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Hemophilia a with Left Knee and Elbow Hemarthrosis: A Case Report

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ABSTRACT

The most significant characteristic of hemophilia A, an X-linked recessive bleeding disorders, is a deficiency of factor VIII clotting activity, which results in prolonged bleeding after wounds, tooth extractions, or operations, as well as delayed or recurring bleeding before the wound heals fully. Less than 1% latent factor activity in severe sickness results in bleeding symptoms, predominantly in the joints (hemarthrosis) but also occasionally in the muscles, soft tissues, and potentially lethal places (e.g., central nervous system), frequently without any visible injuries. This is a case of 6-year-old male child with severe hemophilia of factor VIII activity less than 1% presented with unilateral hemarthrosis of left knee and elbow. The child has a history of multiple infusions of factor VIII, despite having multiple infusions the child has shown recurring episodes of hemarthrosis. The child was indicated with replacement therapy, limb elevation, cold compression and immobility. About 80% of individuals who experience recurrent joint bleeding develop joint injury, with the knees, elbows, and ankles being the most frequently affected joints. Joint dysfunction, persistent discomfort, and a decreased quality of life are the hallmarks of hemophilic arthropathy, a debilitating illness. This case highlights the need for careful monitoring and tailored therapies to maximize outcomes for young individuals with severe hemophilia A.

KEYWORDS: Hemophilia A, hemarthrosis, clotting factor VIII, Von Willebrand Factor, APTT.

INTRODUCTION

Hemophilia A is an X-linked recessive bleeding disorder with main characteristic feature of a deficit of factor VIII clotting activity, which causes delayed or recurrent bleeding before the wound heals completely and prolonged bleeding following wounds, teeth extractions, or surgeries. Sometimes it may also be confused by Von Willebrand disease or deficiency of factor XI. Severe hemophilia A usually appears in the first two years of life amid soft tissue or oral bleeding, either as an outcome of procedures or because of a known family history of hemophilia. In the absence of preventative care, people may experience two to five episodes of spontaneous bleeding each month on average. Severe illness, with less than 1% latent factor activity, causes manifestations of bleeding, usually without apparent trauma, mainly in the joints (hemarthrosis), however, occasionally in the muscles, soft tissues, and susceptible to fatal areas (e.g., central nervous system) (1). Frequent hemarthroses trigger hypertrophic

synovitis, gradual reduction of cartilage, and the long-term consequences of hemophilic arthropathy, which include joint mobility deterioration and chronic discomfort (2). While the exact pathophysiology of hemophilic arthropathy is unknown, it seems to possess similar characteristics with the inflammatory processes linked to rheumatoid arthritis and the progressive degeneration of joints that results from osteoarthritis. Apart from clotting factor concentrates, which are typically prescribed by the hematologist, the orthopedic surgeon and physiotherapist must work closely together to manage acute hemarthrosis, the collaboration of all these specialists with consistent therapy are main role in the management of hemarthrosis, assessing the joint mobility and relieving joint discomfort. Intravenous injection of inadequate clotting factor is the basis for the treatment of hemophilia. This is also known as replacement therapy.

CASE DESCRIPTION

A 6-year-old male child 2nd in birth order, born out of non-consanguineous marriage, previously diagnosed for severe hemophilia A (<1%) at the age of 1year, presented with swelling of left knee joint and left elbow since 4 days, diffused swelling, restricted movements, tenderness at affected region, left knee girth of 27cms, elbow girth of 17cms. Fever since 1day, low grade, on and off, aggravated by pain in joint and relived by medication. On admission the child was afebrile, pallor, and all other vitals were within normal range. History of similar complaints at age of 4years admitted in hospital and administered factor VIII, repeated similar complaints on and off every 2months (11 times), h/o factor VIII administration 11 times, blood transfusion of 1-pint packed cells a year ago. A family history of similar complaints in his uncle of age 40 years.

Table 1: parameters observed at the age of 1 year

Tests	Presented value at age of 1year
PT	12.4sec
INR	0.98sec
APTT	83.9sec
Thrombin time	18.4sec
Plasma fibrinogen	228mg/dl
Factor VIII	<1%
Von willebrand factor	106.5%

The laboratory findings on admission revealed that there was a remarkable increase in the APTT (76.2 sec), with the International normalized ratio and prothrombin time (INR -0.99sec; PT-10.9sec). It is found that the factor VIII activity has significantly low (<2%), and inhibitor negative which is a favorable as the inhibitors doesn't affect the replacement therapy for clotting factor. Initial laboratory tests revealed mild anemia (hemoglobin 9.8 gm/dL) with low mean corpuscular volume (MCV) and mean corpuscular hemoglobin (MCH), with low hematocrit (HCT), while WBC count(8810/µL) were within the normal range. The laboratory findings at the age of 1 year old were International normalized ratio and prothrombin time (INR -0.98sec; PT-12.4sec), APTT (83.9sec), Factor VIII activity was found to be <1%, and Von Willebrand Factor (VWF) activity was 106.5%. Ultrasound scan showed the signs of bleeding in knee joint.

Initially the patient was administered with factor VIII injection at 50mg/kg, (770IU/Slow IV/stat), followed by 20mg/kg (310IU/slow IV/stat), injection paracetamol at 10mg/kg/dose (16ml/IV/QID, 1ml=10mg), followed by 16ml/IV/SOS. Limb elevation, Cold compression and immobilization are suggested. Regular monitoring of vitals was performed. Tablet tranexamic acid 250mg/PO/BD are administered. The patient was scheduled for follow-up appointments and regular care for the arthrosis and regular monitoring of factor VIII activity.

DISCUSSION

A deficit or the absence of coagulation factor VIII (FVIII) is the hallmark of hemophilia A, an X-linked recessive bleeding disorder that increases the risk of bleeding episodes, especially into joints (hemarthrosis). Hemophilic arthropathy, a persistent and incapacitating illness, can result from recurrent hemarthrosis and spontaneous bleeding in severe cases, where FVIII activity is less than 1% (3).

Recurrent joint bleedings induce joint damage in about eighty percent of the patients, with knees, elbows, and ankles as preferential target joints. Hemophilic arthropathy is a disabling condition characterized by joint impairment, chronic pain, and reduced quality of life (4).

The disease condition Is classified as mild, moderate, or severe based on the different levels of FVIII insufficiency. Basal levels of FVIII of less than 1% indicate severe hemophilia, those between 1% and 5% indicate moderate hemophilia, and those between 6% and 40% indicate mild hemophilia. In spite of getting FVIII replacement therapy eleven times, the 6-year-old boy with severe hemophilia A presents with recurrent hemarthrosis. Concerns over the effectiveness of the current treatment plan and despite being negative inhibitor which may not cause any effect in replacement therapy, are raised by the continuance of joint bleeding (5).

Individuals with severe hemophilia are especially symptomatic and manifest spontaneous bleeding from an early age, mostly in the musculoskeletal system (large joints and muscles) (80% to 90% of profuse bleeding events), with mucosal or cerebral bleeding occurring occasionally. Hemarthrosis, or intra-articular bleeding, results in cartilage degradation and synovial enlargement, which leads to hemophilic arthropathy, a progressive but irreversible joint deterioration. (6)

To treat hemophilia, inadequate clotting factor is injected intravenously. This is also known as replacement therapy. To treat or prevent occasional bleeding episodes, the therapy can be given on-demand in multiple sessions. Replacement therapy is given prophylactically (or preventively) to individuals who are severely affected; this usually involves two to three injections per week (7).

In these situations, managing recurrent hemarthrosis calls for a diverse plan of action. The mainstay of care to avoid episodes of bleeding and joint injury is prophylactic FVIII therapy, which is periodically given to keep FVIII levels above 1%. Alternative treatments such emicizumab, a monoclonal antibody that mimics FVIII function, may be tried for patients who are on inhibitors. Additionally, patients with persistent synovitis who do not respond to conservative therapy may benefit from surgical procedures such as synovectomy (8).

This case highlights the need for careful monitoring and customized interventions to maximize outcomes for children with severe hemophilia A. Early and accurate diagnosis, as well as individualized treatment plans, are critical in managing severe hemophilia A and preventing long-term complications. Comprehensive management includes multidisciplinary care involving hematologists, orthopedic specialists, and physiotherapists, as well as regular monitoring of FVIII levels and joint health assessments.

CONCLUSION

The characteristic of hemophilia A, an X-linked recessive bleeding condition that raises the possibility of bleeding episodes, particularly into joints (hemarthrosis), is a deficiency or lack of coagulation factor VIII (FVIII). Recurrent hemarthrosis and, in severe cases, spontaneous bleeding can cause hemophilic arthropathy, a chronic and debilitating condition. To optimize results for children with severe hemophilia A, this instance emphasizes the necessity of close observation and tailored therapies. To manage severe hemophilia A and avoid long-term consequences, early and correct diagnosis is essential, as are customized treatment programs. The regular patient

follow up should ideally schedule. In addition to routine FVIII level monitoring and joint health evaluations, comprehensive management entails multidisciplinary care involving hematologists, orthopaedic specialists, and physiotherapists.

Declaration by Authors

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