Case Report

Spontaneous Regression of a Hemophilic Pseudotumor

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Hemophilic pseudotumor is a lesion resulting from subcutaneous fat, interfascial, subperiosteal, and intrasosseous hemorrhage. This article reports spontaneous regression of a hemophilic pseudotumor affecting the femur in a 2-year-old boy.

CASE REPORT

A 2-year-old boy presented with left knee pain and swelling. One week prior to presentation, his mother noticed he was limping. The mother reported three episodes of left knee swelling over the past 6 months. The patient had a history of hemophilia A that was diagnosed after spontaneous gingival hemorrhage 10 months earlier. After diagnosis, the patient was treated with intermittent injections of 250 U of freeze-dried human blood coagulation factor VIII concentrate after minor traumas. Blood tests at that time were normal except for a prolonged partial thromboplastin time of 76.9 seconds. Partial thrombin time was normal. Factor VIII activity was 1.4%.

Physical examination demonstrated swelling and warmth of the supra-/sternal region without joint effusion or limitation of knee motion. Radiographs revealed an osteolytic lesion of the distal metaphysis of the left femur associated with a popliteal soft-tissue mass and periosteal reaction (Figure 1).

T1-weighted MRI (TR 500 msec, TE 20 msec) revealed a multilocular mass composed of two main components. The posterior half was characterized by three zones of different signal intensity: a surrounding rim of low signal intensity, a peripheral area of increased signal intensity, and a central area of isointensity. On T2-weighted MRI (TR 2000 msec, TE 80 msec), the central and peripheral areas showed high signal intensity. The anterior half of the mass had two zones of signal intensity: a surrounding rim and central area. The central area was homogenous, showing isointensity on T1-weighted images and high intensity on T2-weighted images. The mass was located deep to the quadriceps and hamstring muscles. The cortex of the femur was irregular and eroded. The bone marrow had slightly lower signal intensity than normal fat on T1-weighted images, but was isointense to fat on T2-weighted images (Figure 2). These findings represented hematoma between the periosteal new bone and cortex without bone marrow involvement.

A diagnosis of hemophilic pseudotumor was made. The diagnosis was confirmed on follow-up radiographs, which showed the healing process of the lesion.

Replacement therapy with 250 U of freeze-dried human blood coagulation factor VIII concentrate was performed 11 times over the following 6 months. The knee was not immobilized because the pain and swelling became less severe. Biopsy or surgery was not performed. Radiographic follow-up was performed every 2 months. The lesion healed spontaneously over a period of 14 months after diagnosis (Figure 3).

DISCUSSION

Hemophilic pseudotumor is an infrequent complication in hemophiliacs and is accompanied by radiographic evidence of bone involvement. The rate of pseudotumor in hemophiliacs has been reported to be 1%-2% in individuals with severe factor VIII or IX deficiency (clotting factor levels <1% of normal). Magallon et al reported patient age at the time of diagnosis averaged 40 years (range: 8-61 years). In the present case, the patient had moderate hemophilia, but the activity of factor VIII bordered on severe.

Pseudotumors in children are rare, and the clinical presentation is different. Most of these lesions occur distal to the elbow and knee in children. Our patient was younger than most reported cases. Most authors agree the formation of a pseudotumor differs based on the anatomic site involved. Histologic examination has shown that a pseudotumor is an encapsulated hematoma. A thick fibrous capsule surrounds the hematoma in varying stages of organization. Calcification and ossification may be seen within it.

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Matthews suggested the cyst in their case was subperiosteal in origin. Intraosseous hemorrhage also has been implicated as an initial cause of this condition. A subperiosteal hematoma of the femur resulting from hemorrhosis of the knee joint has been reported; the joint nearest the pseudotumor often is uninvolved. The MRI findings in the present case demonstrated the lesion started as a subperiosteal hemorrhage because the muscles, bone marrow, and knee joint were intact.

Computed tomography and ultrasonography are effective in detecting and evaluating pseudotumors. Magnetic resonance imaging has been used to evaluate pseudotumors in the bones and soft tissues of the pelvis and thigh muscles. Magnetic resonance imaging also has assisted in monitoring therapy response by detecting recent blood within the lesion and evaluating the size of the hematoma in regions difficult to access by physical examination.

The appearance of hematomas on MRI is controversial. Hahn et al. reported intra-abdominal chronic hematomas had a characteristic concentric ring sign with a thin, dark peripheral rim on all pulse sequences and a bright inner ring on T1-weighted images. The central core was intermediate intensity on T1-weighted image. These authors considered the peripheral rim corresponded to hemosiderin digested by phagocytic cells surrounding the hematoma. The bright inner ring was attributed to paramagnetic effects of hemoglobin degradation products such as methemoglobin. The central core corresponded to a gelatinous clot.

In an MRI study of intrapelvic hematomas, Yamashita et al. hypothesized the intermediate intensity on T1-weighted images resulted from a lysed clot containing breakdown products of methemoglobin rather than remaining deoxyhemoglobin. This assumption was based on the fact that both central and peripheral areas of the hematoma were hyperintense, although deoxyhemoglobin is hypointense on T2-weighted images. The MRI findings in the present case were similar to usual hematomas. Hematomas can undergo progressive breakdown, resorption, and dilution without additional bleeding. On the contrary, pseudotumors extend due to recurrent bleeding of different ages.

Management of hemophilic pseudotumors remains controversial. Magallon et al. reported replacement therapy was successful in only 2 of 15 patients, and therapy ultimately depends on careful case-by-case evaluation. They concluded surgical management was the most effective treatment for pseudotumor, although more conservative treatment should not be overlooked in selected cases.

Hilgartner and Arnold reported a hemophilic pseudotumor affecting the femur in a 2-year-old boy. As replacement therapy alone was not effective, radiation therapy was used. In the present case, the young age of the patient may be one reason why the pseudotumor regressed spontaneously, as it is likely the pseudotumor was detected at an early stage.

A review of the literature revealed clinical parameters such as young age, distal location, small size, and early diagnosis may indicate a better prognosis of pseudotumors. Imaging has not played a role in determining whether conservative treatment or surgery is indicated for these lesions.

REFERENCES