Orthopaedic Case of the Month

Elbow Pain in a 9-year-old Boy

Mathew Hamula MS, Nick Pappas MD,
Kristen Thomas MD, John Dormans MD

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History and Physical Examination

A 9-year-old right-hand-dominant boy presented to an outside clinic with a 1-week history of right elbow pain. He experienced trauma to the elbow several weeks before but did not believe this was the cause of his current pain. He and his parents denied any significant medical history, allergies, or medications.

On physical examination the patient was afebrile with stable vital signs. His right distal humeral region was swollen and tender to palpation. It was warm but without erythema or induration. His elbow ROM was limited by approximately 15° in terminal flexion and extension.

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Each author certifies that his or her institution approved the reporting of this case report and that all investigations were conducted in conformity with ethical principles of research.

M. Hamula, N. Pappas, K. Thomas, J. Dormans
University of Pennsylvania School of Medicine,
Philadelphia, PA, USA

N. Pappas
Division of Orthopaedic Surgery, Hospital of the University
of Pennsylvania, Philadelphia, PA, USA

K. Thomas
Division of Pathology, Children's Hospital of Philadelphia,
Philadelphia, PA, USA

J. Dormans (✉)
Division of Orthopaedic Surgery, Children's Hospital
of Philadelphia, Second Floor, Wood Building, 34th Street
and Civic Center Boulevard, Philadelphia, PA 19104, USA
e-mail: dormans@email.chop.edu

He had no micromotion tenderness at the elbow. Plain films of the elbow were obtained (Fig. 1) and interpreted as an age-indeterminate, nondisplaced fracture of the supracondylar humerus. A long arm cast was placed on his arm and the patient was instructed to return for followup in 1 month for a repeat evaluation.

During the next 3 weeks, he began to experience worsening elbow pain and intermittent low-grade fevers to 101.0 °F. He presented to our clinic after 1 month. Repeat elbow films were obtained (Fig. 2). MRI subsequently was performed (Fig. 3). Laboratory results, including leukocyte (WBC) count, erythrocyte sedimentation rate (ESR), and c-reactive protein (CRP), were obtained and were within normal limits.

Based on the history, physical examination, laboratory studies, and imaging studies, what is the differential diagnosis?

Imaging Interpretation

Initial plain radiographs of the elbow (Fig. 1) revealed a radiolucent lytic lesion in the distal humeral diaphysis with ill-defined margins and possible erosion of the medial humeral cortex. Additionally, there is unilaminar periosteal reaction medially and anteriorly. These images were interpreted as indicative of a healing supracondylar fracture of the humerus. Repeat radiographs obtained only 1 month later (Fig. 2) showed a noticeable increase in the size of the radiolucent lesion to include the lateral aspect of the distal humerus with more abundant periosteal reaction suggestive of Codman's triangle superomedially. There is also suggestion of a cortical break medially with adjacent soft tissue swelling and a possible effusion.

Fig. 1A–B (A) AP and (B) lateral radiographs show a lytic lesion in the right distal humerus.

Fig. 2A–B (A) AP and (B) lateral radiographs taken 1 month after initial films show the lesion has increased in size.

Fig. 3A–C Noncontrast T2-weighted (A) axial, (B) coronal, and (C) sagittal MRI scans of the right elbow show an increased signal located at the distal humerus. Periosteal elevation and soft tissue edema can be seen flanking the distal humerus.

A noncontrast MRI scan (Fig. 3) of the right elbow was taken with T-2 weighted axial (Fig. 3A), sagittal (Fig. 3B), and coronal (Fig. 3C) cuts showing a discrete area of high signal indicating marrow replacement over the area

corresponding to the lytic lesion on the plain radiograph. Codman's triangle is apparent with periosteal elevation on both sides of the distal humerus. Additionally, soft tissue edema is present. There is also a moderate effusion with no

fluid-filled levels consistent with blood and water layering in the joint.

Differential Diagnosis

Osteomyelitis
Ewing's sarcoma
Osteosarcoma
Lymphoma
Langerhans cell histiocytosis

Histology Interpretation

Open biopsy of the distal humerus revealed a sheet-like diffuse proliferation of loosely cohesive, intermediate to large polygonal mononuclear cells with abundant eosinophilic cytoplasm. Their nuclei often have irregular contours,

with frequent clefts and coffee bean-like longitudinal grooves (Fig. 4). These cells stained positive for CD1a (Fig. 5). The inflammatory background is comprised predominantly of eosinophils, with scattered lymphocytes and rare plasma cells. Multinucleated giant cells also are seen.

Based on the history, physical examination, radiographic studies, and histology, what is the diagnosis and how should it be treated?

Diagnosis

Primary musculoskeletal Langerhans cell histiocytosis (LCH).

Discussion and Treatment

Based on the history, physical examination, imaging studies which showed an enlarging lytic lesion in the distal

Fig. 4A–B (A) Original magnification and (B) 10× magnification of tissue specimen showing Langerhans cells surrounded by eosinophils (Stain, hematoxylin & eosin).

Fig. 5A–B (A) Original magnification and (B) 20× magnification confirming the presence of Langerhans cells (Stain, CD1a).

humeral metaphysis, and a histologic specimen that had Langerhans cells surrounded by eosinophils, a diagnosis of primary musculoskeletal LCH was made. The biopsy proved definitive in this case, ruling out the rest of the differential diagnoses.

The other potential diagnoses included osteomyelitis, Ewing's sarcoma, osteosarcoma, and lymphoma. Each of these did not fit in this case for one or more reasons. The clinical presentation of osteomyelitis can be similar to that of primary musculoskeletal LCH. In both, a patient may complain of fevers, localized pain, and have radiographs showing a lytic lesion of the bone with adjacent periosteal reaction [2]. Inflammatory markers (ESR, CRP, WBC count) also may be elevated in each, although usually more so with osteomyelitis [2]. Because of their similar presentations, the most reliable means of differentiating the two is through a tissue biopsy. In our case, the fact that no organisms were present on either Gram stain or culture and the histologic analysis failed to show abundant neutrophils or lymphocytes eliminated osteomyelitis as the culprit [2]. Another possibility, Ewing's sarcoma, is the second most common primary bone sarcoma in children [8, 10]. Like primary musculoskeletal LCH it can appear on radiographs as a lytic lesion in the long bone of a child with a prominent adjacent periosteal reaction [8]. However, it more commonly has an associated soft tissue mass and its histologic features are vastly different, with small round blue cells as opposed to Langerhans cells and eosinophils [8, 10]. Osteosarcoma was also high on the list of potential diagnoses given the finding of Codman's triangle. As with primary musculoskeletal LCH, osteomyelitis, and Ewing's sarcoma, it can appear as a lytic lesion in the metaphysis of a long bone with adjacent periosteal reaction. Again, biopsy is the key to differentiating osteosarcoma from eosinophilic granuloma. Under the microscope, osteosarcoma is characterized by the presence of osteoid produced by anaplastic cells varying in size with atypical mitotic figures. Osteoid has more irregular, amorphous and eosinophilic trabeculae than normal bone tissue [5, 8, 10]. The differential diagnosis also included lymphoma, but the histologic specimen showed no small round blue cells, the hallmark of lymphoma [6].

Primary musculoskeletal LCH is one of three known types of LCH. It is important to distinguish it from the other two types, Letterer-Siwe and Hand-Schüller-Christian disease, as primary musculoskeletal LCH has a dramatically better prognosis. Letterer-Siwe disease has an earlier age of onset and produces symptoms such as dermatitis, recurrent bacteremia, and diffuse lymphadenopathy. Hand-Schüller-Christian disease typically occurs in children younger than 3 years and may present with a triad of skull lesions, exophthalmos, and diabetes insipidus [1, 3]. Some patients have widespread visceral involvement. In

contrast, eosinophilic granuloma is benign, localized to bone, and often self-limiting [1, 3]. It often affects children 5 to 10 years of age and can present as a solitary or multiple lesions. Greater than 1/2 of all cases of eosinophilic granuloma involve the skull, spine, pelvis, ribs, and mandible [3]. When it does localize to long bones, it most commonly is found in the diaphyses [3], which is different from our case. Patients typically present with localized pain, swelling, and occasional fevers. The initial workup involves basic radiographic imaging and laboratory studies to look for inflammation (ESR, CRP, WBC count). As in our case, radiographs often show a lytic-appearing lesion surrounded by reactive bone. MRI is also useful, as it can determine whether there is any soft tissue involvement, which is less common with primary musculoskeletal LCH than either osteosarcoma or Ewing's sarcoma. The biopsy is definitive for primary musculoskeletal LCH when Langerhans cells are observed flanked by eosinophils [1, 3]. Once the diagnosis has been made, it is advisable to get a bone scan or skeletal series to rule out multiple lesions.

Primary musculoskeletal LCH often resolves spontaneously. Lesions typically begin to regress after 3 months but can take as long as 2 years to resolve completely [1, 3]. Nine months after biopsy, our patient was asymptomatic with a substantial decrease in the radiolucency of the lesion in the distal humerus and an intact cortex (Fig. 6). Most cases will resolve gradually on their own. However, a substantial number require additional intervention. In these instances, curettage with or without a bone graft can be performed and often is curative. Indications for curettage

Fig. 6A–B (A) AP and (B) lateral radiographs of the humerus taken 9 months after biopsy show near complete resolution of the lesion.

include risk of deformity or pathologic fracture from the lesion [1]. This procedure was not required in our case. Other less common treatment options include corticosteroid injections [4], chemotherapy [1, 9], and radiation [1, 9]. Local injection of corticosteroids such as methylprednisolone at the site of the bone lesion reportedly relieves pain and discomfort less than a week after the injection [4]. However it is unclear whether local steroid injections alter the natural history of localized LCH. Additionally, approximately 1/4 of patients will require a second administration [4]. It is useful as a first-line treatment because it has fewer complications and is more cost effective than surgery or radiotherapy [4]. Radiation therapy at doses of 150 cGy/day for 4 days has been used as a last-line defense once corticosteroid injection and curettage have failed, but it is contraindicated in children and rarely necessary for localized disease [1, 9].

Primary musculoskeletal LCH is an uncommon disease and can be challenging to diagnose at initial presentation, as it was in our case. Known as one of the great imitators, the presentation of primary musculoskeletal LCH is similar to many other disease processes, such as tumor or infection. The key to differentiating eosinophilic granuloma from other conditions is histology, using open or needle biopsy. Diagnosing primary musculoskeletal LCH can spare the patient from receiving potentially harmful treatment for a benign and self-limiting condition.

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