Introduction

Chondroblastoma, first described in 1927 by Kolodny and further detailed by Codman in 1931 and Lichtenstein and Jaffer more recently, is a benign ephiphyseal/apophyseal tumor primarily affecting the long bones.1-2 It consists of 1-2% of all primary bone tumors and 5% of benign bone tumors.3-5 The male to female ratio is 2:1.3-8 The greater majority of patients are less than 20 years old.9,10 The bones most affected are the femur and humerus.5-10 While less common, chondroblastoma tumors can be found in the tibia, foot, flat bones, and vertebrae.11,12

One single, specific cytogenetic abnormality that can lead to chondroblastoma has yet to be described. However, research looking at cartilage signal molecules is proving promising. Studies have shown the tumor to be of mesenchymal cell origin, dedicated to chondrogenic life cycle via signals from active growth plate pathways, thus verifying the intricate relationship of chondroblastoma to the epiphysis.14,15

Grossly, chondroblastoma is a gray-white tumor with yellowish areas of calcification.2 Microscopically, it has compact areas of chondroblasts with 1-2 nuclei, few mitotic figures, and the occasional multinucleated osteoclast like giant cells.6 The textbook finding of dystrophic calcification "chicken wire" may also be present. Nearly one third of cases have secondary aneurysmal bone cysts.6-8,10 Aggressive and metastatic tumors have been shown to have the same histology as indolent and solitary tumors.16

Chondroblastomas are classified according to the Enneking benign tumor classification.23 Clinical presentation is usually insidious in nature with progression of pain from mild to significant both locally and in the adjacent joint.3,4,6,8-10 Patients can present with a soft tissue mass or pathological fracture though this is uncommon with highest incidence of this in tumors of the foot. They usually have decreased range of motion and muscle wasting.4,6,8-11

Radiographically they are well-defined, eccentric oval or round lytic lesions of the epiphysis adjacent to an open growth plate.3 Margins are generally sclerotic and sharp, sometimes having intralesional mottling with or without calcification. Most tumors are less than 4cm in size.2,4,6,9 It is common to find surrounding soft tissue and marrow edema, but rare to find periosteal bone formation.17 While some ephiphyseal lesions will cross the physis and invade the metaphyseal bone, true metaphyseal and diaphyseal chondroblastomas are rare but have been described.14,18 CT scan is ideal for identifying the tumors anatomic limits including distance from the growth plate.11,12 Chondroblastoma is hypointense on T1 MRI and variable hypo-to hyperintense on T2. MRI is ideal for evaluating the status of surrounding soft tissue and the extent of marrow edema.17

The natural history of chondroblastoma has shown that it does not spontaneously resolve and has not been shown to respond to medical management.3 The standard of care at this point is intralesional excision via curettage and bone graft with care to avoid the growth plate in skeletally immature individuals.6-12 Excision augmented with high speed burr, electrocautery, phenol, argon beam, and cryotherapy are all described appropriate treatments.19 Radiofrequency has been shown to have success with small tumors (<2 cm) that are away from the joint.20 For aggressive tumors and recurrences, limb-sparing surgery and endoprosthetic reconstruction are now options.4,10 Due to late recurrence and possibility of lung metastases, most recommend an annual follow up for at least 5 years with chest x-rays at each annual visit.7

The most common complication for chondroblastoma is recurrence, with studies reporting a 5-40% recurrence rate. However, the rate of recurrence is not related to one specific mode of management, tumor size, patient sex, or duration of follow-up.6,8,9 The most common reason for recurrence is incomplete resection and biologic aggressiveness.7 Less than 1% of chondroblastomas metastasize. There is no reported relationship of metastatic lesions to previous surgical or nonsurgical treatment, tumor location, or patient age.16,21,22 The average time to metastasis is 8 years.16 Common locations are the lungs, other bones, soft tissue, skin, and liver.16,21 Patients with metastatic chondroblastoma continue to have long survival rates and no benefit from chemotherapy.

Chondroblastoma of the Humeral Epiphysis in a 15-Year-Old Female: A Case Report

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Case

EG is a 15-year-old female, right hand dominant, who presented to the pediatric upper extremity clinic in August of 2013 with a chief complaint of eight months of left shoulder pain that was constant and exacerbated with use. Without a history of trauma, her pain was anterior, worse with overhead activity, decreased range of motion and significant muscle atrophy. She had mild relief with anti-inflammatory medication. She presented with a diagnosis of rotator cuff tear. Past medical, surgical, birth, and developmental history were all normal. There is no family history of cancer.

On physical exam, she was found to have pain with range of motion and tenderness to palpation at the left proximal humerus without a palpable mass. Pain significantly limited her range of motion with forward flexion and abduction at the left shoulder less than 90 degrees actively and passively. She had external rotation to neutral. She had noted atrophy of the left shoulder, arm, and forearm with decreased strength.

Radiographs of the left shoulder and humerus revealed a well-circumscribed lucent lesion within the proximal humeral epiphysis (Figure 1). There was no cortical disruption noted, but the lesion did extend very near the glenohumeral articular surface. A 3.0 Tesla MRI revealed a 3.5cm x 3.1cm, x 2.8cm heterogeneous, well-circumscribed lesion of the epiphysis that invaded the open physis and extended into the metaphyseal region. There was notable marrow and soft tissue edema without axillary lymphadenopathy and a moderate joint effusion (Figures 2 & 3). CT of the chest was normal without metastatic lesions to the lungs. Full body bone scan was negative for multifocal disease.

One week after presentation, the patient had a CT-guided core needle biopsy performed by interventional radiology (IR). Pathology findings were consistent with chondroblastoma. Three weeks after her initial presentation, she underwent cryoablation. She experienced temporary partial relief of her pain from the cryoablation but her symptoms returned to baseline level in two months.

The patient subsequently underwent two cycles of radiofrequency ablation three months after her initial presentation. She again experienced minimal relief and was taking maximum doses of anti-inflammatory medications.

She was re-evaluated four months after the biopsy and repeat x-rays, CT, and MRI showed mildly increased marginal sclerosis as a result of the ablations but the tumor had increased in size now measuring 3.6cm x 3.5cm x 3.2cm and was very near the articular surface with more reactive soft tissue and marrow edema. There were also axillary reactive lymph nodes present.

On month 7 from presentation, she underwent intralesional curettage and grafting of the proximal humerus with bone graft substitute of calcium phosphate and calcium sulfate. The deltopectoral approach was used and an anterior cortical window was created. Curettage was done using a high-speed burr with hydrogen peroxide for irrigation. Meticulous care was used to avoid the articular surface. (Figure 4)

Intraoperative pathology results show no cytogenetic abnormalities on genotyping with classic microscopic features of chondroblastoma (including molecular marker S-100 and DOG-1 +) in addition to hypocellular fibrosis.
and reactive and new-bone osteoid matrix. Her histology showed areas of necrosis from the cryoablation and pathological multinucleated cells. (Figure 5)

At her most recent follow-up two weeks post-operative, she had significant decrease from her baseline level of pain. She was showing increased strength with good effort. She had the same limited range of motion but minimal pain through the range.

Discussion

This patient represents a typical presentation of chondroblastoma. It was explained to the patient and her family at the time of presentation that intralesional curettage and bone graft would be the definitive treatment. A big concern for the patient was finishing out her school year before undergoing surgery and getting her pain controlled. Of concern to the surgeons was the proximity of the tumor to the articular surface of the humerus. Thus, in an effort to help sustain the articular surface, decrease the patient’s pain, and to hopefully buy her some time until the school year was completed, cryoablation was done. Given the patient had some relief but her pain returned, radiofrequency ablation was attempted. Repeat imaging after both procedures were completed showed the tumor had increased in size. These interventions were not as successful as we had hoped, but the team was aware prior to the intervention that studies weren’t reporting much success with tumors the size of the patient’s in terms of decreasing size or symptom relief. The post-operative pathology showing hypocellular fibrosis indicates, however, that there were changes to the tumor as a result of the ablations.

In an effort to maintain the articular surface of the proximal humerus, the most superior aspect of the tumor was not entirely excised and this is evident on post-operative x-rays. This potentially does place the patient at increased risk for recurrence. The amount of symptom relief the patient experienced is promising. She will be followed closely in the post-operative period and then annually for at least 5 years. A follow-up case report is planned, particularly looking at recurrence, residual shoulder dysfunction, and maintenance of the glenohumeral joint surface.

References


