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Hemophilic Pseudotumor Of HIP: A Case Report

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I. Introduction

Hemophilia is a spectrum of clotting disorders that is caused by deficiency of factor VIII (Hemophilia A) and factor IX (Hemophilia B) respectively. Clinical features varies with the severity of the disease. Severe type of hemophilia usually presents with spontaneous bleeding into joints and muscles. Hemophilic pseudotumor develops as a result of repeated episodes of bleeding at a fracture site in bone or as a result of subperiosteal haemorrhage or bleeding into soft tissue. Inadequate resorption of extravasated blood results in an encapsulated area of clotted blood and necrosed tissue. With successive hemorrhagic episodes, these lesions increase in size over time and eventually causing symptoms over mass effect.

Majority of haemophilic pseudo-tumors that have reported have been involving the musculoskeletal system. Although their incidence have decreased over time with advanced mode of treatment of haemophilia. Still these pseudo tumors are significant in cases of hemophilia. In this case report we describe a patient with massive pseudo tumor in a known case of Hemophilia A which involves the pelvic bone with intra abdominal extension.

II. Case Report

Our patient is a 38 year old male with Factor VIII deficiency who presented to our institution in out patient department with complaints of swelling over right hip and difficulty in walking since last 3 years. In his childhood he had several episodes of bruise over his body with trivial trauma for which he underwent investigations as advised by physician and he was eventually diagnosed with Hemophilia A. Throughout his childhood he had several episodes of hemarthrosis and eventually developed fibrosis in bilateral knee joints. In his early adulthood since last 15 years he started having swelling over right iliac region which gradually increased over time. Initially he was having a feeling of fullness over the right iliac fossa. It was associated with pain in right hip region. Patient sought medical care then and he underwent radio logical investigation which was inconclusive and then further MRI was done which confirmed presence of sinusoidal fluid filled lesion over right iliac bone. Patient took factor replacement regularly and the swelling was managed conservatively.

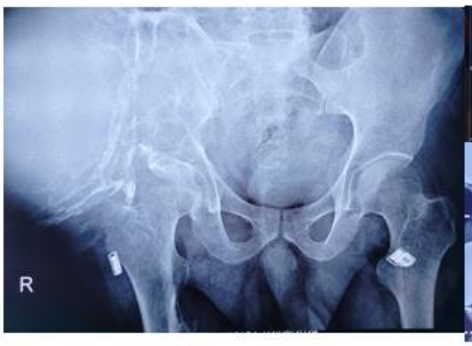

Fig no 1 : X ray showing Right Ilium pseudotumor

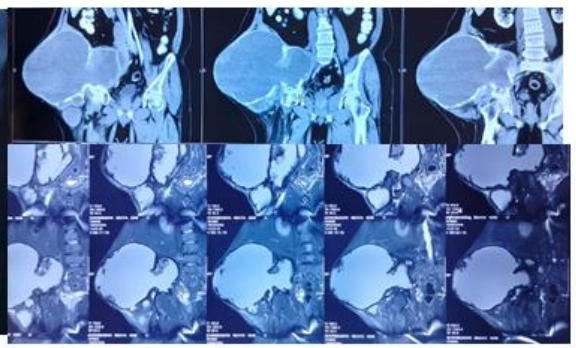

Fig no 2: CT Scan showing right Ilium homogeneous mass hyperintense with contrast

Since last 3 years the swelling rapidly increased in size and is associated with pain and difficulty in walking. Pain however worsened over time and then patient sought medical care. Radiological investigations were done which is suggestive of a large homogeneous radiopaque lesion

overlying right iliac bone extending laterally and subcutaneously. A fresh MRI was done which is suggestive of a large expansile lytic multi cystic lobulated lesion of about 26*18*25 cm involving right iliac bone, acetabulum, right sacroiliac joint, right Ala of sacrum extending intra abdominally. A CT Angiogram was done to assess the vascularity which is suggestive of haemophilic pseudotumor with multiple clots and having calcified wall. The lesion is supplied by right external iliac artery and right superficial femoral artery. Considering the investigations, a surgical intervention was planned. In the mean time patient continued receiving factor replacement at regular intervals. Pre operative angioembolisation of the feeder vessels were done. Intra operatively, a large 30*25*25 cm lesion was seen over lateral aspect of right iliac fossa involving lateral aspect of right iliac bone overlying the inguinal ligaments and extending through the greater sciatic notch into abdominal cavity. The capsule of the tumour is found adhered to the peritoneum. Volume of the tumour cavity is about 5 liters. Multiple clots are found adhered to the tumour wall. Tumor debulking is done.

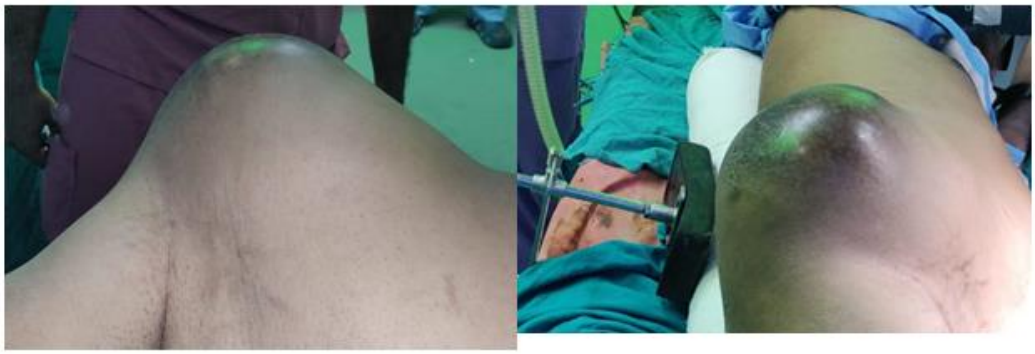

Fig no 3: Clinical picture of pseudotumor

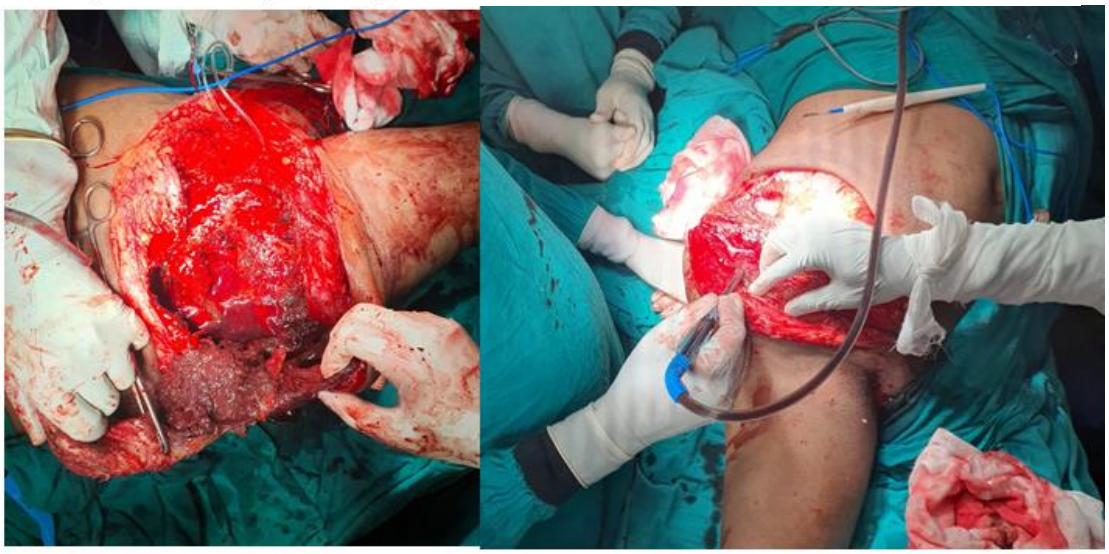

Fig no 4: Tumor cavity with thinned out bony margins

Fig no 5: Dark chocolate coloured fluid of pseudotumor

III. Discussion

The hemophilic pseudotumor was first identified by Starker, who reported the condition in the femur of a 14-year-old boy. These pseudotumors, also known as "blood cysts," are a rare complication of hemophilia, occurring in only 1% to 2% of patients with severe forms of the disease. Typically, affected individuals are diagnosed hemophiliacs who have experienced multiple hemorrhagic episodes at various sites over several years.

With the advent of factor replacement therapy, the frequency of pseudotumors may have declined. However, this therapy proves ineffective in patients who develop autoantibodies against factor VIII or factor IX, where bleeding episodes are often more severe and can occur in a wider range of anatomical locations.

Pseudotumors are composed of extravasated, clotted blood encapsulated by fibrous tissue. While they most commonly affect bones, they have also been reported in the lungs, abdomen, and within the wall of the

stomach. The bones most frequently affected include those most prone to trauma, such as the femur, pelvis, tibia, and, less commonly, the small bones of the hand, listed in decreasing order of frequency.

IV. Conclusion

Hemophilic pseudotumors are a well-documented complication of bleeding disorders, and clinicians should remain vigilant for this condition when treating affected patients. Early evaluation of signs and symptoms of compression should include initial imaging with CT, followed by MRI for more detailed assessment. Once the diagnosis is confirmed, prompt surgical consultation and factor replacement therapy are crucial. In cases of advanced disease or complications, a conservative approach may be considered, but further decision-making should involve an interdisciplinary team.

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