JAMA Clinical Challenge

Radiographic Absence of the Left Humeral Head

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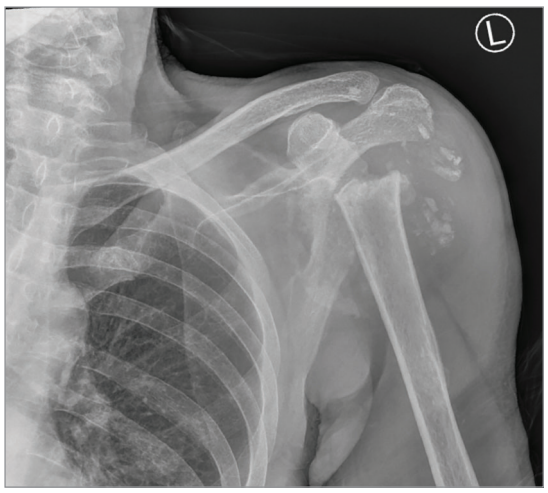

Figure 1. Anteroposterior radiograph of the patient's left shoulder demonstrating complete destruction of the proximal humeral head.

A 67-year-old man with thoracolumbar scoliosis, poor mobility, and history of frequent falls presented to the emergency department with 2 months of left shoulder pain, stiffness and reduced range of motion, and numbness and paresthesias in his left upper extremity. Ten years prior to presentation, he underwent surgical decompression for syringomyelia. A magnetic resonance imaging (MRI) scan of his cervical and thoracic spine performed 2 years prior to presentation revealed a recurrent syrinx extending from C1 to T11, which was not resected because it did not cause symptoms at that time. On physical examination, he had mild tenderness to palpation and reduced range of motion of the left shoulder with abduction and flexion limited to 120° (normal range of motion, 180°). The left scapular muscles were atrophic, and pain and temperature sensation were reduced in his proximal left arm, and dorsal aspect of his left shoulder. His complete blood cell count, serum glucose levels, C-reactive protein levels, and erythrocyte sedimentation rate were normal. Results of tests for rheumatoid factor and antinuclear antibody were negative. Left shoulder radiograph showed complete absence of the left humeral head and a well-demarcated smooth osseous margin of the proximal humerus with associated soft tissue swelling and periarticular calcification (**Figure 1**). A chest radiograph taken 2 years prior revealed a normal left shoulder joint. The patient was hospitalized for further evaluation and treatment.

WHAT WOULD YOU DO NEXT?

A. Obtain MRI of the left shoulder

B. Order serum tumor marker testing

C. Orthopedic surgery evaluation for shoulder replacement surgery

D. Perform an urgent left shoulder joint aspiration

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Diagnosis

Neuropathic arthropathy of the shoulder (Charcot shoulder)

What to Do Next

A. Obtain MRI of the left shoulder

The key to the correct diagnosis of neuropathic arthropathy of the shoulder is the radiographic finding of an absent left humeral head in a patient with a history of syringomyelia. Choices B and D are incorrect because the well-defined linear margins of the proximal humerus made septic arthritis or osteolysis due to malignancy unlikely. Shoulder replacement surgery (choice C) is not recommended for most patients with Charcot shoulder.

Discussion

Neuropathic arthropathy of the shoulder joint, also known as Charcot shoulder, is a rare progressive disorder characterized by rapid joint destruction and associated soft tissue swelling.[1,2] Patients typically present with gradually increasing shoulder swelling, which may or may not be painful, weakness in the shoulder, and decreased range of motion of the shoulder. Patients may report paresthesias or numbness in the affected upper extremity.[1,2] Approximately 80% of people with Charcot shoulder have syringomyelia, characterized by a fluid-filled cavity (syrinx) in the spinal cord.[2] Other conditions associated with Charcot shoulder include diabetes, syphilis, alcohol use disorder, Arnold-Chiari malformations, cervical spondylosis, amyloidosis,

end-stage kidney disease, myelodysplasia, multiple sclerosis, peripheral neuropathy, intra-articular steroid injection, tuberculosis, leprosy (Hansen disease), and gigantism.[2-4]

Syringomyelia may be congenital or secondary to trauma, infection, tumors, vascular abnormalities, or spinal degeneration.[4] As it expands, a syrinx damages spinothalamic tract fibers and may compress dorsal column and anterior horn cells, resulting in loss of sensation of pain and temperature, decreased muscle strength, muscle atrophy, and arreflexia.[5,6] Approximately 6% of patients with syringomyelia develop Charcot shoulder, and 80% have involvement of only 1 joint.[1,6]

The pathophysiology of neuropathic arthropathy, such as Charcot shoulder, is incompletely understood. A syrinx may decrease shoulder joint sensory innervation, leading to loss of somatic muscle reflexes that protect the joint from injury, resulting in recurrent subclinical trauma that causes inflammation and progressive shoulder joint destruction.[2,6] A syrinx may also cause loss of autonomic vascular control, increasing blood flow to the joint, and activating osteoclasts, which results in accelerated bone resorption.[1,7]

Patients with Charcot shoulder may have atrophic changes on radiographs, consisting of substantial bone resorption or shoulder joint destruction, and hypertrophic reactions, with osteophytes, sclerosis, and accumulation of osseous debris.[2] MRI of the shoulder can confirm the presence of Charcot shoulder and facilitates assessments for other associated conditions, such as rotator cuff tear.[2]

The diagnosis of Charcot shoulder should be made only after exclusion of other causes of joint destruction, including septic arthritis, osteolytic lesions from malignancy, rheumatoid arthritis, synovial chondromatosis, trauma, soft tissue sarcoma, and Milwaukee shoulder syndrome, a destructive arthropathy caused by deposition of hydroxyapatite crystals.[1,3]

Upon diagnosis of Charcot shoulder, the underlying cause should be identified and treated, in an attempt to slow or halt further joint destruction. Surgical resection of a syrinx in patients with Charcot shoulder due to syringomyelia may slow joint deterioration and improve neurologic symptoms.[2] Treatments for Charcot shoulder include nonsteroidal anti-inflammatory medications, joint aspiration if an effusion is present, patient education about avoidance of mechanical trauma, physical therapy, and rehabiliation.[1-3] Shoulder surgery is not typically recommended, but may be considered for a subset of patients with Charcot shoulder who do not improve with nonoperative management.[2,7]

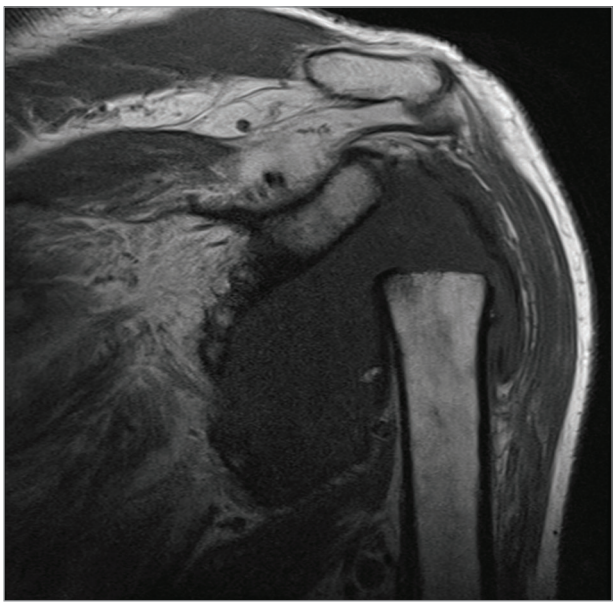

Figure 2. T1-weighted MRI of the left shoulder showing destructive osteolysis of the left proximal humeral head with clear linear margins and associated joint effusion.

Patient Outcome

The patient was treated with acetaminophen (paracetamol), topical nonsteroidal anti-inflammatory drugs, and short-acting opioids as needed. MRI showed destructive osteolysis of the left humeral head with well-defined, linearly demarcated margins of the proximal humerus with surrounding osseous debris and a moderate glenohumeral joint effusion (**Figure 2**). After consultation by clinicians in neurosurgery, orthopedic surgery, geriatrics, and radiology, the patient was advised to undergo intensive physical and occupational therapy. After 4 weeks of inpatient physical therapy 5 days per week, his left shoulder pain resolved, and left shoulder flexion and abduction was 160° to 170°. At his most recent outpatient clinic visit, 3 months after hospital discharge, the patient reported no left shoulder pain and had no change in shoulder range of motion. A follow up radiograph revealed a slight decrease in the left glenohumeral effusion.

ARTICLE INFORMATION

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Submissions: We encourage authors to submit papers for consideration as a JAMA Clinical Challenge. Please contact Dr Walter at kristin.walter@jamanetwork.org.

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