Case Report
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Early IVC thrombosis after central shunt surgery in young adult with uncorrected tetralogy of Fallot

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As there is paucity of data about thrombosis of inferior vena cava (IVC) in adult patients with uncorrected cyanotic heart defects, including those with tetralogy of Fallot, revealing and reporting of such cases is necessary for development of their proper clinical management.

Because cardiac surgery is relatively new in Azerbaijan late uncorrected cyanotic cases with subsequent complications are still an issue in our country.

Here we describe a case of IVC thrombosis after placement of systemic-pulmonary bypass (Davidson's shunt) in a 26-year-old patient diagnosed with Fallot's tetralogy. As there were signs of pulmonary embolism in early postoperative period, the patient has undergone vena cava filter placement.

Keywords: IVC thrombosis, congenital heart disease in young adult, systemic to pulmonary shunts, tetralogy of Fallot.

Introduction

Venous thromboembolism can be described as a state of deep venous thrombosis (DVT) with possibility of migration of thrombi through veins toward pulmonary artery.

Usually, in cyanotic congenital heart defects there is a connection between atria or ventricles that in case of regurgitation from right to left causes embolism of brain vessels with subsequent stroke [1, 2].

Inferior vena cava (IVC) thrombosis is quite alike DVT concerning etiology. The hypercoagulation state associated with hematological problems, neoplasm, venous stasis, surgery or a trauma is a main causative factor [3, 4].

Also patients with cyanotic heart defects have compensatory increased hematocrit that in turn increases blood viscosity, which contributes to easy formation of thrombi. There are also anomalies of coagulation and haemostatic, including those of fibrinolysis, in patients with congenital heart defects. The rate of mentioned anomalies is between 20 to 60%. Children with congenital heart defects that have hematocrit more than 60% often prone to develop thrombosis. [5]

Case Presentation

A 26-year-old male patient applied to our clinic complaining of breathlessness, cyanosis aggravated by motion, loose cough and stiffness in right extremities. His medical history revealed left-sided ischemic brain stroke 3 months before admission. Also, 2 months before admission he had an episode of massive haemoptysis. Physical examination revealed lean body constitution with weak development of subcutaneous fat and muscles. There were deformations of rib cage, acrocyanosis and nail clubbing. Oxygen saturation was 60-67% during the rest and inhalation of 2 l/min oxygen. Echocardiography confirmed diagnosis of Fallot's tetralogy with double vessels emerging from right ventricle (DORV Fallot type), and with hypoplastic pulmonary arteries. Right

(RPA), left (LPA) and main (MPA) pulmonary arteries measured 7, 4 and 16 mm, respectively. Contrast enhanced computer tomography (CT) revealed multiple aorta-pulmonary collaterals, thus further confirming the diagnosis. According to CT scans, the dimensions of pulmonary artery were determined as following: RPA – 9 mm, LPA – 4.5 mm, MPA – 18 mm. His blood and biochemistry tests were as following: haematocrit – 44.7%, heamaglobin – 14g/dl, platelets- 269 ×10^9/1, international normalized ratio – 1.18, prothrombin index – 78%, creatinine – 73 μmol/l, total bilirubin – 16.2 μmol/l, alanine transferase (ALT) – 9.3 IU/ml, aspartate transaminase (AST) – 38.8 IU/ml.

Weighing up all pros and cons the patient was recommended to undergo central aortopulmonary shunt (Davidson) procedure.

On 06.09.2017 patient had been placed the Davidson's shunt number 10. During postoperative period, he received heparin infusion at 15 U/kg/h rate [6]. Despite of recommended dose we couldn't get appropriate control of coagulation. His saturation during the rest and inhalation of 2 l/min oxygen increased up to 80%. The patient was mobilized and postural drainage was performed on 2nd postoperative day. Beginning from the 2nd postoperative day heparin infusion was stopped and the patient was administered Aspirin at 100 mg/day. On the 3rd postoperative day, the patient was transferred to in-patient treatment unit. On the 4th postoperative day, the scleral icterus, oliguria, hepatomegaly, apathy, breathlessness and decrease in saturation (despite of increase of delivery of oxygen up to 60%) were revealed. Blood tests were as following: total bilirubin – 60.4mkmol/l, unconjugated bilirubin – 36.4 μmol/l, conjugated bilirubin – 24 μmol/l, ALT – 87.5 IU/ml, D-dimer – 3500ng/ml). Echocardiography revealed big and mobile thrombus in IVC. Taking in account clinical signs and all possible risks, we decided on urgent placement of cava filter. The procedure was followed by transfusion of heparin with subsequent administration of Warfarine, Clexane and Aspirin. On the 9th day, the patient's blood tests were as following: total bilirubin – 29 μmol/l, ALT – 35.2 IU/ml, AST – 20.7 IU/ml. The patient was discharged without any clinical signs (his saturation was 78-85% without additional delivery of oxygen).

Discussion

It can be speculated that thrombosis in our case was caused by high viscosity of the blood, as a consequence of the main disease. Also patient denied mobilization in early postoperative period. As it is noted in the guideline of 2014th year [8], patients with venous thromboembolism are at high risk for ischemic brain stroke. That is also true for our patient; as he had stroke in the past probably due to DVT. We took in account the risk of repeated venous thrombosis. However, according to literature, the risk of subsequent venous thrombosis in such patients is 2% during 2 weeks, 6.4% during three months and 8% during 6 months, despite of usage of anticoagulants. Many patients with clinically manifesting DVT experience a thrombosis of pulmonary artery in 40-50% of cases. [7-9].

Husler et al., described 40 cases of thrombosis of inferior vena cava, including 6 patients with cyanotic and 4 patients with non-cyanotic heart defects [8].

Our patient developed thrombi despite of constant 2 days-long infusion of anticoagulant in sufficient doses. As there was a history of ischemic brain stroke, most probably the patient already had the DVT before admission. Also patient might have resistance to heparin. The most common cause of resistance to heparin is deficiency of anti-thrombin, which is necessary for anticoagulant effect of heparin. The deficiency can be congenital due to low synthesis, or it can be acquired because of disseminated coagulation syndrome or, due to high doses of heparin used during operation for extracorporeal circulation [10].

As there were not external factors listed above, we consider that our patient had congenital deficiency of anti thrombin. However, we don't have appropriate facilities to confirm this.

After placement of filter in IVC and administration of varfarine the thrombus has decreased in dimension and no episodes of recurrent thrombosis were noted.

Conclusion

According to described experience, one can speculate about high risk of deep vein thrombosis and IVC thrombosis in patients with cyanotic heart defects, particularly in adult cases. Predisposing factors may include disturbances of coagulation and insufficiency of anti thrombin and, hypodinamia, which we consider as a key factor in our case.

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