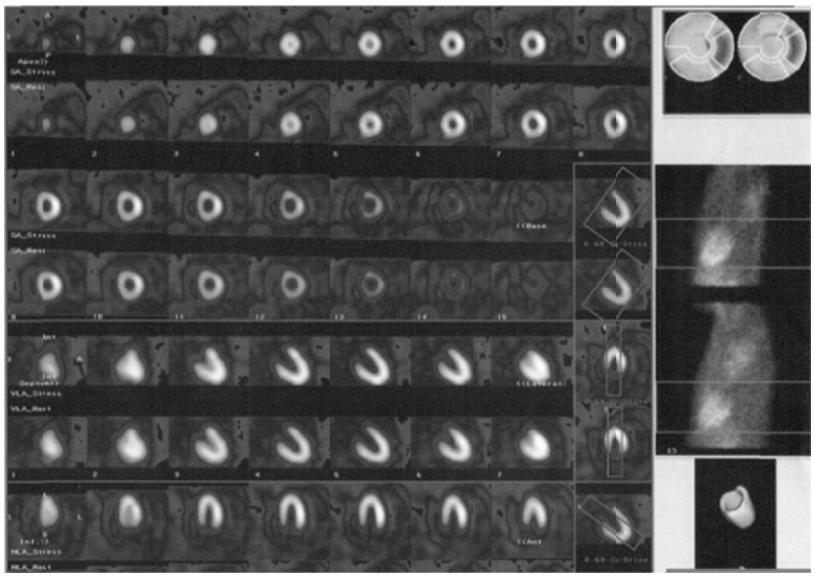

Figure 4. Stress/Rest Tc-99m Myoview perfusion SPECT shows no stress-related perfusion defects.

In purpose to evaluate a possible cause of syncope the patient underwent a standard evaluation protocol. Twice performed holter ECG monitoring showed no evidences of the possible arrhythmogenic nature of syncope episode. Patient's response to head-up tilt testing and to the carotid sinus massage was physiological. Programmed atrial stimulation revealed no evidence of conductivity abnormalities. From these results, the patient was diagnosed as having a coronary artery to pulmonary artery 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. After a three year follow-up patient remains asymptomatic still participating in cycling tours and other sports events. Recently performed control myocardial Tc-99m Myoview perfusion SPECT showed no evidence of stress related myocardial ischemia.

Discussion. CAF are rare predominantly congenital or occasionally acquired (such as stab or projectile injuries, post-coronary angioplasty) 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. The evolution of the coronary artery network involves complex embryology. 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 fistulae [3].

The incidence of this anomaly ranges from 0.2-0.4% in selected series of congenital heart disease patients, and its incidence in the overall population is estimated to be about 0.002%. During 18,272 diagnostic cardiac catheterizations, CAF were identified incidentally in 10 patients (0.05%) [2]. The majority of these fistulas arise from the right coronary artery. Left CAF is less common, but usually drains into the right ventricle or right atrium [4]. 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 pulmonary artery occurs in only 17% [13].

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 dyspnoea, 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 [5, 8]. Our patient presented with an isolated CAF but no other coexisting heart abnormalities and it was not fully understood whether a syncope episode was directly related to the fistula. Clinically manifested myocardial ischemia is resulting from fistula steal phenomenon. Coexisting congenital or acquired heart disease is found in 40% of patients; however, no associated lesion was found to have a specific relationship with CAF.

Transthoracic echocardiography with colour flow imaging by an experienced echocardiographer is an ideal first-line definitive diagnostic tool in the evaluation and diagnosis of CAF [1]. The diagnosis of coronary fistula to the main pulmonary artery can usually be made by Doppler colour 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 pulmonary artery may be difficult to detect [9]. Nowadays, a live three-dimensional transthoracic echocardiography can be useful in demonstration of coronary artery to pulmonary artery fistula [10].

Transoesophageal echocardiography is capable of precisely demonstrating the origin, the course and the drainage site of the fistula [12], 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. Ergometry or stress/rest Tc-99m Myoview SPECT may be useful for assessing the hemodynamic significance of the coronary flow and the fistula-related myocardial ischemia. No evidence of the stress induced myocardial ischemia was found in presented case.

In patients with non-significant CAF, conservative follow-up is strongly suggested and intervention procedures may be unnecessary [7]. Elective closure of coronary artery fistulas by percutaneous transcatheter techniques or surgery is generally accepted in the presence of symptoms, but controversies exist in the management of asymptomatic patients [11]. Based on the experience, there is no evidence that clinically silent CAF diagnosed incidentally by colour Doppler echocardiography are associated with adverse clinical outcome. Conservative management with continued follow-up of these patients appears to be appropriate [14]. Potentially serious and hemodynamically significant CAF require accurate recognition, and at times, percutaneous or surgical correction [15].

Conclusion. In conclusion, several imaging techniques are needed for an accurate diagnosis of coronary fistula and for the suggestion of proper further management. The patient presented in this case was asymptomatic at the time of diagnosis and consequently no intervention was recommended. The patient is seen in follow-up and is doing fine, still participating in sports reporting no complaints, but he might require transcatheter closure or surgical treatment when he becomes symptomatic.

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A. KISKO, L. DERNAROVA, J. KMEC, M. BABCAK, A. HUDAKOVA

Presov University in Presov, Faculty of Health Care, Cardiology Clinic, Presov, Slovakia

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A 32-year-old active cyclist was referred for the evaluation because of syncope he had performed in preseason. He was found to be normostenic, acyanotic, normotensive with clear lungs and a regular pulse of 60 bpm, with normal dual heart sounds and a grade Levine 2/6 continuous diastolic murmur in the second intercostal space of the left parasternal area. He had no family history of premature cardiac death and his lipids were normal. Rest ECG showed a regular sinus rhythm of 62 bpm with incomplete right bundle branch block and no significant ST-T changes.

A transthoracic ECHO in parasternal short-axis view revealed an anomalous colour flow jet in diastole arising from the lateral wall into the main pulmonary artery and coronary artery fistula came under suspicion. In contrast, it has not been confirmed clearly by the transoesophageal ECHO. Coronary angiography was without coronary stenosis and confirmed a ‘serpentine’ anomalous drainage supposedly from left anterior descending artery to the main pulmonary artery. A complex anatomy of sacculary dilated fistula that originates from the proximal left anterior descending artery and drainages the main pulmonary artery was showed in detail by a MDCT scanning. Myocardial Tc-99m Myoview perfusion SPECT imaging showed no perfusion defects in maximal physical stress and follow-up without intervention was suggested. In addition, due to the results of holter ECG monitoring, head-up tilt testing, carotid sinus massage and programmed atrial stimulation syncope was concluded as vasovagal. After a three year follow-up patient remains asymptomatic and recently performed control myocardial perfusion SPECT showed no signs of stress related myocardial ischemia.

In conclusion, several imaging techniques are needed for an accurate diagnosis of coronary fistula and for the suggestion of proper further management.

Key words: coronary artery fistula, syncope, Doppler colour flow imaging, Tc-99m Myoview perfusion SPECT

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