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Acquired hemophilia A , Unusual age onset: young girl with Intramuscular Hematoma

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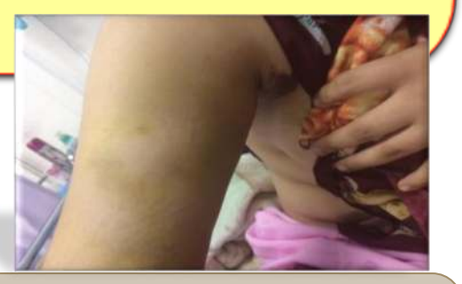
Background

Acquired factor VIII (FVIII) deficiency as a life threatening antibody mediated autoimmune disease is rare. FVIII is the most common target of autoantibodies which involve in clotting activity, although the particular reason for this immunologic interference is not still clearly defined. In light of recent efforts, some dark aspects of the pathophysiology of the disease are discovered and some authors suggested the role of genopolymorphisms and CD4+ T lymphocytes in triggering the autoreactive cascade of this autoimmune condition.[1,2] Regarding the predisposing factors of the disease, it has been observed that the pathology mostly flares up in females older than 50 at postpartum periods.[3] In addition, rheumatoid arthritis, malignancy, systemic lupus erythematosus, and drug reactions, have been suggested as other predisposing factors. However, in some subjects, no underlying condition could be found (idiopathic type). [2,4] We present a case of FVIII deficiency whose clinical manifestations of disease were intramuscular hematoma and epistaxis.

Case presentation

A 13-year-old girl with no significant medical history presented with frequent epistaxis from 2 weeks ago that takes long time to stop bleeding.She was visited in the emergency department of Imam Khomeini Hospital with painless swelling, ecchymosis and pain in her right arm. She had not experienced any trauma or fever and had not undergone any operation in the past month. She doesn't have personal or familial history of bleeding disorders. Her problem was initiated 2 weeks ago with unusual epistaxis and diffused ecchymosis in dorsal side of left leg with swelling and pain. After first visit she was refered to surveyed in central hematologic department . On examination, her blood pressure was 110/80, heart rate was 106 beat per minute and body temperature was 36/8.There was antecubital fossa ecchymosis (size: 5cm×4cm) and right axillary fossa ecchymosis (size: 10cm* 6cm). Non-pitting swelling in her arm was disseminated right hemithorax; without swelling of hand and forearm. Radial and ulnar pulses were normal. There was no pain and abnormal neurologic finding in distal of upperlimbs. According to ultrasonography study; jugular venous and proximal of subclavian vein were compressible and had normal flow. Doppler sonography veins and artries of right upper limb was normal and hematoma in soft tissue. Following the mentioned examinations, disseminated superficial ecchymosis was accepted. Patients' coagulation test and factor assay Laboratory test resultsrevealed a hemoglobinlevel: 5,6g/dL (normal:12-16 g/dL), hematocrit:16.1% (normal: 36-48%), a von Willebrand factor activity: 93% (normal: 50-160%),aPTT level: 104.6 seconds (normal: 32-40 seconds), mixed .aPTT: 52 seconds (high- not corrected), PT level: 13.4 seconds (normal: 12.3-14.5 seconds), Bleeding time: 3minute (Normal: 2-7 minute), Factor 8 inhibitor: 16.8 Bethesba (<0.4 none). Plateletcount and liver function were also within the normal ranges.



Activity of factor 2, 5,7, 9, 10 and 11was normal. Anti-TPO: 1.8 (negetive), ANA: 2.3 (negative<10),Anti dsDNA: 0.6 (normal<100), Anti Phospholipid IGM: 2.7(negetiv<10), Anti Phosphplipid IgG: 2.6 (negative<10), Anti cardiolipin IgM: 3(negative <7), Anti-cardiolipin IgG: 2.4(negative<10).These results confirmed the diagnosisof acquired hemophilia A. Treatment with IVIG and continued by oral prednisolon 1mg/kg/day and Immunosuppressive agent (cyclosporine) was started for controlling the symptoms.In respons to the treatment, the aPTT normalized and symptoms were relifed. FVIII level was increased (1%>>16%>>20%>>60%) at several month later and Inhibitor level was decreased.(16.8>>>8.29>>2.5>>1.09>>0.64). the patient fallowed by physical exam, coagulation tests and educated for symptoms.

Discussion

Acquired hemophilia A is an autoimmune mediated disease and antibodies that inhibit the factor or inactivated it. Although the reasons of producing these autoantibodies is un known, but some gen polymorphisms and autoreactive CD4+ T lymphocytes, may involved acquired hemophilia A is very uncommon.There is a scarcity of acquired haemophilia A studies from Asian countries. This study reveals a novel finding of younger age at diagnosis of acquired haemophilia A among Asian patients. The weighted mean (SD) age at diagnosis was 58.10 (16.96) years compared with 75.70 (14.47) years in the European series (absolute difference 17.6 years)·This clinical manifestation is in contrast with congenital hemophilia A, which commonly presents with hemorrhage in joint articulations, muscles and other soft tissues that are manageable with conservative treatment. Some of the rheumatic

disorders associated with AHA are rheumatic arthritis, systemic lupus erythematosus and autoimmune hypo/hyperthyroidism. To confirm the presence of AHA, the clinical indications must be exhibited in addition with the specialized laboratory testing such as the Factor VIII inhibitory assay and Bethesda Unit quantification assay. This patient met with the previous prerequisites. This is followed with the ruling out of anticoagulant medications as the culprit of the bleeding episodes. It is important to note, however, some possibilities that can explain the diagnosis in this patient. One of these would be the presence of a potential neoplasia in the liver that could have given way to the appearance of AHA in this patient.

Finally, the difficulty in implementing hemostasis with conventional treatment protocols in patients with this anomalous condition needs to be considered. Training hospitalists and family medicine specialists in recognizing the clinical appearance of this bleeding disorder is key in determining the swift resolution of this treatable autoimmune disease.