Case Report

Acute cardiac tamponade due to spontaneous bleeding in a child with Haemophilia A

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Abstract

In severe haemophilia A, patients, start from the first years of life, with spontaneous bleeding and require transfusion. However, cardiac tamponade due to spontaneous pericardial bleeding is rare. An 11-year-old boy receiving haemophilia A treatment was referred to the Department of Paediatric Haematology with pneumonia, fever, dyspnoea, and palpitation. In his PA chest radiograph, pneumonic infiltration in the right lung and enlargement in the pericardial area were found. On his echocardiograph, pericardial effusion reaching 3.9 cm and other findings of tamponade were detected. APTT was outside the measurable range. It was deranged to >120 seconds. The patient received 1000 U of factor VIII intravenously. A pericardial window was made via left anterior mini thoracotomy due to fluid drained.

In his control echocardiograph taken after one month, no pathology was found. At 50th day, the patient showed left pleural serohaemorrhagic effusion, which was treated with tube thoracostomy. In haemophilia A patients, either pericardiocentesis or subxiphoid pericardial drainage or pericardial window creation via thoracotomy may be applied, depending on the primary pathology. In paediatric cases, pericardial window creation via mini thoracotomy can be an alternative treatment of choice considering complications such as recurring bleeding and effusion during pericardiocentesis.

Introduction

Haemophilia A is an X-linked recessive bleeding disorder characterized by qualitative and quantitative deficiency of factor VIII resulting from heterogeneous mutations in the factor VIII gene (F8C) located in the Xq28 region. Small/large gene deletions, insertions and gross gene rearrangements underlie the molecular pathogenesis of the disease. Haemophilia A is more common than haemophilia B, representing 80-85% of the total. The number of persons affected worldwide is estimated to be about 400,000.[1]

A bleeding episode generally occurs before 18 months of age and may follow a minor injury in haemophilia patients. A child with haemophilia bruises easily, even an injection into a muscle can cause bleeding that results in a large bruise and haematoma.[2,3]

In severe haemophilia A, life-threatening haemorrhages may occur such as central neuron system haemorrhage, deep muscle, joint haemorrhage, ocular haematomas, retroperitoneal haemorrhage, bleeding following tooth extraction, postsurgical bleeding, easy bruising, and mucosal bleeding. However, spontaneous intrapericardial haemorrhage and tamponade occur rarely.[3]

Case Report

The patient was an 11-year-old boy diagnosed and followed up by the Department of Paediatric Haematology with severe haemophilia A whose factor VIII levels in the blood varied between 0.8% and 4.8%. He was hospitalized due to pneumonia with fever, dyspnoea and palpitation. There was no history of trauma. He had growth retardation, with weight 20 kg and height 125 cm. The body surface area of the patient was 0.835. On his PA chest radiograph, of right lower lobe, localized pneumonic infiltration and enlargement of the pericardial shadow was seen (Figure-1). On his echocardiograph, signs of tamponade and pericardial fluid 3.9 cm in size were found (Diastolic right ventricular collapse, pseudo systolic anterior motion on anterior mitral valve and

swinging heart) (Figure-2). Blood pressure was 90/50 mmHg, pulse rate 153/min and fever 37.6°C. APTT was markedly deranged. Haemoglobin level was 8.71 g/dL. No history of trauma or medical intervention was present. The patient received intravenous 1000 U of factor VIII. An anterior mini thoracotomy was performed through the fourth left intercostal space. The pericardium was strained and livid. A pericardial window was created over the left phrenic nerve path. Approximately two hundred cc fluid was drained. The colour of the fluid was haemorrhagic. All tamponade signs disappeared. No bleeding was seen in the cardiac structure or main vessels. Histopathological findings of the pericardium were non-specific and secondary to bleeding. Pericardial fluid was sent for microbiological examination, but was negative. On the first postoperative day in the intensive care, a total of 380 ml drainage was measured. On the second day, the patient was transferred to the ward and the total drainage was 70 ml/day. The haematocrit was 29.6 and haemoglobin was 8.9 g/dL following which one unit of erythrocyte suspension was transfused. APTT was 44.9 sec. On the third postoperative day, there was no drainage at all and the drain was removed. Starting at the eighth postoperative hour, initially 2 X 1000 U factor VIII was given intravenously for two days, and 3 X 500 U for the following 3 days. Factor VIII was given totally for 5 days. The patient was discharged from the hospital on the fifth postoperative day. There was no pathology on the echocardiograph after one month. On day 50, dyspnoea with left pleural serohaemorrhagic effusion was detected, patient was hospitalized and tube thoracostomy was done. During the procedure, replacement of the factor VIII continued. After 5 days, the drains were removed and the patient was discharged with full recovery. In the subsequent 12 months, the patient did not have any other major spontaneous bleeding episodes.

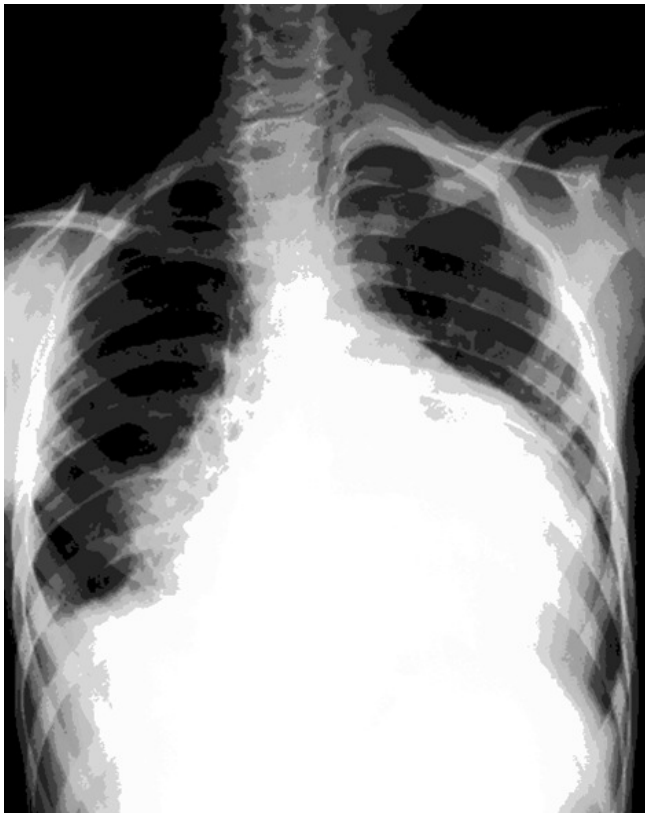

Figure-1: PA chest radiograph, showed that on the right lower lobe, localized pneumonic infiltration and enlargement on the pericardial shadow.

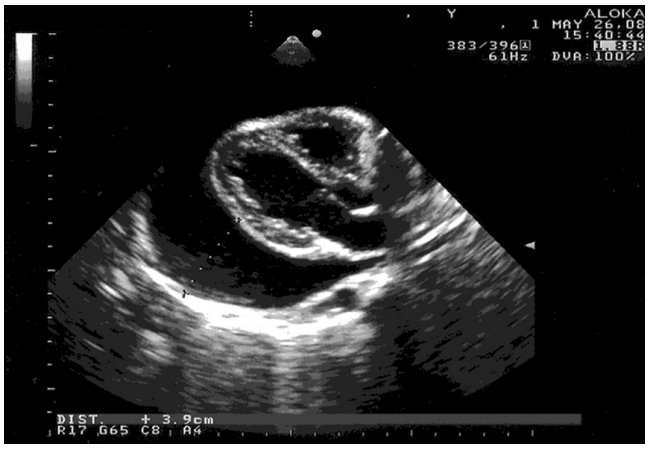

Figure-2: Echocardiograph, signs of tamponade (swinging heart and diastolic right ventricular collapse) and pericardial fluid 3.9 cm in size were found.

Discussion

The most common sites of bleeding in a person with haemophilia are the joints and muscles of the extremities. The bleeding episodes are frequent and spontaneous in severe haemophilia depending on the severity of disease factor levels. Prophylaxis is the standard of care for treatment of children with severe haemophilia and consists of regular infusion of factor VIII concentrates to avoid bleeding. Bleeding should be treated with factor replacement therapy at the earliest possible moment, preferably within 2 hours of the onset of the symptoms.

Cardiac tamponade is defined as a clinical entity in which the intrapericardial pressure elevates due to extra fluid accumulation, the diastolic ventricular filling is impaired and the stroke volume and cardiac output decrease. Since cardiac tamponade is life-threatening, early diagnosis and prompt therapy are needed. Critical tamponade is a form of cardiogenic shock where the symptoms and signs are related to the physiological degradation speed. The treatment of tamponade involves the immediate evacuation of the pericardial fluid.

One of the following interventions is applied: pericardiocentesis, subxiphoid drainage, or pericardial window creation via thoracotomy, depending upon primary

pathology. Physicians prefer pericardiocentesis as a better treatment approach in tamponade cases complicated by spontaneous pericardial bleeding, which rarely occurs. In contrast, we suggest that pericardial window creation could be a good choice, considering the complications that can occur during pericardiocentesis and at the follow-up, such as recurrent bleeding and effusion.[4,5]

Tamponade caused by spontaneous intrapericardial bleeding in haemophilia A patients is very rare. Still, haemophilia-associated haemorrhage is preventable if clotting factor replacement therapy is given to achieve and maintain normal physiological levels of factor VIII. In the presented case, even though the factor levels were routinely measured and replacement therapy was applied to prevent bleeding, the factor level decreased below the critical level due to an infection in the respiratory system. Therefore, before starting the surgery, blood tests such as, blood factor levels, APTT, fibrinogen, prothrombin time, bleeding and coagulation time, platelet count, liver function tests and fibrin degradation products should be performed. In our case, the needed factor VIII was given intravenously before the operation and continued in the days following the operation. In severe haemophilia A cases, pain control during the invasive procedures is very important. Therefore, all products that cause platelet dysfunction, especially those containing Acetylsolicylie Acid (ASA), should be avoided and non-steroidal anti-inflammatory drugs should be used with caution. Paracetamol, with or without narcotic analogues, is usually effective in controlling pain. Also, intramuscular injections should be avoided. In the presented case, we preferred tramadol (Tramadol hydrochloride) for analgesia of the postoperative thoracotomy pain.

Conclusion

Severe haemophilia A patients can develop spontaneous pericardial bleeding causing tamponade, which should be treated with replacement therapy and pericardial window creation. This is a good choice in paediatric patients to avoid early and late complications.

Referances

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