

Thrombosis within Glenn anastomosis (RCD code: IV-1D.1b)

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Abstract

Thrombosis secondary to Glenn anastomosis in Ebstein's anomaly has been reported very rarely, treatment depends on presenting symptoms and risk factors. In this article we presented the case of 63-year-old woman with Ebstein's anomaly (implantation of biological tricuspid valve, Glenn procedure, closure of patent foramen ovale and implantation of DDD type pacemaker with epicardial electrodes) who complained on neck discomfort while tilting head forward. Eventually in whom thrombosis within Glenn anastomosis was diagnosed by angio-CT scan. JRC D 2015; 2 (3): 89–91

Key words: Ebstein's anomaly, Glenn procedure, thrombosis

Case presentation

We present case of a 63-year-old female patient with Ebstein's anomaly [1–4], treated with implantation of biological valve Mosaic 33 mm [5,6] Glenn procedure [7–10], closure of patent foramen ovale [11,12] and implantation of DDD type pacemaker (Biotronik) with epicardial electrodes in 2011. Prior to the procedure she suffered from paroxysmal atrial flutter/fibrillation, diabetes mellitus type 2 on oral treatment. She underwent radiofrequency catheter ablation for atrial fibrillation performed in 2009. On routine post operative check up one year later patient complained on neck discomfort while tilting head forward. There were also transient facial oedema and attacks of irregular palpitations for approximately 3 months. Anyway, she had better exercise tolerance and less dyspnea than before surgery.

On physical examination she had regular heart rate 70 beats per minute, normal lungs on auscultation, soft abdomen, no evidence of liver or spleen enlargement. Distended jugular veins, no peripheral oedema.

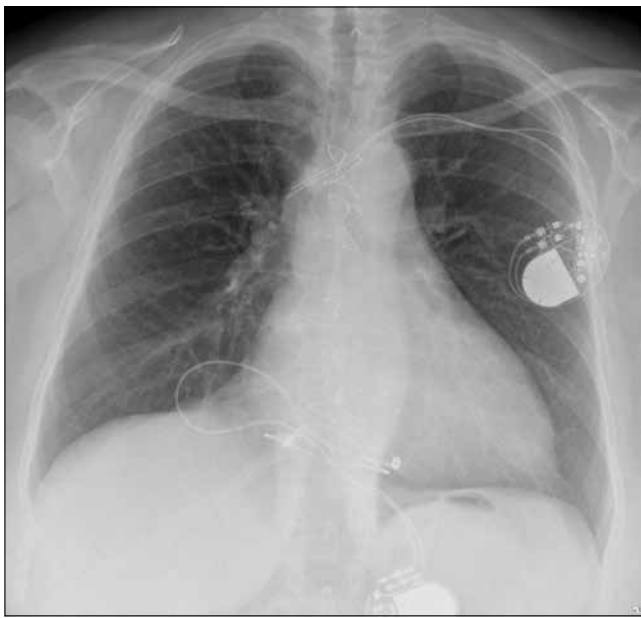
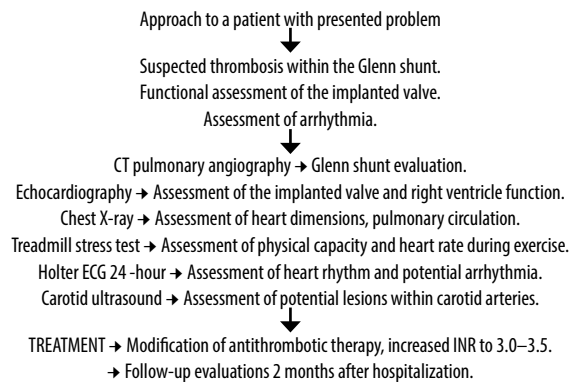
Resting electrocardiogram (ECG) showed VVI stimulation with pace of 69 bpm. Results of blood tests were: red blood cell (RBC) 4,57 million cells/mcL; hemoglobin (Hgb) 13,2 g/dL; hematocrit (HCT) 40,1 percent; platelet count 161 000/mcL; activated partial thromboplastin time (APTT) 44,79%, international normalized ratio (INR) 3,06. Chest X-ray revealed two-electrode

pacemaker implanted into abdominal wall tissues with epicardial electrodes, as well as bio-prosthesis in the tricuspid valve orifice. The heart silhouette was enlarged with normal aorta and pulmonary vascular markings (Figure 1). Transthoracic two-dimensional echocardiography showed normal morphology and function of the implanted valve – maximal tricuspid gradient 6.9 mm Hg and mean tricuspid gradient 3.5 mmHg; pulsatile blood flow within the Glenn shunt was demonstrated [13], with a backward systolic flow of approximately 1.3 m/s, which was respiratory related and reduced during apnoea. Superior vena cava was not dilated. Right ventricle (RV) was moderately enlarged (RV outflow track 38 mm with RV inflow track 43 mm), Tricuspid Annular Plane Systolic Excursion (TAPSE) 8 mm corresponding with diminished contraction of the RV. Left ventricular dimensions were within normal limits – (diastole 50 mm, systole 30 mm) with normal position of interventricular septum. Mild mitral regurgitation was noticed with dilated left atrium – atrial area 25 cm². Inferior vena cava was of normal size with regular respiratory variation (Figure 2). Angio computed tomography (angio-CT) of the pulmonary artery demonstrated anastomosis of the superior vena cava with right pulmonary artery. In addition, there was a clot (11 mm in diameter) attached to the posterior wall of the right pulmonary artery in front of vena cava superior. Prosthesis of the tricuspid valve and pacemaker with its epicardial electrode inserted into RV apex were demonstrated. There were also two former electrodes within the superior vena cava which were the remnants of the former pac-

Conflict of interest: none declared.

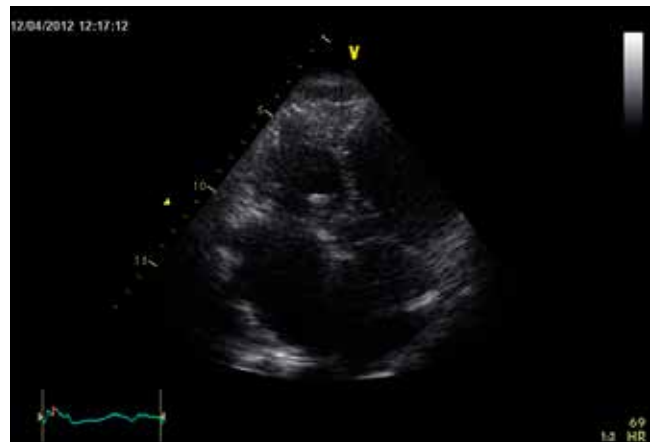
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Table 1. Patient management and follow-up**Figure 1.** Chest X-ray. Substantially increased diameters of the heart

ing system (Figure 3). On 24-hour Holter ECG, a sinus rhythm with episodes of paroxysmal /atrial fibrillation was found, as well as normal stimulation with an average ventricular rhythm of 66 bpm, minimum 59 bpm and maximum 87 bpm, 4 premature ventricular beats a day, and atrial fibrillation recorded in periods of reported palpitations. A treadmill stress test was performed on a bicycle ergometer: exercise was discontinued after 3 minutes and 19 seconds (9/10 Borg scale degrees), with maximum load of 100 W = approx. 5.4 METS, corresponding to 90% of normal limit for the age and sex, resting heart rate (HR) 69 bpm, maximum 70 bpm, corresponding to 44% of the limit. There was no blood pressure increase during exercise and no evidence of arrhythmia. Colour 2D Doppler imaging of peripheral vessels with spectral analysis of the carotid arteries showed insignificant atherosclerotic lesions within the both carotid arteries. Similarly, vertebral arteries showed normal flow.

On the ground of angio-CT thrombus within the right pulmonary artery was diagnosed year after Glenn procedure in patient

**Figure 2.** Transthoracic echocardiography. Apical 4-chamber view. Enlarged right heart chambers

with Ebstein's anomaly. Anti-thrombotic treatment was modified [14]. Laboratory tests did not correspond with pulmonary embolism [15]. The patient was discussed with cardiac surgeon who advised continuation of medical treatment. Other tests did not reveal any abnormalities. Patient was scheduled for follow-up [16] at the out-patient clinic of our department.

At discharge she was on: acenocumarol controlled by INR with target value of 3.0–3.5, extended release metoprolol succinate 95 mg, furosemide 40 mg 2 tablets twice daily (without this dose of furosemide patient has oedema), spironolactone 25 mg twice a day, metformin 500 mg a day, potassium supplementation.

Thrombosis secondary to Glenn anastomosis has been reported very rarely. Most of the cases have been found in the anastomosis region [17–20]. Main risk factors of thrombosis are female sex, increasing age, elevated right atrial and ventricular end-diastolic pressure (before surgery), elevated superior vena cava pressure and poor ventricular function (after surgery) [21–23]. Treatment depends on presenting symptoms and risk factors. It is recommended that anti-thrombotic therapy should be considered in patients who are suspected of having thrombosis within the Glenn anastomosis. Patient described had already been on anti-thrombotic treatment



Figure 3. Computed tomography. A blood clot or post-operative lesion in right pulmonary artery

so we decided to intensify it with higher dosage of acenocumarol and INR values between 3.0–3.5. The potential effect of this strategy will be evaluated by next angio-CT scan.

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