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HEMOPHILIC PSEUDOTUMOR: A CASE REPORT*

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Hemophilic pseudotumor has been defined as a progressive cystic swelling involving muscle which is produced by recurrent hemorrhage and accompanied by roentgenographic evidence of bone involvement. Pseudotumor is a rare complication of hemophilia, and, therefore, we present a case of a six-year-old male hemophiliac with a cyst in the left distal radius. *Key words: bone and joint changes, hemophilia, hemophilic cyst of bone.*

The term hemophilic pseudotumor describes a progressive cystic swelling involving muscle which is produced by recurrent hemorrhage and accompanied by radiographic evidence of bone involvement[1]. Masses develop most commonly in the pelvis, thigh or calf and consist of a slowly expanding encapsulated coagulum of chronic and acute hemorrhage[2,3]. In this paper, we describe a hemophiliac with a cyst in the left distal radius.

Case Report

A six-year-old boy was admitted to the hospital with the complaint of swelling of the left arm. The diagnosis of Factor VIII deficiency was established when he was two years of age. He had suffered a trauma of the left arm nine months ago. Although, he had been symptom-free in recent months, a swelling appeared on the same site of his arm.

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Physical examination revealed no abnormalities except for swelling and skin necrosis on the distal part of the left arm (Fig. 1). Laboratory examination revealed a hemoglobin level of 11.5 g/dl, white blood cell count 11200/mm^3, and differential count within normal limits. The prothrombin time was 14 sec and partial thromboplastin time 100 sec. The platelet count was 175000/mm^3. He had mild hemophilia type A, with Factor VIII being more than five percent.

Roentgenograms showed a hemophilic pseudotumor in the distal part of the left radius (Fig. 2).

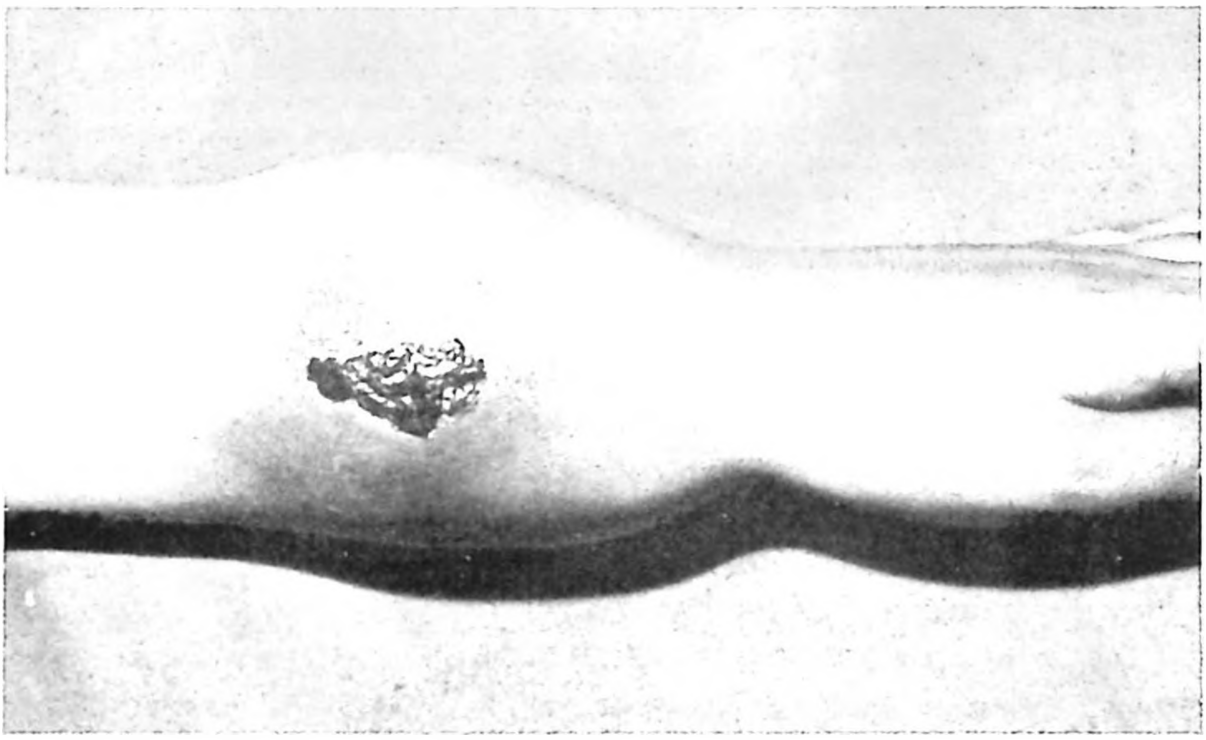

Fig. 1: Swelling and skin necrosis on the distal part of the left arm.

Fig. 2: Hemophilic pseudotumor on the distal part of the left radius.

Discussion

Pseudotumor is an unusual complication of hemophilia[3]. Gunning[4] estimated that one percent of severe hemophiliacs may have pseudotumors. However, its pathogenesis is not fully understood, and confirmed trauma is a factor in some patients[2].

Bone destruction has undoubtedly also played a role in the formation of pseudotumors, particularly in cases preceded by fracture. Intraosseous hemorrhage may also be a factor in the development of the hemophilic cyst, and with continued or repeated hemorrhage, these cysts may then increase in size causing expansion of the bone and thinning of the bony cortex. Reported cases indicate that lesions occur most commonly in the femur. However, they have also been reported in the pelvis, tibia, small bones of the hand, feet, calcaneus, humerus, olecranon and radius[2]. These lesions may also occur in more than one bone[3].

A pseudotumor can remain asymptomatic and unchanged for decades and then may suddenly be the site of bleeding and perforation. Fernandez de Valderrama[1] is of the opinion that there are three main types of hemophilic cysts involving muscle which can be defined by their mode of origin and subsequent course. Gilbert[5] described two types of pseudotumor: a proximal type located in the pelvis and a distal type tumor localized mainly in the hands and feet. The former type appears mainly in adults, while the latter, with a better prognosis, appears only in children.

Gross pathologic examination of these pseudotumors has revealed thick-walled cysts filled with old clot and serosanguinous fluid. The walls of the cysts are extremely rigid, firm, and thick. Microscopic examination has revealed multiple organizing hematomas consistent with both recent and old hemorrhage[6].

Characteristic roentgenographic features of a soft tissue mass containing coarse calcifications, periosteal elevation, new bone formation in the configuration of bony "struts", and varying degrees of bone destruction are usually sufficiently distinctive to indicate the diagnosis[2].

Non-invasive imaging of these lesions by computerized tomography and sonography have proved useful in diagnosis and in planning treatment[6-9]. Early periosteal elevation resembles that of osteomyelitis, Ewing's sarcoma and metastatic neuroblastoma. Also, the osteolytic bone destruction which occurs resembles the lesions seen in primary bone sarcomas, tuberculosis, coccidioidomycosis, echinococcosis, aneurysmal bone cyst, giant cell tumor, plasmacytoma (myeloma) and metastatic neoplasms[2].

Hemophilic pseudotumors have been treated surgically, conservatively, and with radiation therapy. Prior to the availability of Factor VIII transfusions, surgical treatment was usually fatal[4]. Surgery appears to be the treatment of choice when

the lesions have been present for many years and non-surgical management is unsuccessful[4]. Needle or aspiration biopsies may result in chronic fistulous tracks and surgical scars that fail to heal, in addition to increasing the possibility of secondary infection[2]. Brant and Jordan[10], who believe that doses of 1000 to 2000 rads result in an endarteritis in an acutely bleeding hematoma, have noticed regression of the soft tissue mass in acute lesions. However, radiation was ineffective in the treatment of pseudotumors with longstanding bone changes that had been present for years[2]. Our patient refused hospitalization, and no specific surgical treatment was given.

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