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COR TRIATRIATUM SINISTER AS AN INCIDENTAL FINDING IN ELDERLY WOMAN

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SUMMARY

Cor triatriatum is a rare condition. It comprises around 0.1% to 0.4% of all congenital heart malformations. There are two types of this anomaly: cor triatriatum sinister (CTS), which is more common and accounts for 92% of all cases, and cor triatriatum dexter (CTD). This condition usually presents at an early age and is diagnosed mainly during early childhood. In some patients, who have less severe cases of CTS, diagnosis could be made in adulthood. We report an unusual case of a 78-year-old woman who was diagnosed with CTS for the first time. The patient was admitted to the cardiology department with symptoms of chest pain and dyspnoea. Physical examination revealed diffusely diminished breath sounds, with focal wheezing, and irregular heart rate, with no murmurs, while blood pressure was 140/90 mmHg. A transthoracic echocardiogram revealed an accessory membrane in the left atria suggestive of CTS.

Keywords: cor triatriatum sinister, hypertension, atrial fibrillation, vaginal hemorrhagia, anticoagulant therapy

SRPSKI

SAŽETAK

Srce sa tri pretkomore je retko stanje. Čini oko 0,1% do 0,4% svih urođenih srčanih malformacija. Postoje dva tipa ove anomalije: levo srce sa tri pretkomore (lat. cor triatriatum sinister, CTS) koje je češće i javlja se u 92% svih slučajeva i desno srce sa tri pretkomore (lat. cor triatriatum dexter, CTD). Ovo stanje se obično prezentuje u ranom dobu i dijagnostikuje se uglavnom u ranom detinjstvu. Kod nekih pacijenata, koji imaju manje teške slučajeve CTS, dijagnoza se može postaviti u odraslom dobu. Prikazujemo neobičan slučaj 78-godišnje žene kojoj je prvi put dijagnostikovan CTS. Pacijentkinja je primljena na odeljenje kardiologije sa simptomima dispneje i bola u grudima. Fizikalnim pregledom nađen difuzno oslabljen disajni šum sa vizingom, nepravilan srčani rad, bez šumova, dok je krvni pritisak bio 140/90 mmHg. Transtorakalni ehokardiogram otkrio je dodatnu membranu u levoj pretkomori koja ukazuje na CTS.

Ključne reči: levo srce sa tri pretkomore, hipertenzija, atrijalna fibrilacija, vaginalno krvarenje, antikoagulantna terapija

CASE REPORT

A 78-year-old woman was admitted to the cardiology department of the general hospital in Novi Pazar after couple of days of chest pain and dyspnoea. Patient history revealed hypertension and chronic obstructive pulmonary disease. A week prior to admission she was treated for persisting vaginal hemorrhage. After thorough diagnostics, and consultation with radiologists, urologist, endocrinologist, pulmonologist, surgeon and dermatologist, the gynecologist made a diagnosis of vaginal varicose veins. The patient was treated for thrombophlebitis and acroangiodermatitis at the time as well. After recuperation of all symptoms patient was discharged, but seeing vascular surgeon was strongly advised. Five days later, symptoms of chest pain and dyspnoea brought her back to the hospital.

Physical examination revealed diffusely diminished breath sounds, with focal wheezing, irregular heart rate, with no murmurs, while blood pressure was 140/90 mmHg. Varicose veins were present on lower extremities as well.

Electrocardiography showed atrial fibrillation, ventricular rate of 65 bpm, ST depression in inferior leads, as well as, precordial V4-V6 leads. T-wave was negative in V2 and V3 leads (Figure 1).

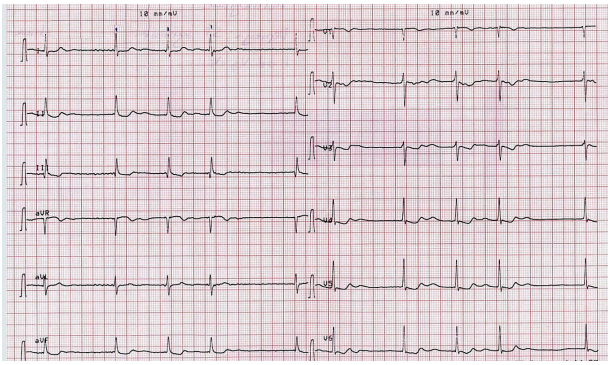

Figure 1 *- ECG*

Chest radiography showed enlargement of cardiac silhouette, mainly across right cardiac border, increase in width of pulmonary vascular pedicle, and redistribution of blood flow into upper parts of lung.

Transthoracic echocardiogram (TTE) showed severely abnormal left atrial diameter of 50 mm. In the left atria accessory membrane with small fenestration, suggestive of cor triatriatum sinister (CTS) was observed. Small fenestration present on intra-atrial septum indicated that this case of CTS was Loeffler class II. Thickening of anterior mitral cusp was also present, as were moderate mitral and tricuspid regurgitation (MR 1-2 +; TR 2+), and moderate pulmonary hypertension (right ventricle systolic pressure 48mmHg, mean trans-membrane pressure gradient was 38mmHg). Interventircular septum was 10mm thick, left ventricle posterior wall was 9mm, enddiastolic diameter of left ventricle was 50 mm, end-systolic diameter of left ventricle was 32 mm, ejection fraction was 55%, interatrial septum was intact and there was no free fluid in the pericardium (Figure 2 and Figure 3).

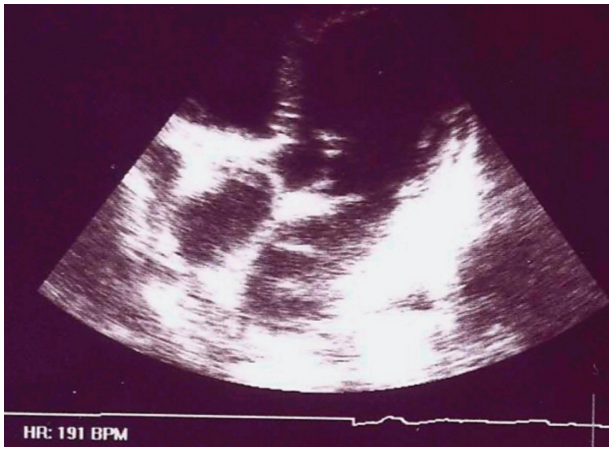

Figure 2 *- Transthoracic echocardiogram*

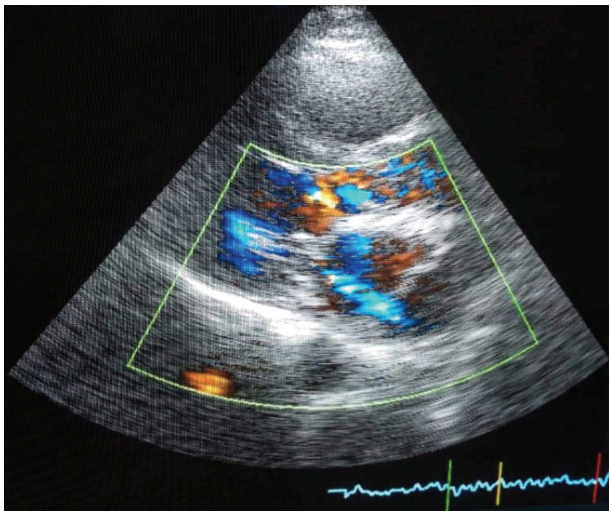

Figure 3 *- Transthoracic echocardiogram with Doppler*

Transtesophagaeal echocardiography (TEE) was indicated to confirm the diagnosis, as well as coronarography for thorough examination of the cause for ECG changes. Patient refused these procedures and consequently no TEE no coronarography were done.

CHA2DS2-VASc score was 5. Due to previous history of vaginal haemorrhagia, prescription of anticoagulant therapy was thought through. Decision to prescribe vitamin K antagonist (warfarin) was made after consultation, since there was a risk for cerebrovascular insult and benefits outweight possible risks. Patient was treated conservatively in the hospital. Diuretic, aldosterone antagonist, bronchodilator therapy, angiotensin II receptors antagonist and non-dihydropyridine calcium antagonist (verapamil) were prescribed for the symptoms as well as warfarin. After seven days in hospital patient recuperated and was discharged without symptoms of cardiac distress.

DISCUSSION

Cor triatriatum accounts for around 0.1% up to 0.4% of all congenital heart malformations [1]. Cor triatriatum sinister (CTS) is more frequent than cor triatriatum dexter (CTD), and accounts for 92% of all cases of this condition reported

so far [2]. This represents rare cardiac malformation and its embryological background is not well described, with numerous theories reported [3]. Its prevalence is almost equal in both sexes (with slightly higher prevalence among males 1.5:1) and there is no known genetic predisposition [3].

The simplest classification of CTS is based on size and number of fenestrations in fibro-muscular membrane and is more well-known as Loeffler's classification [2]. In the Loeffler class one are cases without any communication between the accessory chamber and normal left atrium, although accessory chamber might be connected to right atrium or there might be pulmonary veins draining the blood in anomalous fashion. Loeffler class two includes cases with one or more small fenestration on accessory membrane, which allows some communication between two chambers. Loefller class three includes cases with large single opening which allows wide communication between normal left atrium and accessory chamber.

Cor triatriatum is usually diagnosed in children, since the symptoms tend to present at an early age [3]. Symptoms are most often a consequence of obstruction of pulmonary venous flow or concomitant abnormalities. Usual concomitant heart malformations are ostium secundum atrial septal defect or patent foramen ovale followed by left superior vena cava, anomalous pulmonary venous return, mitral valve regurgitation, isolated pulmonary artery stenosis, tetralogy of fallot, and double outlet right ventricle [4-7].

Less severe cases could be diagnosed in adulthood, and there is an increasing trend in diagnosis of these cases in previous years due to improvements of diagnostic procedures [8]. Diagnosis in adult patient in usually made incidentally or patient was examined by cardiologist due to symptoms of mitral stenosis or any of the following: dyspneoa, orthopnoea, hemoptysis, palpitations, transient ischemic attack, syncope, right heart failure or hypoxemia [9, 10, 11]. Generally, clinical presentation of CTS may vary. It could present as chronic obstructive pulmonary disease, atrial fibrillation, cardio-embolic stroke, pulmonary artery thrombosis or esophageal varices [2]. Occurrence of symptoms in adults could be led by development of valvular insufficiency, atrial arrhythmias or fibrosis of fenestrations of fibro-muscular septum, which mimics mitral stenosis. Mitral regurgitation could also develop, which is thought to be a consequence of myxomatous valve degeneration. Adults could develop symptoms after excessive volume administration, during pregnancy or postpartum period [3].

Diagnosis of cor triatriatum, today, is made with different diagnostic imaging procedures: transthoracic echocardiography, transesophageal echocardiography, computed tomography, catheter angiography, and magnetic resonance imaging [3]. The most commonly used is transthoracic echocardiography, which could be enough for the most cases of CTS, but transesophageal echocardiography is used for better visualization and better differentiation from other structures [2].

ECG in cases of CTS is usually normal. If pulmonary artery hypertension develops, right atrial hypertrophy, right axis deviation and S1Q3 pattern could be found [12]. There might also be atrial tachycardia and atrial fibrillation.

In the previous decades, cardiac angiography was also used, since it could show differential filling of the two distinct atrial chambers [12].

Management of an early diagnosed cor triatriatum sinister without other cardiac abnormalities is surgical [3]. In the centers where there are surgeons familiar with this procedure, surgical outcome is generally favorable [12]. The reported cases so far indicate all patients to be asymptomatic at follow up and overall five-year survival rate to be more than 90% [4, 13]. In older adults, like in the case of our patient, due elevated risk for adverse surgical outcomes and usually less serious obstruction due to CTS, follow up and symptomatic therapy are sufficient [2]. Our patient had CHA2DS2-VASc score of 5, which is considered very high and signals high risk for thromboembolism [14].

This is an extremely unusual case of incidental diagnosis of cor triatriatum sinister in an 78-year old patient without previously known heart malformations. Patient was admitted with unspecific cardiac symptoms of chest pain and dyspnoea. She was treated for hypertension for 16 years and for chronic pulmonary disease for 10 years. There was nothing that could indicate existence of cor triatriatum sinister in patient history. By the time she came into cardiologist office she already developed symptoms of heart failure, ischemia and chronic pulmonary disease.

CONCLUSION

Cor triatriatum sinister diagnosed in an elderly patient, which was the case here, where the patient was 78-year-old with no previous history of heart disease, is a rare finding and could be presented with wide range of symptoms of different cardiac or pulmonary diseases. First line for diagnosis is TTE, followed by TEE and treatment should be conservative in elderly patients with strict follow ups by attending physician.

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