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Photo Clinic

Hemophilic Pseudotumor of Humerus in an Adolescent with Severe Hemophilia A

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A 15-year-old boy, known case of severe hemophilia A, was referred with the complaint of a painful swollen shoulder. He had experienced several bleeding episodes in multiple joints. Because of developing high titer inhibitor, he had undergone a course of immune tolerance induction that was unsuccessful. As a result, since then he was managed on-demand by receiving bypassing agents for his bleeding episodes. On physical examination, decreased range of motion in right shoulder was remarkable. Anteroposterior radiograph of the shoulder showed a well-defined osteolytic lesion in the right humeral head surrounded by a thin sclerotic rim with incomplete septa-like structures mimicking a subchondral bone cyst. The radiography was considered as a hemophilic pseudotumor (Figure 1).

He was hospitalized and scheduled to receive factor eight inhibitor bypassing agent (FEIBA) for a couple of days to control hemostasis and improve the range of motion of the shoulder joint. After about a week, range of motion of the shoulder joint was improved. He was scheduled to receive physiotherapy sessions under bypassing agents. He was considered as a candidate for curettage and bone grafting of the humerus pseudotumor.

Repeated bleeding into the joints often occurs in the first decade of life in patients with hemophilia. Joint degeneration is progressive, and although early treatment can slow the process, destruction of the joint is unavoidable. Hemophilic pseudotumors (HPT) are developed due to recurrent bleeding from extra-articular bone or soft tissues. The prevalence of HPTs is about 1–2%.[1]

On radiography, intraosseous pseudotumors produce a well-defined, unilobular or multilobular, expanding

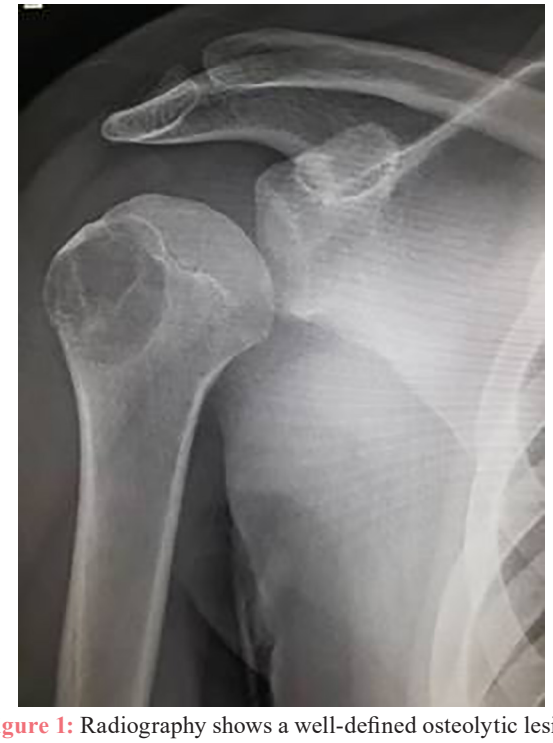

Figure 1: Radiography shows a well-defined osteolytic lesion in right humeral head surrounded by a thin sclerotic rim with incomplete septalike structures, considered as hemophilic pseudotumor.

lytic lesion of variable sizes. They occur in any part of the tubular bones; metadiaphysis or epiphysis.[2] Gilbert et al. described two clinical types of pseudotumors in patients with hemophilia. Proximal and distal pseudotumors. Proximal pseudotumors occur in the proximal skeleton, mostly around the femur and pelvis, while distal pseudotumors occur distal to the wrist and ankle. Distal pseudotumors are primarily seen in children and adolescents.[3]

Hemophilic arthropathy of the shoulder is a well-known complication in patients with hemophilia. MacDonald and colleagues reported rotator cuff tear as the most common abnormality noted in half of the symptomatic shoulders.[4] In a study to evaluate the clinical, radiographic, and ultrasonographic characteristics of the shoulder in a cohort of patients with hemophilia, seventy adult patients with hemophilia were assessed.[5] 35 patients had experienced shoulder bleeding. 18 (26%) of the 70 patients had abnormal radiographic findings. According to the Pettersson score, a variety of radiographic abnormalities including osteoporosis, osteophytes, narrowing of the joint space, subchondral irregularity and cyst formation, erosion of the joint margins, and bone remodeling could be defined.[6]

Subchondral cysts usually develop at joints which are weight bearing. MR imaging could help to differentiate blood from clear fluid before the cyst becomes obvious on the plain radiographs. Bone cysts caused by trabecular bone resorption are well visualized on CT scans.[7]

There are two types of subchondral and intraosseous cysts in patients with hemophilia. Subchondral cysts in hemophiliac patients have a direct connection with joint damage arthropathy. Intraosseous cysts are different because they are caused by an intraosseous haemorrhage, usually at the entrance of the bone nutrient artery. Unlike subchondral cysts, they have no connection to the joint and usually are not associated with arthropathy.[8]

Radiography is a sufficient technique to delineate the location of the subchondral cyst. CT scan is necessary for the preoperative planning, it discovers the size of the cyst, its actual location and the communication with adjacent structures. MRI informs about the cyst contents, which should be aspirated. When the cyst is in the active process, it is usually full of blood. However, when the joint is in an advanced degenerative process, the cyst is usually empty and there is no edema in the surrounding tissue.[9]

Conflict of Interest: None declared.

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