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Shoulder arthroplasty in the setting of polyostotic fibrous dysplasia

Andrew R. Jensen, MD, MBE [a], John W. Sperling, MD [b,*]

[a] Department of Orthopaedic Surgery, University of California, Los Angeles, CA, USA
[b] Department of Orthopedic Surgery, Mayo Clinic, Rochester, MN, USA

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Fibrous dysplasia is a benign intraosseous tumor in which marrow and normal cancellous bone are pathologically replaced with fibrous tissue, weakening the structural integrity of the bone.[1,3] Genetically, fibrous dysplasia is due to a mosaic mutation in G-protein alpha subunit that occurs during embryogenesis.[1,7]

Fibrous dysplasia is monostotic in 80% of cases, with the most commonly affected bones including the ribs, femur, tibia, mandible, skull, and humerus.[3] Fibrous dysplasia lesions in long bones typically affect the diaphysis or metaphysis.[8] In 20% of cases, the tumors are polyostotic. Patients with polyostotic disease and with lower extremity involvement are more likely to have pain and deformity.[10] Polyostotic disease can also be associated with McCune-Albright syndrome, which is a constellation of polyostotic fibrous dysplasia, precocious puberty, and café-au-lait spots, and Mazabraud's syndrome, which presents with polyostotic fibrous dysplasia and soft tissue myxomas, although both of these conditions are rare.[9] In addition, fibrous dysplasia, particularly the polyostotic form, may be associated with the presence of an aneurysmal bone cyst (ABC).[6]

Patients with fibrous dysplasia may present to the clinic due to a consequence of their fibrous dysplasia, such as with pain due to a pathologic fracture.[14] This is particularly common in the lower extremity, where patients can develop a varus deformity of the proximal femur known as a Shephard's Crook deformity.[8] A patient with new onset pain without a pathologic fracture should be evaluated for malignant transformation of the lesion, although this is fortunately rare.[13] Often, patients who present with fibrous dysplasia, be it due to a pathologic fracture or worsening pain, are treated with local curettage with or without internal fixation.[2,7,8,14]

Alternatively, patients with fibrous dysplasia may present to the clinic for workup of a separate pathology and be found to have fibrous dysplasia incidentally. This can occur in the work up of patients with end-stage arthritis, for instance, who are candidates for arthroplasty surgery. Because arthroplasty of the hip and knee have both historically been more common than shoulder arthroplasty, all of the literature to date on joint arthroplasty in the setting of fibrous dysplasia has been in the context of total hip or knee arthroplasty.[5,11,12,15,16]

Owing to the increased incidence of shoulder arthroplast utilization, however, we anticipate that more patients with fibrous dysplasia will be evaluated for shoulder arthroplasty in the coming years. These patients have unique clinical manifestations that the shoulder arthroplasty surgeon should be aware of. Therefore, we now present a case of a patient with rotator cuff arthropathy and polyostotic fibrous dysplasia to highlight the surgical challenges of shoulder arthroplasty in the setting of fibrous dysplasia.

Case report

Preoperative evaluation

A 75-year-old right-hand-dominant retired gentleman presented to the orthopedic clinic complaining of 2 years of progressively worsening left shoulder pain. He was unable to sleep or perform his activities of daily life without severe pain. He had failed an attempt at nonoperative management, including activity modifications, oral NSAID use, and an intraarticular corticosteroid injection.

The patient's past medical and surgical history was significant for a known diagnosis of nonsyndromic polyostotic fibrous dysplasia of his left upper and lower extremities. This was first

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* Corresponding author: John W. Sperling, MD, Mayo Clinic, 200 First Street SW, Rochester, MN, 55905, USA.
E-mail address: sperling.john@mayo.edu (J.W. Sperling).

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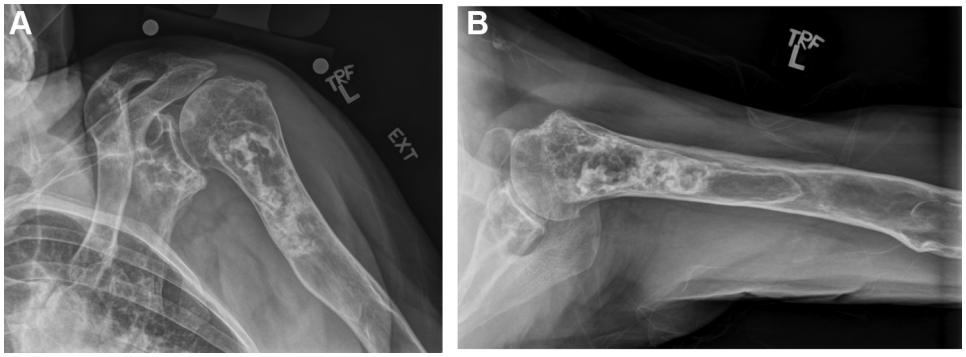

Figure 1 Preoperative Grashey AP (**A**) and axillary (**B**) radiographs demonstrated rotator cuff arthropathy as well as polyostotic fibrous dysplasia involving the clavicle, scapula, and proximal humerus.

diagnosed during his adolescence due to progressive left hip pain and deformity, for which he had multiple surgeries in his 20s. As is common in polyostotic fibrous dysplasia, his osseous lesions were contained to just one side; his right upper and lower extremities were spared.

Visual examination of his shoulder girdle demonstrated no skin abnormalities or notable muscle atrophy. He had 50° of active forward elevation, 90° of passive forward elevation, 20° of active external rotation (ER), and active internal rotation (IR) to the sacrum. He had crepitus and pain with passive glenohumeral motion. His strength was 4/5 in forward elevation and 3/5 in abduction, ER, and IR. There was no ER lag sign. Distally he was neurovascularly intact.

Plain radiographs of the left shoulder were obtained and demonstrated evidence of rotator cuff arthropathy, including glenohumeral joint space narrowing, inferior glenoid and humeral neck osteophytes, and proximal migration of the humerus with disruption of the Gothic arch (Fig. 1). In addition, there was widespread tumor involvement at the proximal humerus, scapula, and clavicle. The proximal humerus demonstrated endosteal scalloping with cortical thinning and intramedullary cortical rimming around the tumor.

An MRI was obtained to evaluate for possible extraosseous soft tissue tumor extension, which would be concerning for malignant transformation, and to assess his rotator cuff integrity (Fig. 2). There were no cortical breaches by the tumor. The MRI demonstrated a massive supraspinatus tear with retraction medial to the glenohumeral joint, severe muscle atrophy, and Goutallier grade 3 fatty degeneration. A CT scan was also performed for preoperative planning (Fig. 3).

The patient was diagnosed with left shoulder Hamada grade 2 rotator cuff arthropathy in the presence of fibrous dysplasia involving both the proximal humerus and glenoid vault. He was consented for reverse shoulder arthroplasty.

Surgery

After induction of general anesthesia, the patient was positioned in the beach chair position. Intravenous prophylactic antibiotics were administered, and the operative site was prepared and draped in the standard sterile fashion.

A deltopectoral approach was utilized. His skin incision was carried deep to the deltopectoral fascia, where the cephalic vein was identified and mobilized medially. The subscapularis tendon was identified and a tenotomy was performed. The humeral head was dislocated anteriorly and a standard humeral head cut was made.

At this point, a soft tissue lesion within the humeral metaphysis consistent with fibrous dysplasia was noted. There was no evidence of an ABC. Intramedullary tissue samples were obtained and sent to pathology, which were consistent with fibrous dysplasia and negative for a secondary malignancy. The humeral canal was then prepared in the standard fashion, using reamers and broaches for a short humeral component stem to avoid the sclerotic intramedullary cortical rims distally.

After preparation of the humerus, attention was turned to the glenoid, which was cleared of soft tissue and adequately visualized with appropriate retractor placement. Care was taken to ream the glenoid gently as to prevent fracture of the pathologically weakened bone. A standard baseplate was placed at the inferior aspect of the glenoid with a bicortical central screw and peripheral locking screws. A trial glenosphere was placed and attention was returned to the humerus.

Owing to the presence of fibrous dysplasia, a cemented humeral stem was used. After confirmation of appropriate stability and soft tissue balance with the trial components, the final components were inserted and the humeral stem was cemented in 30° of retroversion. After the cement had hardened, the final humeral liner was placed and the prosthesis was reduced.

The subscapularis tenotomy was repaired with interrupted absorbable sutures. The wound was thoroughly irrigated throughout the surgery and again at closure. The deltopectoral interval was reapproximated, the skin was closed, and a sterile dressing was applied. The patient was placed in a simple sling, awoken from anesthesia, and brought to the recovery room.

Postoperative course

The patient had an uneventful recovery in the post-anesthesia care unit and was transferred to the orthopedic ward. After achieving the appropriate milestones, he was discharged home on the morning of postoperative day 1.

He was advanced out of his sling at 6 weeks and gradually returned to his activities of daily living thereafter. At the patient's two-year clinic follow-up, he reported that he had no pain in his left shoulder, that he was now able to sleep comfortably, and that he had returned to playing racquet sports. On physical examination, the patient's range of motion had improved to 160° of active forward elevation, 40° of active ER, and active IR to L5. His strength was graded as 5/5 in all directions. The patient's subjective shoulder value increased from 10% preoperatively to 70% postoperatively, and his postoperative ASES shoulder score was 90. His postoperative radiographs demonstrated no evidence of glenoid or humeral component loosening (Fig. 4).

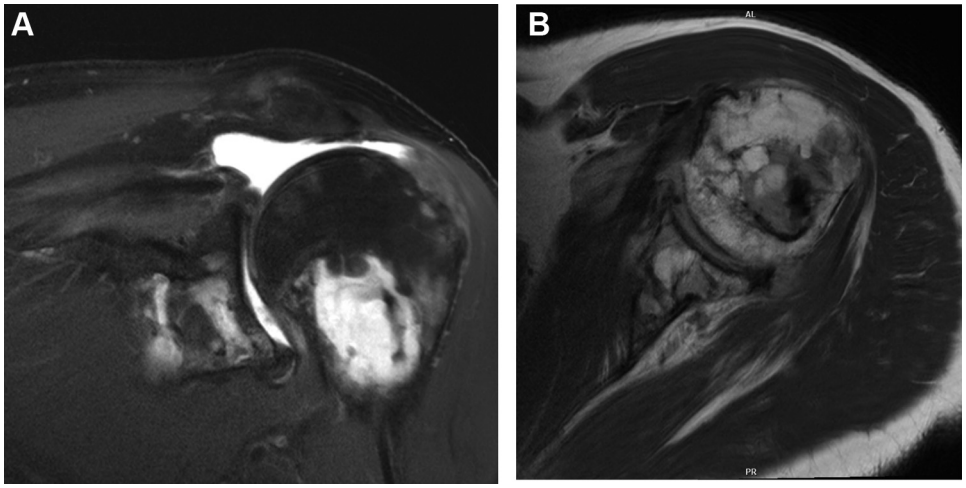

Figure 2 Representative images of a preoperative MRI. T2 coronal image (**A**) confirming the presence of a chronic, massive supraspinatus tendon tear with retraction, muscle atrophy, and fatty degeneration. T1 axial image (**B**) demonstrating fibrous dysplasia present in the proximal humeral metaphysis, the glenoid vault, and the coracoid. There were no cortical perforations or soft tissue extension of the fibrous dysplasia lesions to suggest malignant transformation.

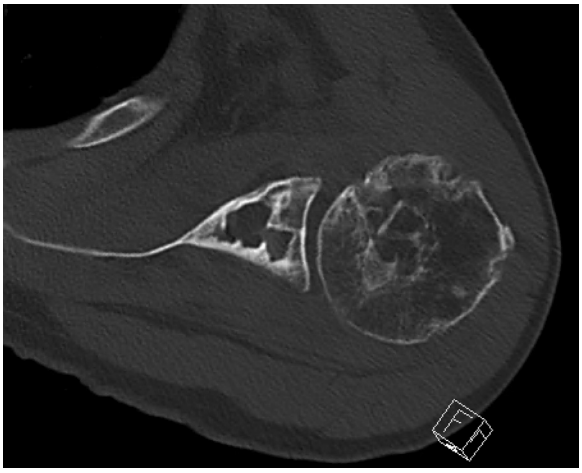

Figure 3 Preoperative axial CT image demonstrating fibrous dysplasia present in the humeral head and the glenoid vault.

Discussion

Although fibrous dysplasia is a rare entity and more commonly symptomatic in the lower extremities, it is likely that fibrous dysplasia lesions will increasingly complicate shoulder arthroplasty as the incidence of shoulder arthroplasty continues to rise. It is important for the shoulder arthroplasty surgeon to understand the clinical manifestations of fibrous dysplasia. The unique surgical challenges in shoulder arthroplasty posed by the presence of fibrous dysplasia include obtaining adequate humeral stem fixation, managing humeral deformity, obtaining adequate glenoid component fixation, avoiding intraoperative fracture, and being prepared for potentially significant blood loss (Table I).

The shoulder arthroplasty surgeon should be familiar with identifying fibrous dysplasia lesions during preoperative workup. On radiographs, fibrous dysplasia is notable for an intramedullary location with expansile endosteal scalloping and a ground-glass appearance.[3] Lesions can have a variable appearance on MRI but are often hypointense on T1-weighted images, and fluid-fluid levels can be suggestive of a coexisting ABC lesion.[3] Patients with fibrous dysplasia of the proximal humerus or glenoid may have had a previous pathologic fracture with resulting abnormal geometry, and previous surgical attempts at bone grafting often result in recurrence of the fibrous tissue. Malignant transformation, typically in the form of osteosarcoma, is rare but may present with cortical perforation and extension into the soft tissues.[13]

Although no prior study has reported the results of shoulder arthroplasty in the setting of fibrous dysplasia, there are a number of case studies published on hip and knee arthroplasty for patients with fibrous dysplasia from which we can glean relevant insights.[2,7,11,12,15,16] For example, Sierra et al reported on 12 total hip arthroplasties (THAs) performed in patients with fibrous dysplasia.[15] Of the 5 THAs in which cementless, press-fit femoral components were used, 3 required early revision for loosening and there was one intraoperative femur fracture. Comparatively, of the 7 THAs in which cemented femoral components were used, there were no revisions within 10 years.

Because of these findings and awareness of the underlying pathogenesis of fibrous dysplasia, we recommend using cement fixation for the humeral component. Pressfit humeral stems with grit-blasted or plasma spray coatings rely on native cancellous bone growth to obtain ongrowth stem fixation and implant stability. In patients with fibrous dysplasia, there is an intrinsic deficiency of the affected medullary bone to grow a normal architecture, and so the likelihood that successful ongrowth will occur to a sufficient degree to impart implant stability in these patients is low. Despite a lack of basic science or implant retrieval studies in patients with fibrous dysplasia evaluating the ability of affected bone to achieve successful ongrowth with pressfit arthroplasty stems, the high clinical rate of aseptic loosening of pressfit components seen in the THA literature would seem to indicate that cement fixation is the more reliable option.

In addition, fibrous dysplasia results in 2 related osseous abnormalities on the humeral side that can complicate shoulder arthroplasty. For 1, patients may have proximal humeral deformities, either due to metaphyseal expansion or previous pathologic fractures. These geometric abnormalities may require the use of a short stemmed or stemless humeral implant to avoid irregular cortical contours distally. Intramedullary cortical rims of bone can also complicate standard shoulder arthroplasty. These rims of bone form around the slow growing fibrous dysplasia lesions and can deflect medullary reamers during humeral preparation, leading to a cortical perforation. Implants

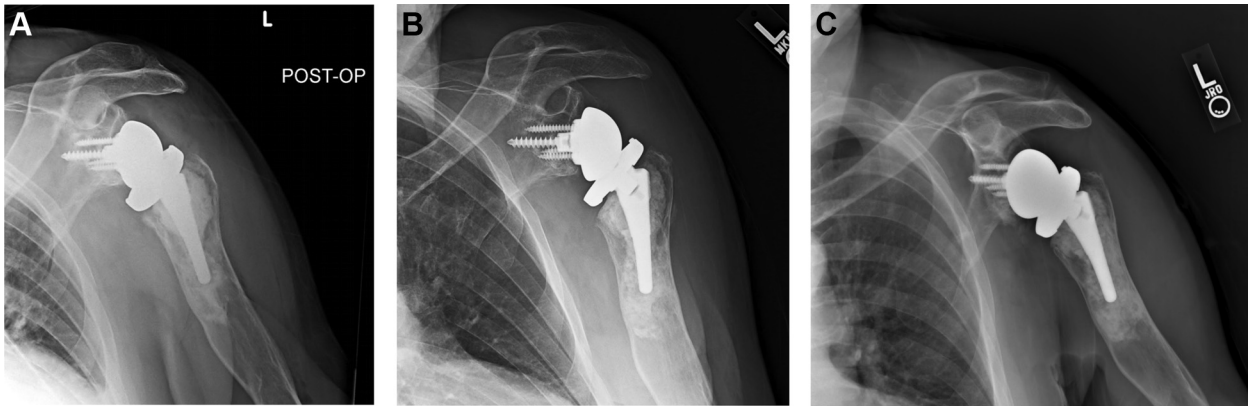

Figure 4 Immediate (**A**), six-week (**B**), and two-year (**C**) postoperative Grashey AP radiographs demonstrated excellent positioning of reverse shoulder prosthesis implant with no evidence of glenoid or humeral component loosening.

Table I
Unique surgical challenges for shoulder arthroplasty in patients with fibrous dysplasia.

Challenge	Description	Solutions
Obtaining adequate humeral stem fixation	• Compromised medullary bone growth prevents adequate ongrowth	• Use of a cemented humeral component
Managing humeral deformity	• Altered proximal humeral geometry prevents long stems • Intramedullary sclerotic rims of bone prevent safe distal reaming	• Use of short-stem implants to avoid distal cortical irregularities • If reaming is required, consider fluoroscopic guidance
Obtaining adequate glenoid component fixation	• Compromised medullary bone prevents adequate glenoid component fixation	• Removal of fibrous tissue from the glenoid vault • Cancellous bone grafting for small, nonstructural defects • Use of an augmented baseplate or corticocancellous bone graft for eccentric defects • Prepared to use revision fixation constructs as needed
Avoiding intraoperative fracture	• Thin cortical bone surrounding tumor increases the risk of humeral and/or glenoid intraoperative fractures	• Humeral stem cementation • Gentle glenoid reaming • Prepared to convert to hemiarthroplasty if unsalvageable glenoid fracture occurs
Preparing for potentially significant blood loss	• Aneurysmal bone cysts may be present in conjunction with fibrous dysplasia	• Prepared during osseous preparation for use of hemostatic agents, packing, and blood product transfusion • Perioperative use of tranexamic acid

that avoid these intramedullary cortical rims, such as the aforementioned short or stemless implants, can prevent this complication. If cortical rims are unavoidable during humeral preparation, we suggest the use of fluoroscopy for confirmation of intraosseous reamer and implant placement.

In patients with glenoid involvement, the first surgical step is to remove all fibrous tissue from the glenoid vault that would otherwise compromise glenoid component fixation. The resulting defect can then be treated in accordance with the glenoid bone defect classification system proposed by Gupta et al.[4] For central defects, without loss of cortical structural integrity, cancellous bone impaction can be utilized to fill the defect followed by implantation of a standard baseplate, although there is a risk of eventual fibrous transformation of the impacted bone graft. For eccentric defects, glenoid fixation options include use of an augmented glenoid baseplate or structural corticocancellous graft. In the rare circumstance of a completely unsalvageable remnant glenoid, the surgeon can consider revision techniques such as use of the alternate scapular line, of a two-staged glenoid reconstruction, of a glenoid vault reconstruction prosthesis, or of utilization of a hemiarthroplasty. For anatomic TSA, after removal of fibrous tissue within the glenoid vault, consideration should again be given to cancellous bone grafting for small, nonstructural defects before cementation of the glenoid component. However, if the cortical rim of the glenoid vault is violated or tenuously thin, we recommend implanting a reverse prosthesis with the aforementioned reconstructive techniques.

Finally, although neither occurred in the presented case, the shoulder arthroplasty surgeon should be aware of the increased possibility of intraoperative fracture or significant bleeding in fibrous dysplasia cases. Intraoperative fractures may occur due to pathologic thinning of the cortices from endosteal scalloping. With regard to the humerus, this is another reason to use a cemented implant in lieu of a press-fit one. Care should be taken in patients with glenoid fibrous dysplasia to avoid intraoperative fractures during glenoid preparation and reaming, and the surgeon should be prepared to convert to a hemiarthroplasty if an unsalvageable glenoid fracture were to occur. In addition to a heightened awareness of intraoperative fracture risk, the surgeon must also be prepared for potentially significant blood loss due to the presence of an ABC within a fibrous dysplasia tumor. This brisk bleeding could occur during humeral preparation, for instance, and may require hemostatic agents, packing, and the use of blood transfusion products.

Conclusion

Although rare, it is likely that more patients with fibrous dysplasia of the shoulder girdle will be indicated for shoulder arthroplasty as the incidence of shoulder arthroplasty continues to increase. Shoulder surgeons should be familiar with the clinical manifestations of fibrous dysplasia and the unique surgical challenges they present.

Conflicts of interest

None of the authors have anything to disclose regarding this article.

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Patient consent

Obtained.

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