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A Unique Case of Embolic Myocardial Infarction in a 22 Years Old Womanwith Fontan Circulation

Case Report

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Abstract

Coronary artery embolism is an uncommon cause of acute myocardial infarction. We present a case of 22 years old patient with double inlet left ventricle (DILV), ventricular septal defect and pulmonary stenosis after Fontan repair with an acute embolic inferior ST- segment elevation myocardial infarction (STEMI). In addition to the known risks of thrombotic complications associated with Fontan circulation, potential predisposing factors to thromboembolism in this patient included thrombus formation located inthe inferior vena cava and the lack ofanticoagulation therapy. This is one of the first reported embolic causes of STEMI in a patient with a Fontan circulation. Up to date, there is no consensus that anticoagulation therapy with warfarin is superior to aspirin in primary prevention of thromboembolismin patients with Fontan circulation.

Introduction

Fontan circulation is associated with higher risk of thromboembolic events. However, the incidence of embolic myocardial infarction (MI) is very rare. We describe the unique case of young patient after fenestrated Fontan repair, presenting with embolic inferior STEMI. The antitrombotic treatment strategy plays pivotal role in primary and secondary prevention of tromboembolism.

Case report

We present a 22 years old woman with double inlet left ventricle, pulmonary stenosis, and ventricular septal defect, who underwent superior cavo-pulmonary shunt (Glenn) at the age of one yearand atrio-pulmonary (Fontan) connection at the age of 4 years recurrent supraventricular tachycardia necessitated multiple electro physiological studies, with ablation of posteroseptal accessory pathway at the age of 11.

In 2017, she was seenatthe emergency department for acute chest pain radiating to the neck. Examination showedno signs of heart failure, blood pressure was 100/60 mmHg. On ECG significant ST segment elevations

in limb leads II, III and aVFwere seen (Figure 1). A transthoracic echocardiogram (TTE) showed preserved left ventricularejection fraction(EF LV50%), hypokinesis of inferior wall, trivial mitral and tricuspidal regurgitation, suspection of trombus formation of width 5mm and length 38mm in inferior vena cava (IVC) (Figure 2 &3). Her chronic medical therapy included sotalol (2x80mg) and acetylsalicylic acid (1x100mg). She was transported to the East Slovak Institute of Cardiovascular Diseases and was medicated according to the STEMI guidelines with heparin and dual antiplatelet therapy. Her creatine kinase peaked to 5,72µkat/l, MB fraction of creatine kinase peaked to 0,46, high sensitive toponin T (hsTn-T)peaked to

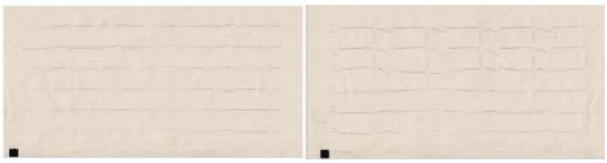

Figure 1: ECG showed signs of inferior STEMI

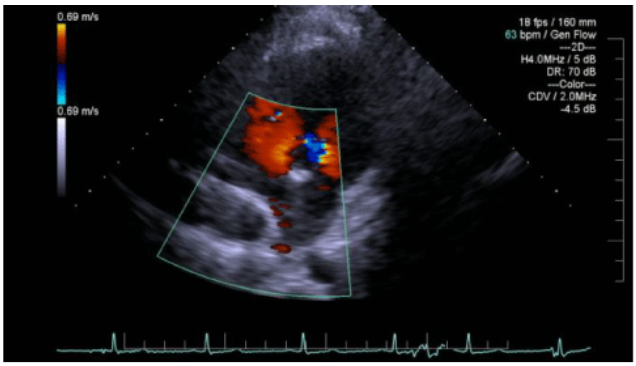

Figure 2: Echocardiography - four-chamber view

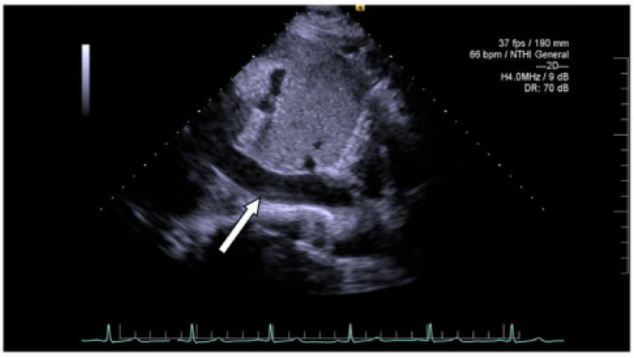

Figure 3: Echocardiography - trombus formation located in inferior vena cava

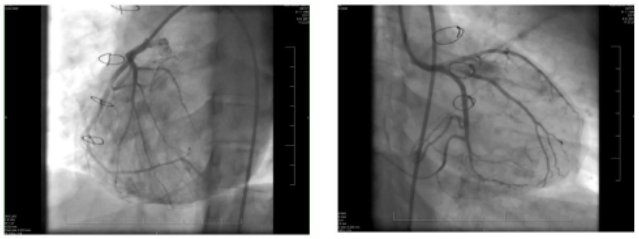

Figure 4: Selective coronary angiography – normal finding

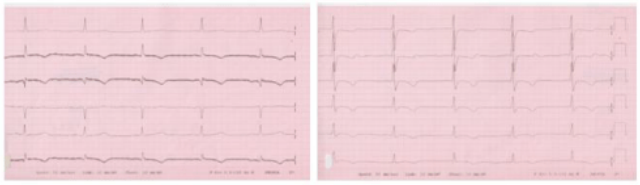

Figure 5: ECG after selective coronary angiography

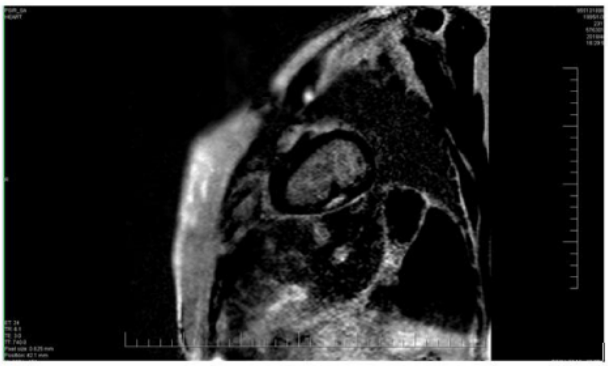

Figure 6: Cardiac magnetic resonance imaging (MRI), the late gadolinium enhancement showed the signs of past inferior transmural ischemia.

0,390ug/l. NT pro B-type natriuretic peptide was 137,8pg/ml and D-dimers 0,36mg/l, a complete blood count was normal. The symptoms of patient, TTE with regional wall motions abnormalities finding and elevation of hsTn-T over 99 percentile are consistent withdefinition of acute STEMI. Selective coronary angiography showed normal findings. No coronary artery anomalies were detected (Figure 4), which raised the possibility of spasm or thromboembolism. Serial electrocardiograms showed the typical changes of acute myocardial infarction (Figure 5).

Ultrasonographic examination of lower extremities did not confirmsuspicion of deep venous thrombosis. Cardiac magnetic resonance imaging (CMR) perfomed thereafter

showed a patent Fontan pathway, no thrombi and normal pulmonary arborization. The left ventricular ejection fraction was 50% butthe late gadolinium enhancement showed the clear signs of past inferior transmural ischemia (Figure 6). The patient had a complete haematological examination, without finding of thrombophilia. Recommended therapy on discharge was: acetylsalicylic acid100mg, clopidogrel 75mg, inhibitor of proton pump 20mg, sotalol 2x80mg, cardilan 175mg and atorvastatin 80mg. Because of the second type of MI and the trombus formation in IVCseen on the first TTE, antithrombotic therapy was changed in outpatient care for warfarin without evidence of thrombotic reccurenceduring a follow-up ogthree yearswith this secondary prophylactic strategy.

Discussion

The Fontan procedure was first performed in 1968 in patient with tricuspid atresia and has transformed the lives of children born with single-ventricle physiology [1]. The earlier Fontan connected the right atrial appendage to the pulmonary artery (PA). The current procedure is an extracardiac conduit going from the inferior vena cava to the PA, and a Glenn connection joining the superior vena cava to the right PA [2]. Most of patients with Fontan circulation are surviving into adulthood. However, due to the abnormal circulation, they have an increased risk of complications. The frequent late complications are arrhythmias, thromboembolism, protein-losing enteropathy, heart failure and plastic bronchitis [3,4].

Table 1: Summary of case report´s findings of embolic MI incidence after Fontan procedure.

Author	Basic diagnosis before FP	Type of MI	Age in admission	Antitrombotic treatment used before admission/after discharge	Recurrence of trombembolic event reported
Hastings et al.(7)	Pulmonary atresia, severe right heart hypoplasia	Inferior STEMI	19 yo	Aspirin/Combined aspirin and warfarin therapy	No
Deshpande et al.(8)	Tricuspid atresia,coarctation of the aorta, single ventricle	Anterolateral STEMI	17 yo	NA/NA	NA
Subahi et al.(9)	Hypoplastic left heart syndrome	Anterolateral STEMI /the only one with stent implantation	10 yo	aspirin/aspirin, clopidogrel and warfarin	No
Shamoon et al.(10)	Hypoplastic left heart syndrome	Inferolateral STEMI	24 yo	NA/aspirin, ticagrelol and warfarin	NA
Noonan et al.(11)	Hypoplastic left heart syndrome	Inferior STEMI	3 yo	None/aspirin and warfarin	NA
Wilson et al.(12)	Atresia of tricuspid valve, ventriculoparterial discordance, coarctation of the aorta	Anterior STEMI	3 yo	Aspirin/low dose aspirin and warfarin	Yes
Meier et al.(13)	Double outlet right ventricle, ventricular septal defect and straddling atrioventricular valves	Lateral STEMI	21 yo	NA/NA	NA

The most common etiological factors of thrombembolism are endothelial dysfunction, abnormal blood flow, hypercoagulability, presence of a right-to-left shunt [5]. The recognised locations of trombembolisation are caval veins, right atrium, pulmonary artery, venous chamber and cerebral arteries [6]. MI due to embolic occlusion of coronary arteries post-Fontan surgery is very rare and was described only in a few patients with repaired hypoplastic left heart syndrome, patients with history of double outlet right ventricle, ventricular septal defect and patients with pulmonary atresia and severe right heart hypoplasia, who underwent extra-cardiac Fontan operation. Table 1 shows a summary of published case reports dealing with this topic.

We present a unique case of embolic inferior STEMI in a 22 years old patient with fenestrated Fontan circulation due to double inlet left ventricle, ventricular septal defect and pulmonary stenosis. The potential source of embolism was the thrombus formation located in the IVC. We suppose that after the heparin and dual antiplatelet treatment in the acute phase of MI, the coronary bed was without any residual thrombotic occlusions. Moreover, MRI and ultrasound examinations didn´t confirm any residual vascular or intracardiac trombus formation. However, late gadolinium enhancement showed clear signs of past inferior transmural ischemia. The thrombus formation could be caused by the sluggish circulation and stagnation as a result of absence of the right ventricular pump. Alterations of coagulant factors are also one of the explanations but the hematological examination didn´t prove any anomaly. To date, however, there has been no reported difference in the incidence of thrombotic events between fenestrated and nonfenestrated Fontan circuits in recent trials [7]. The tromboprophylaxis after Fontan repair is very important to prevent thromembolism [8]. Up to date, there was no significant proven evidence that anticoagulation with warfarin is superior to aspirinin preventing thromboembolic events in patients with Fontan circulation [9,10]. However, in our case, there was the thrombus formation while on antiaggregation monotherapy with aspirin. Therefore, the treatment with warfarin was started without any recurrence of thrombosis during the three years period of anticoagulation therapy. Anticoagulation monotherapy was chosen instead of simultaneous treatment with warfarin and aspirin used in previous reported cases, because of the higher risk of bleeding.

Conclusion

The main goal of this case report is to highlight the potential need of anticoagulation therapy instead of antiplatelet treatment in primary and secondary prevention of thrombembolic complications after Fontan repair. There is a need for a large multicentered randomized control trials comparing the two treatment strategies and focusing on the perspective of new oral anticoagulants. Secondary, we want to emphasize, that acute MI of embolic origin is one of the very rare and life threatening complications of Fontan circulation, even in very young patients, regardless of basic diagnosis requiring Fontan operation in childhood.

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