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CASE REPORTS

Treatment of proximal humeral dysplasia epiphysealis hemimelica with custom hemiarthroplasty: a case report

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Dysplasia epiphysealis hemimelica is a rare disorder involving alterations in the growth of the epiphyses. It is more frequently encountered in the lower extremity, and shoulder girdle involvement is less common.[1,2] Diagnosis is usually made by radiographically identifying epiphyseal lesions of varying size that alter joint size and shape. Often, these lesions progress to limit joint motion and can lead to pain. Presented here is a patient in whom epiphysiodesis failed and who underwent shoulder hemiarthroplasty after humeral head collapse.

Case report

A 14-year-old boy was seen in our upper extremity clinic after being observed by another provider for his known diagnosis of dysplasia epiphysealis hemimelica since the initial radiographic diagnosis at the age of 3 years. At presentation, his radiographs showed a large, asymmetric growth of the proximal humerus and involvement of the distal clavicle. He had pain that localized to the clavicular lesion. At that time, motion was limited in rotation to neutral, but forward elevation (170°) and abduction (165°) were well preserved.

Because of the size of the humeral lesion, he underwent epiphysiodesis. His pain in the shoulder worsened over time, and at age 16, showed humeral head collapse after continued growth of the humeral epiphysis (Fig. 1), which was confirmed by computed tomography scanning. Furthermore, the glenoid was dysmorphic with shallow concavity and a small excrescence of bone in the center of the glenoid (Fig. 2). Function at that time demonstrated 50° of forward elevation and −20° of external rotation at the side.

The patient simultaneously developed elbow pain. Radiographs at age 16 demonstrated abnormal morphology of the radial head and a loose body proximal to the coronoid fossa (Fig. 3). Flexion was to 95°, and he lacked 10° of full extension. Pronation and supination were normal, at 90° each.

Ultimately, his shoulder pain worsened and he elected to undergo hemiarthroplasty. A custom implant was created because the humeral head measured 80 mm in diameter at its largest dimension on the computed tomography scan. Given the massive size of the humeral head, coupled with the shallow glenoid concavity, it was felt that standard implants would not have provided appropriate stability because of expected capsular laxity after resection of the massive humeral head. A 56-mm head with variable offsets and a custom lockable implant stem were created. The proximal humerus had a significant stenosis of the canal at the humeral head/neck junction that would have precluded traditional implant fixation with circumferential bone−implant contact. To obtain adequate stability with a short, ingrowth stem, locking was selected.

Resection of the massive humeral head showed cartilage throughout the entirety of the humeral metaphysis. The rotator cuff was thin but intact. The supraspinatus was tightly compressed between the acromion and the enlarged humeral head. The glenoid was smaller and shallower than usual, and a small area of convex bone, approximately 1 mm × 2 mm, was curetted to create a concave surface for articulation consistent with preoperative imaging. Excepting this small area, the rest of the glenoid surface was covered with grossly normal articular cartilage, so neither reaming nor interposition arthroplasty were performed.

The humeral capsule was extremely tight. This required release of the anterior and inferior capsule off the humeral neck from where it had been invaginated into the space between the enlarged humeral head and the humeral shaft. Additional release of the inferior capsule from the neck of the glenoid was also required to

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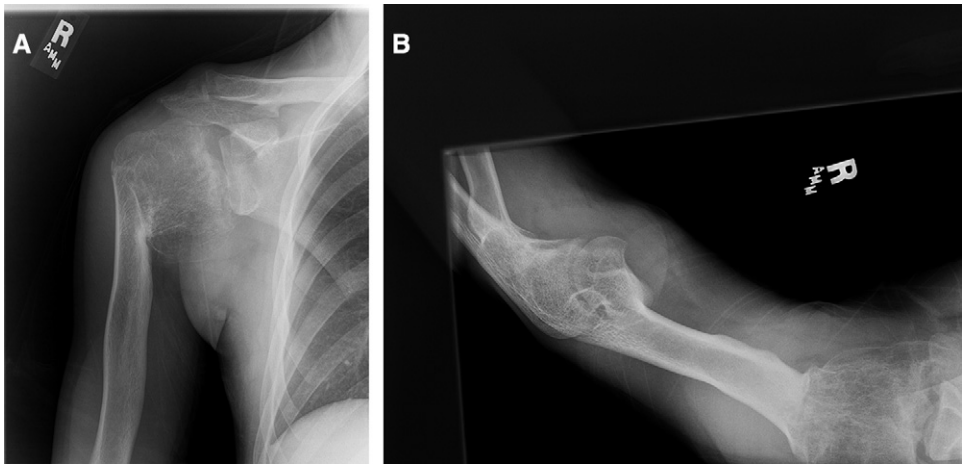

Figure 1 (**A**) Anteroposterior and (**B**) lateral preoperative images.

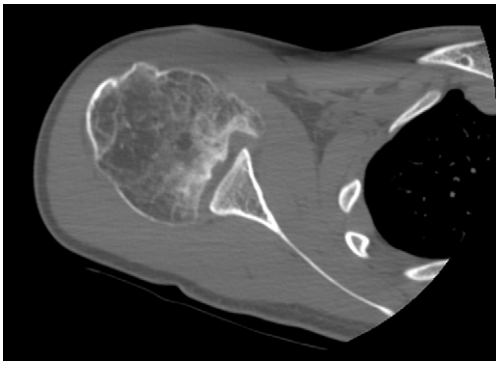

Figure 2 A computed tomography scan shows the dysmorphic glenoid articular surface.

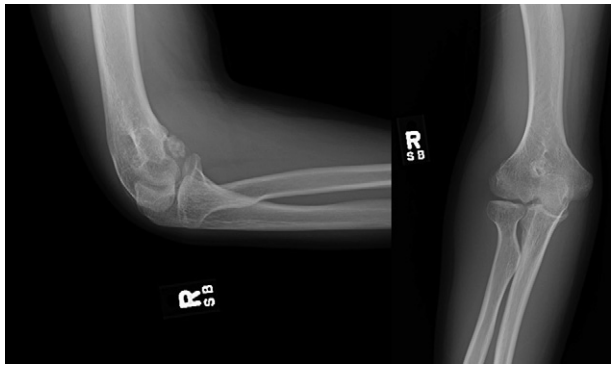

Figure 3 (*Left*) Lateral and (*Right*) anteroposterior images of the elbow.

properly expose the humerus for amputation and reaming of the canal. The preponderance of the posterior capsule was preserved to reduce the risk of posterior instability.

Postoperatively, the patient had substantial reduction of his pain, but return of motion was slow. At age 18 (20 months after hemiarthroplasty), shoulder motion was improved but limited compared with the contralateral side, with 120° of forward elevation (including hyperlordosis) and 30° of external rotation. Forward elevation with scapular stabilization was limited to 80°, however, demonstrating that some of the improvement was from scapulothoracic motion and hyperlordosis of the lumbar spine. Shoulder imaging at that time showed stable position of the custom implant, without glenoid wear (Fig. 4) In addition, the patient's elbow pain had improved substantially, despite no change in motion. Radiographs also remained unchanged.

Discussion

Dysplasia epiphysealis hemimelica is an infrequent lesion involving the epiphysis. Few cases involve the upper extremity and even fewer the shoulder girdle. This patient also had involvement of 3 different joints in the same extremity (acromioclavicular, glenohumeral, and elbow). In this case, the humeral epiphysis progressively enlarged, limiting function and leading to increasing pain. The epiphysiodesis procedure did not slow humeral growth adequately and ultimately led to humeral head collapse. The surrounding tissues at surgery were also abnormally contracted, and motion in the shoulder demonstrated this. The course of nonsurgical and surgical management is described.

Conclusion

The literature contains few examples of patients with epiphyseal growth to the point of collapse requiring arthroplasty. In this unique case, hemiarthroplasty was required because of collapse of the humerus around the glenoid. A custom implant was used to compensate for the massive size of the humeral head and to allow

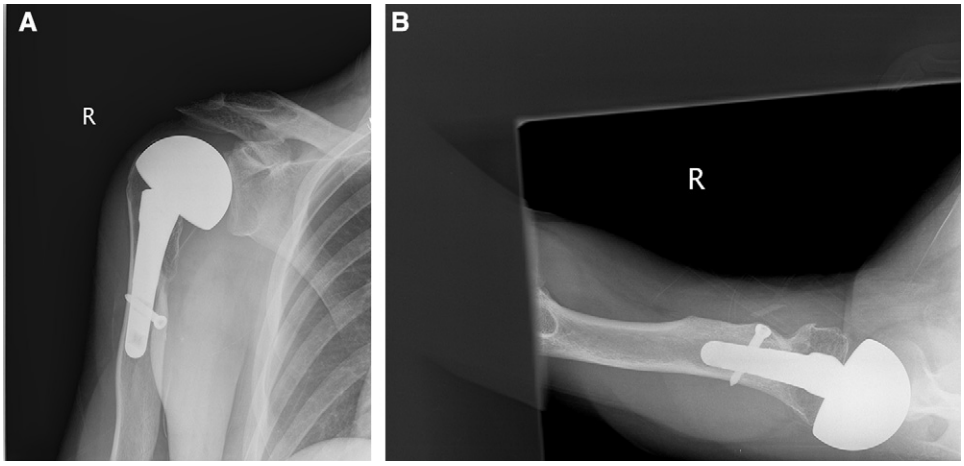

Figure 4 Postoperative (**A**) anteroposterior and (**B**) lateral images show the humeral head replacement.

ingrowth with the complex shape of the proximal humerus. Fortunately, this patient has demonstrated improvement in his motion and pain with surgical treatment. This appears to be a reasonable method to salvage this difficult and challenging problem when nonsurgical management has failed to provide relief.

Disclaimer

The authors, their immediate families, and any research foundations with which they are affiliated have not received any financial payments or other benefits from any commercial entity related to the subject of this article.

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