Left ventricular thrombus in a patient with congenitally corrected transposition of the great arteries

Skrzeplina w lewej komorze u pacjentki z wrodzonym skorygowanym przełożeniem wielkich pni tętniczych

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Abstract

A 27 year-old woman with congenitally corrected transposition of the great arteries (ccTGA), which had been diagnosed five years previously, was admitted to our department because of severe heart failure and a suspected left ventricular thrombus. During the emergency operation, thrombi were removed from the left ventricle and tricuspid annuloplasty was performed. To the best of our knowledge, this is the first case report describing thrombus formation in the pulmonary ventricle in a patient with ccTGA. Most probably, the coexistence of multiple risk factors contributed to the thrombus formation.

Key words: congenitally corrected transposition of the great arteries, thrombus, oral contraceptives, systemic ventricular failure, tricuspid regurgitation

CASE REPORT

A 27 year-old woman with congenitally corrected transposition of the great arteries (ccTGA) was transferred to our department from the cardiology unit, because of hyperechogenic structure in the left ventricle. On admission, she was in sinus tachycardia 120 bpm, with blood pressure 100/60 mm Hg. Due to severe heart failure (NYHA class IV), the patient was maintained with the administration of oxygen, digoxin, furosemide, heparine and dobutamine. Laboratory tests showed: normal platelet count (191 $\times$ 10$^3$/µL), with slightly elevated INR (1.55), fibrinogen (3.74 g/L) and D-dimers (301 µg/L), as well as diminished antithrombin III (36.8 L%) levels.

The ccTGA had been diagnosed five years earlier when her heart failure symptoms had first appeared. The patient had been regularly seen by a cardiologist with great experience in congenital heart diseases and maintained in Warnes-Somerville functional class 2–3. Combined oral contraceptive pills had been started four weeks before admission. The patient had been confined to bed for the previous two weeks due to circulatory deterioration. She had been given standard anti-thrombotic prophylaxis with subcutaneous fractional heparine. No history of deep venous thrombosis was present. Chest computed tomography excluded a pulmonary embolism.

Transthoracic echocardiography revealed a typical ccTGA anatomy, with atrio-ventricular and ventriculo-arterial discordance. The anatomical right ventricle (RV) (systemic ventricle) was severely dilated, with sphericity index 1.2 and severely depressed global systolic function (Fig. 1). The left atrium was severely dilated with prominent appendage. Severe regurgitation of the anatomical tricuspid valve (systemic atrioventricular valve) was present, with dilated annulus and restricted leaflets motion secondary to RV enlargement (Fig. 2). The anatomical left ventricle (LV) (pulmonary ventricle) was normally sized with mildly depressed global systolic function. Hyperechogenic structure (diameter 2.2 $\times$ 3.3 cm) was seen in the LV apex (Fig. 1). When the ultrasound probe was tilted...
in apical views, an additional small round mass attached to the basal interventricular septum (IVS), could be seen. A small pericardial effusion was present around the right atrium.

The patient was operated on using cardiopulmonary bypass, cold crystalloid cardioplegia, and in general hypothermy (28°C). The thrombi, covered with a fibrous pouch (confirmed by post-operative histopathology), were totally removed from the apical and basal septal LV regions. Tricuspid annuloplasty was also performed using a mitral artificial ring (Carpentier-Edwards 32 mm).

The initial post-operative course was complicated by severe low cardiac output syndrome requiring high doses of inotropic support. Rethoracotomy was performed on the fourth post-operative day because of excessive bleeding. The patient remained on prolonged mechanical ventilation, and a tracheotomy was performed ten days after the operation. Acute renal failure (requiring haemodialysis) occurred in the later post-operative course. Rehabilitation proved extremely difficult due to poor co-operation from the patient combined with depression. After three months spent in the cardiac surgery department as well as in the rehabilitation unit, the patient was discharged home. She was subsequently monitored by our centre every three months. During the last control visit (18 months after the operation) the patient was in a very good condition, in Warnes-Somerville functional class 2, and had no symptoms of depression. However, echocardiography showed no improvement in systemic ventricular function, and significant tricuspid regurgitation despite the annuloplasty.

DISCUSSION

This is a 27 year-old woman who developed thrombus formation in the pulmonary ventricle. She had previously been diagnosed with ccTGA and had remained in Warnes-Somerville class 2–3 for about five years. Four weeks before the hospital admission, combined oral contraception had been started. For the previous two weeks, the patient had been confined to bed due to worsening heart failure, but standard anti-thrombotic prophylaxis was implemented.

To the best of our knowledge, this is the first case report describing thrombus formation in the pulmonary ventricle in a patient with ccTGA [1–3]. The patient had at least two moderate (chronic heart failure, oral contraception) and one weak (staying in bed for more than three days) risk factors for deep venous thrombosis (DVT) [4, 5].

Interestingly, she did not develop DVT nor a pulmonary embolism (most probably because of appropriate anti-thrombotic prophylaxis) but did develop thrombi in the pulmonary ventricle. Most probably, the coexistence of multiple risk factors contributed to the thrombus formation (bearing in mind the size and morphology of the thrombi) [4, 5]. The geometry of the pulmonary ventricle in ccTGA is severely altered due to bowing of the interventricular septum. Morphologic left ventricle in ccTGA supplies the pulmonary circulation and is exposed only to pulmonary pressures [2].

In this case, we observed LV cone-shaped apical region with apical and IVS akinesis and blood stasis seen on colour and Pulsed WaveDoppler. An LV endothelial injury secondary to regional wall motion abnormalities and blood stasis cannot be excluded. Temporary hypercoagulopathy (as a result of beginning oral contraception and immobilisation in bed) was the next point of the Virchow’s triad in this case.

Conflict of interest: none declared

References


