A Rare Case of Chondromyxoid Fibroma of the Scapula

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Chondromyxoid fibroma (CMF) is a rare benign tumor, apparently derived from cartilage-forming connective tissue. The name is highly descriptive of this distinctive tumor and has gained acceptance.[1] The entity was first described in 1948 by Jaffe and Lichtenstein,[2] who presented 8 cases and emphasized the danger of mistaking this benign neoplasm for a malignant lesion, chondrosarcoma in particular. Approximately two thirds of the recorded cases of this tumor have been in the long tubular bones and one third in the proximal tibia.[1,3,4] A scapular origin of this tumor is exceedingly rare.[1,5-10]

We report the case of a 13-year-old girl with chondromyxoid fibroma of the scapula. This case is of interest because of the rarity and unusual location of the tumor. The authors have obtained the patient's guardian's written informed consent for print and electronic publication of the case report.

Case Report

A 13-year-old girl presented for evaluation of a lesion in the right scapula. The lesion was noted on magnetic resonance imaging (MRI) of the shoulder, which had been injured in gym class. By the time of presentation, however, the pain had completely resolved. On physical examination, there were no overlying skin changes, no visible masses, and no palpable tenderness in the right shoulder, and range of motion was full and painless. The neurovascular status of the involved extremity was intact, and there was no lymphadenopathy. Laboratory tests, including complete blood cell count, serum chemistries, and erythrocyte sedimentation rate, were all within normal limits. The patient denied fever, chills, weight loss, and systemic symptoms. Her medical, surgical, and family histories were unremarkable.

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Plain radiography (Figures 1A, 1B) and computed tomography (CT) scan (Figure 2) revealed an expansile lesion of the right scapula with central calcification suggesting chondroid-type matrix. There was some thinning of the cortex but no obvious cortical breach or associated soft-tissue mass. MRI (Figure 3) revealed a 5×3×2.5-cm expansile lesion involving the inferior border of the scapula. T_2-weighted images showed a heterogeneous mass with bright signal intensity. There was considerable edema in the teres minor and subscapularis muscle bellies. No fluid–fluid levels were seen. Additional workup included a chest CT scan and a whole-body bone scan. The bone scan revealed increased focal uptake to the right scapula with central photopenia. There was no evidence of disease elsewhere.

After evaluation of imaging studies, we considered a wide array of differentials, ranging from unicameral bone cyst and benign fibrous lesion to locally aggressive lesions, such as aneurysmal bone cyst (ABC) and chondroblastoma. Surgical biopsy was indicated to determine the nature of the lesion, which on microscopic examination was seen to be a highly cellular lesion made up of polygonal cells with indented nuclei and areas of spindle and stellate cells. Rare foci of a myxoid matrix and calcifications were seen. Both chondroblastoma and CMF were included in the differential diagnosis. Once the final tissue diagnosis was available, the definitive surgical procedure was performed as a separate procedure, subtotal scapulec-

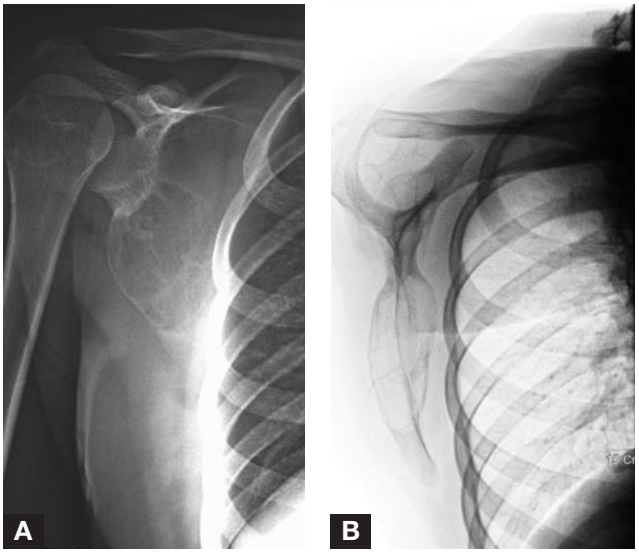

Figure 1. Plain anteroposterior (A) and scapular (B) radiographs show expansile lesion of scapula.

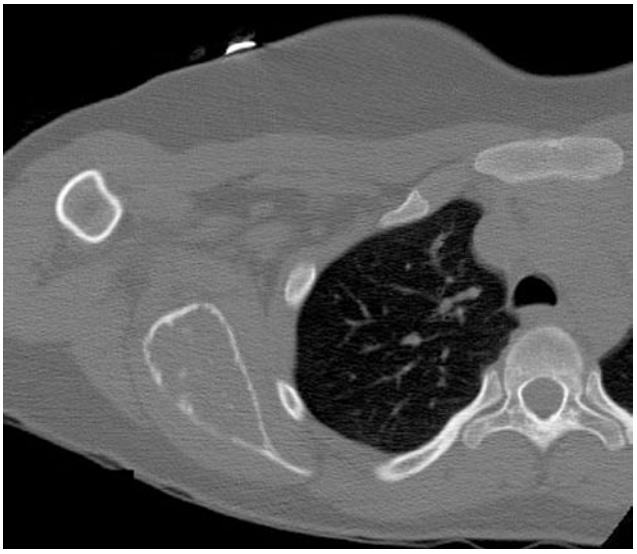

Figure 2. Axial computed tomography scan of shoulder shows expansile lesion of scapula with foci of calcification suggesting chondroid-type matrix.

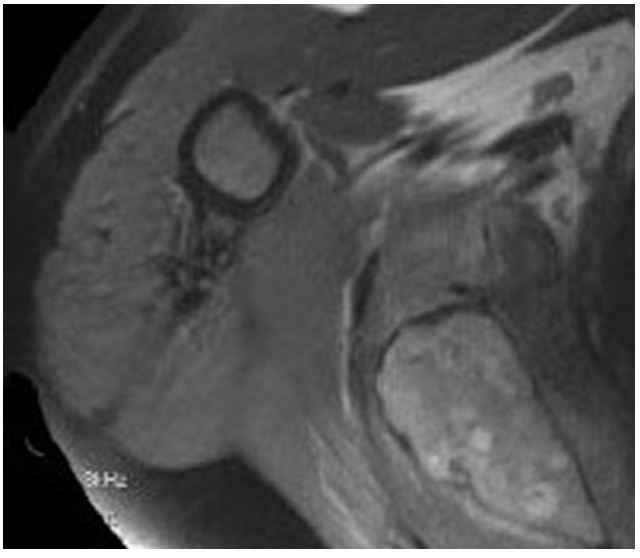

Figure 3. T_2-weighted axial magnetic resonance imaging scan of shoulder shows 5×3×2.5-cm expansile lesion of scapula with heterogenous signal.

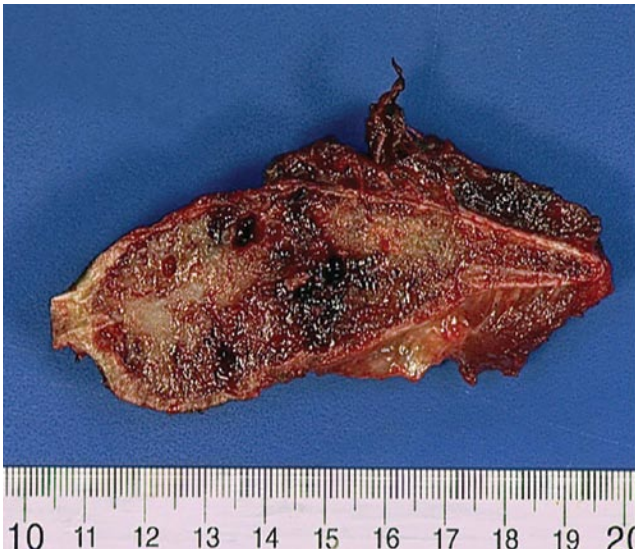

Figure 4. Gross photograph of resected scapula shows replacement of marrow with light tan tumor having a central area of hemorrhagic necrosis.

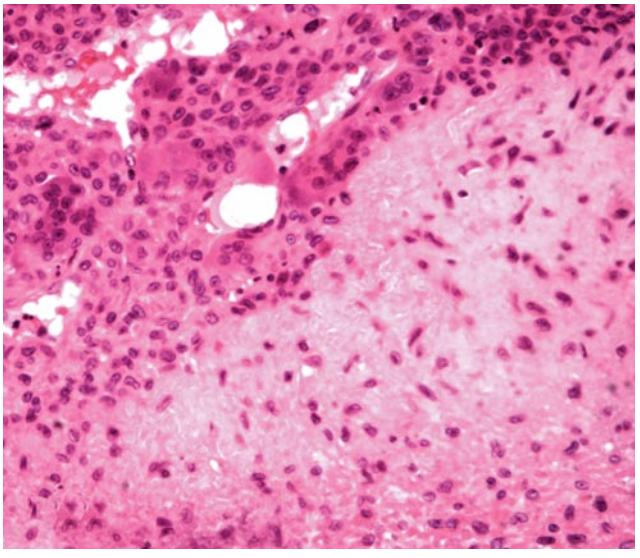

Figure 5. Cellular components made up of polygonal cells, stellate cells, and giant cells (hematoxylin-eosin).

tomy. Resection was carried out in such a way to preserve the glenoid and the glenoid neck to maintain stability of the shoulder joint. Intraoperative pathology consultation indicated negative margins.

Gross examination revealed a lobulated, scapula-expanding neoplasm with a grayish white and flesh-colored surface (Figure 4). Microscopically, the tumor showed lobulation and a myxoid matrix surrounded by high cellular areas containing polygonal and stellate cells (Figure 5). Foci of blood-filled lakes comprised secondary ABC formation. Intratumoral calcification was present in rare foci. Given this histologic appearance, a definitive diagnosis of CMF was made. Cytogenetic analysis and karyotyping of a portion of the tumor revealed rearrangement of chromosome 6q25.

Six months after resection, there was no evidence of recurrence. Range of motion at the right shoulder was normal. Radiographic evaluation at most recent follow-up showed well-maintained glenohumeral articulation with an area of heterotropic ossification (Figure 6).

Discussion

CMF is a cartilaginous tumor of bone with varying amounts of myxoid and fibrous tissue elements, with giant cells in some cases and occasional cellular atypia. Data indicate the benign nature of the lesion, despite an occasional ominous histologic appearance.[10-12] This tumor accounts for only 2% of benign bone tumors and less than 1% of all primary bone tumors.[1,3,4,9,13,14] CMF has a marked predilection for patients

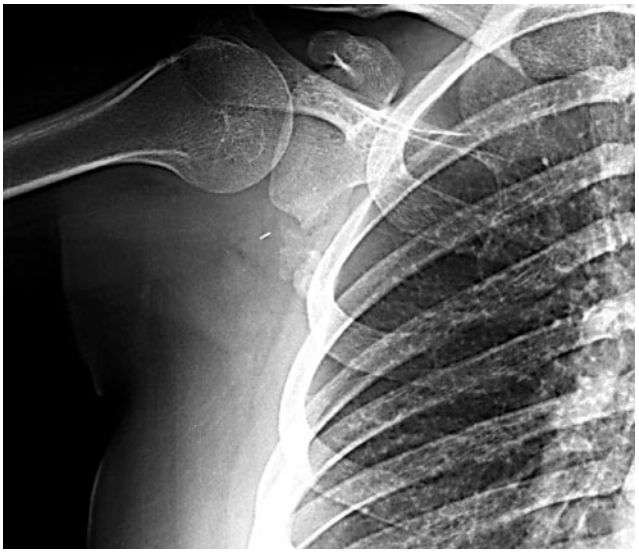

Figure 6. Plain anteroposterior radiograph at most recent follow-up shows well-maintained glenohumeral articulation and an area of heterotropic ossification inferior to glenoid.

in the second and third decades of life.[2,8,15] Most series have demonstrated a definite male predilection.[1,3,4,15]

Few patients with this tumor have no symptoms, but most present with pain of variable duration (2 weeks–20 years). Local swelling may be noticed by a patient whose tumor is not camouflaged by a thick layer of overlying tissue. Occasionally, these tumors are asymptomatic, incidental findings on radiographs.[2,4,8]

The most common site of this tumor is the metaphysis adjacent to the epiphyseal growth plate of long tubular bones, particularly around the knee joint. This finding is consistent with the hypothesis that the tumor arises from remnants of cartilage at this site.[15,16] As mentioned, the scapula is a very rare site, accounting for 0.8% to 3% of all CMF cases.[7]

Radiographic Features. Typical radiographic features of CMF are a single, lobular, eccentric, radiolucent lesion with expansion of the affected bone.[16] The endosteal margin is usually well demarcated, with a sclerotic rim. Although the tumor is benign, the aforementioned radiographic features confirm potentially aggressive behavior. Radiographic evidence of calcification is rare in CMF.[1,7] Of the 191 tumors evaluated by Wu and colleagues,[17] 87% had a purely lucent matrix. Calcified CMFs are reported more commonly in unusual locations, particularly flat bones.[17,18]

Treatment. Treatment of CMF has changed since this entity was recognized in 1948. Wide resection of the tumor is a treatment option. Curettage, though ordinarily successful, is associated with a 12.5% to 25% risk for recurrence. Bone grafting is often necessary after curettage. It has been suggested that bone grafting reduces risk for recurrence. Heydemann and colleagues[22] attributed soft-tissue recurrence of CMF after curettage to spillage of tumor cells during the index procedure—further emphasizing the aggressive nature of the tumor. Fotiadis and colleagues[16] advocated use of polymethylmethacrylate after curettage, as it provides structural strength and extends the margin of tumor cavity by exothermal reaction. Radiation therapy is not indicated, except for the very rare surgically inaccessible lesion.[1,7,13] Age seems to have a significant influence on recurrence rate. In younger patients, the tumor has been found to have mostly myxoid areas and large nuclei. Lowered resistance offered by the thin cortex and spongiosa of young bone has been reported to contribute to the more aggressive nature of the lesion. These cases are most likely to develop recurrences after curettage.[3,13] Few authors have described CMF as having undergone malignant transformation (overall incidence, 1%-2%).[15] Some cases of the malignant behavior of this tumor may be attributed to erroneous classification of low-grade chondrosarcoma as CMF.[16]

The Differential Diagnosis

When a clinicopathologic diagnosis of CMF of the scapula is being considered, the differential diagnosis should include chondroblastoma, osteochondroma, ABC, and chondrosarcoma. Presence of the distinctive "chondroblasts," osteoclast-like giant cells, "chicken wire" calcifications, and lack of lobulations are characteristic features of chondroblastoma. In addition, the chondroblastoma is an epiphyseal lesion, whereas CMF is typically metaphyseally centered. Osteochondroma is the most common primary benign neoplasm of the scapula, with an incidence of 4.6%. Patients with osteochondroma usually present with the complaint of a snapping sensation with movement of the shoulder. ABCs are radiolucent expansile lesions. Fluid–fluid levels are seen with MRI. Microscopically, ABCs are characterized by hemorrhagic tissue with cavernous spaces separated by cellular stroma. It must be noted that lesions such as giant cell tumor, chondroblastoma, fibrous dysplasia, and nonossifying fibroma may have ABC as a secondary component. Chondrosarcoma is the most common primary malignant neoplasm of the scapula. Radiographs may show cortical destruction by a lesion containing a calcified cartilaginous matrix. Microscopically, most chondrosarcomas produce hyaline cartilage. Presence of mitoses and nuclear anaplasia differentiates chondrosarcoma from CMF. Engels and colleagues[19] concluded that, in most cases, adherence to strict histomorphologic criteria leads to a definitive diagnosis of CMF. Other less common differential diagnoses are transformed chondrosarcoma, mesenchymal chondrosarcoma, and a CMF-like variant of low-grade osteosarcoma.[15,19-21]

Prior Reports of Scapular CMF. In our review of the literature, we found only 4 reports of CMF of the scapula. Prichard and colleagues[8] reported a case of CMF of the scapula in a 38-year-old man in 1964 (their case report can be considered the first on this location of this tumor). Uematsu and colleagues[23] reported a case of "malignant" CMF in a 39-year-old woman based on histologic characteristics and tumor invasion to the skeletal muscles. Baklouti and colleagues[5] noticed, in a 23-year-old woman, extensive invasion of the soft tissues—a feature seldom observed in CMF. Mizuno and colleagues[7] reported ABC as a secondary component to CMF and stated that, though uncommon, CMF should be considered in the differential diagnosis of a radiolucent, expansile lesion of the scapula in a young patient.

Summary

CMF is an uncommon benign bone tumor of cartilaginous origin. Its occurrence in the scapula is rare and poses a diagnostic and therapeutic challenge. Considering current information, we recommend that CMF of the scapula be treated with wide surgical excision when possible and that the patient be regularly followed up for recurrence.

Authors' Disclosure Statement

The authors report no actual or potential conflict of interest in relation to this article.

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This paper will be judged for the Resident Writer's Award.