A Rare Case of Langerhans Cell Histiocytosis Presenting as Shoulder Pain

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Introduction

Langerhans cell histiocytosis (LCH) is a rare cancer derived from myeloid progenitor cells. It is characterized by histiocyte infiltration and proliferation, and may manifest as single or multi-system disease. The most commonly observed manifestation of LCH is bone involvement with one or multiple osteolytic lesions. Early detection and risk stratification are critical in appropriately treating LCH.

Here we present a rare case of LCH presenting as shoulder pain in the pediatric patient.

Case Presentation

History

A 10 year old female with no pertinent past medical history presented to the sports medicine clinic with a one month history of pain in her right shoulder and over the distal end of the clavicle. The onset of the pain was acute, and was first noted with an episode of night-time wakening. There was no reported history of trauma to the area. The pain was noted to be persistent, but not worsening. The patient reported pain that is worse at night. There had been no noted swelling or bruising. Conservative therapy with rest and ice had yielded no improvement. The patient and her mother denied associated symptoms such as fever, night sweats, weight changes, numbness/weakness/paresthesias as well as any other sources of joint or musculoskeletal pain.

Physical Exam

Physical exam revealed a patient in no acute distress with normal visual inspection of the shoulder and no appreciable changes such as edema, erythema, malalignment, or atrophy. Tenderness over the distal one third of the right clavicle was noted on bony palpation. However, the patient had no reproducible pain over the AC joint, SC joint, coracoid, or bicipital groove. She demonstrated normal active range of motion and strength of the bilateral shoulders, however experienced pain with right shoulder flexion, abduction, and external rotation. She also endorsed pain with passive range of motion at extremes of motion with both right shoulder flexion and abduction. Anterior and posterior load-and-shift tests were negative and the patient demonstrated a negative sulcus sign. Drop arm test, empty can test, Hawkins-Kennedy test, and Neer test were negative. However piano key sign was positive. Patient demonstrated normal neurological sensation.

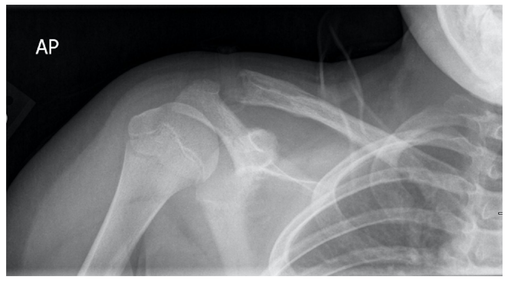
Bilateral clavicular X-Ray completed in office demonstrated thickening diameter with somewhat indistinct and irregular margins at the AC articulation. There was no noted periosteal reaction or focal areas of lucency.

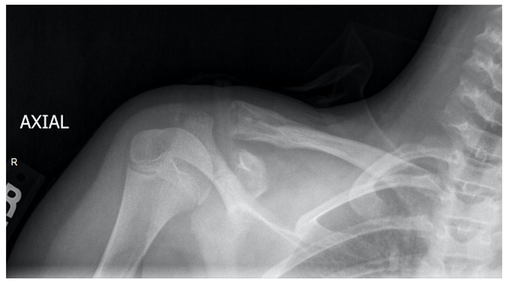
Differential Diagnosis

Tumor (osteosarcoma, osteochondroma, LCH, etc), Osteomyelitis, chronic recurrent multifocal osteomyelitis (CRMO), AC Sprain.

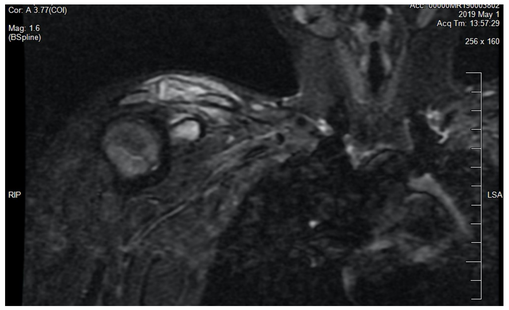
Imaging

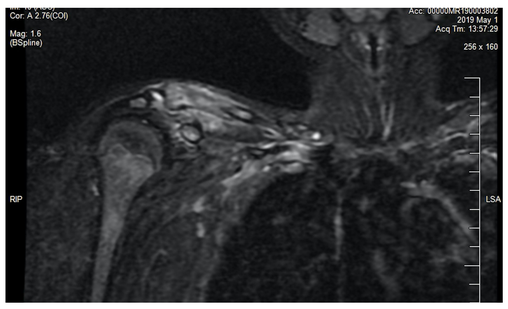
In-Office X-Ray Results



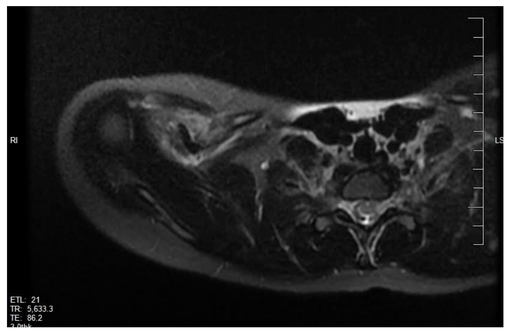


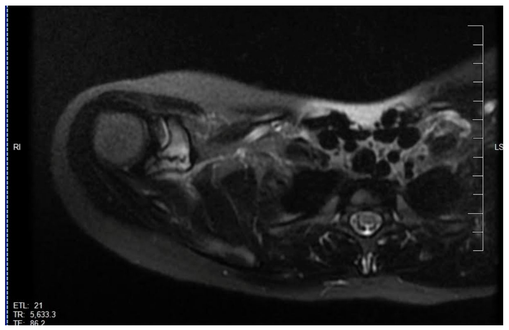
MRI Results



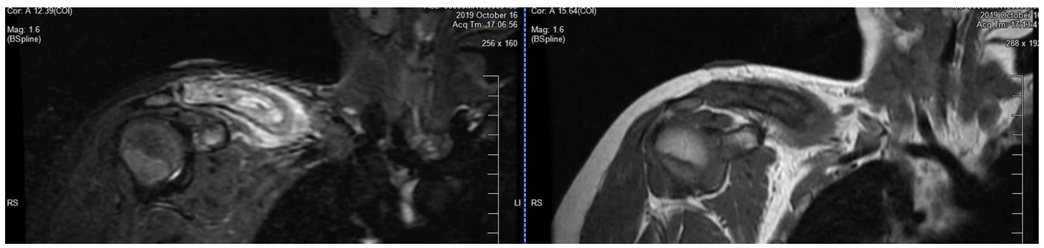


MRI revealed increased caliber of the right clavicle with expansion of clavicular marrow space as well as periosteal reaction. There was also noted to be surrounding soft tissue edema. Coronal Views (Above) Axial Views (Below).





Repeat MRI performed 6 months postoperatively (below) showed increased enhancement and edema with callus formation likely secondary to normal healing process.



Follow Up

Given the patient's presenting symptoms and exam in conjunction with in-office X-Ray results, the patient was expediently referred to orthopedic surgery with follow-up MRI ordered in advance of this visit. The MRI results were found to be suspicious for recurrent multifocal osteomyelitis versus LCH, and the patient was scheduled for biopsy with orthopedic surgery. Lesional biopsy was completed with in-op surgical pathology that was diagnostic for LCH. Frozen section demonstrated multiple histiocytes with a lack of nuclear atypia. The availability of in-operative surgical pathology evaluation facilitated prompt diagnosis and definitive treatment with bone curettage and grafting at the time of biopsy. Of note, the patient's biopsy and curettage with grafting occurred just 8 days after her initial clinic visit. Follow-up PET scan showed no signs of multifocal disease, and the patient was followed by hematology/oncology with enrollment in a surveillance-only study. The patient continues to undergo surveillance at this time.

Discussion

This case of LCH presenting as shoulder pain highlights the importance of considering uncommon causes of musculoskeletal pain, especially in the pediatric population. In multiple studies, it is noted that 96-97% of LCH patients with a solitary bone lesion experience disease free survival after definitive treatment. However, 20-65% of patients with multiple bone lesions or multisystem involvement experience recurrence or complications, further emphasizing the importance of prompt diagnosis.

In this case, the patient's presenting symptoms could have been underestimated and the work-up delayed. Instead, a thorough exam was performed and in-office X-Ray imaging was completed. Prompt follow-up imaging and referral allowed for the patient to be in surgery undergoing definitive diagnosis and treatment just 8 days after her initial presentation to the clinic. Timely radiographic imaging and referral can facilitate early detection and treatment of potentially malignant conditions.

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