Case report

A case report on partial scapulectomy with glenoid preservation for Chondromyxoid fibroma of scapula

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ABSTRACT

Chondromyxoid fibroma is a benign bone tumour accounting for less than 1% of all primary bone tumours. It usually affects the metaphyseal region of long bones in the first or second decade of life. It rarely occurs in scapula. We present a case of 29 year old female with biopsy proven Chondromyxoid fibroma of left scapula. She underwent wide marginal excision by partial scapulectomy with preservation of glenoid. Post operatively she has stable shoulder joint with normal range of movement & no recurrence on regular follow up.

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1. Introduction

Chondromyxoid fibroma is a benign but aggressive bone tumor¹ accounting for less than 1% of all primary bone tumours and only 2% of benign bone tumours^{2–7}. It usually affects metaphysis adjacent to the epiphyseal growth plate of long bones, particularly around the knee joint^{8,9}. Scapula is a very rare site^{2,10–12}, accounting for 0.8% to 3% of all cases of chondromyxoid fibroma¹³. Although it primarily occurs in the second and third decades of life^{8,14,15} yet a second peak in the fifth, sixth and seventh decades is observed^{1,3,16}. As curettage with or without local adjuvant shows a recurrence rate of approximately 25%, the preferred treatment is wide excision. Radiotherapy may be useful in nonresectable cases but bears the risk of radiation induced malignancies¹⁷.

2. Case report

A 29 years old female presented with a painless swelling over left scapular area which was noticed about two years back. It was small in size at the time of first detection and increased in size progressively. She had no difficulty in movement of her left shoulder and could perform all activities of daily living with her left

upper limb. She had no history of trauma, loss of body weight or appetite, fever or any other systemic features.

Examination revealed an oval shaped swelling of about 12 × 8 cm in greatest dimension of length and width respectively arising out of left scapula (Fig. 1). The overlying skin was normal looking and could be lifted off the lesion all throughout. Surface was smooth and margin was well defined. It was firm, non-tender and fixed to left scapula and therefore could not be moved in any direction. No localized signs of inflammation, discharge, ulcer, sinus or fistula was noticed. There was no regional or generalized lymphadenopathy and distal neurovascular deficit. General and systemic examination did not reveal any abnormal findings.

Her radiograph showed an eccentric radiolucent expansile lesion arising from supraspinatus part of left scapula with distended, thinned and partially destroyed cortex (Fig. 2). Her blood parameters like Haemoglobin percentage, Total and Differential Leucocyte Count, Platelet Count; Erythrocyte Sedimentation Rate, Liver Function Test and Kidney Function Test were within normal limits. MRI showed a large lobulated hypo intense lesion T1 weighted images & hyper intense lesion in T2 weighted images. The lesion was of approximately 9.1 × 5.7 × 7.8 cm with a small intralesional bleed & calcification arising from supraspinatus part of scapular body causing its partial destruction and displacing adjacent subscapularis and supraspinatus muscles with evidence of infiltration into inner fibers of supraspinatus muscle (Fig. 3). Her chest radiograph was normal.

The clinicoradiological differential diagnosis considered at this stage before proceeding for biopsy included Chondroblastoma, Chondromyxoid Fibroma, Osteochondroma, Aneurysmal Bone Cyst

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Fig. 1. Pre-operative clinical photograph showing normal shoulder movement.

and Chondrosarcoma. Although chondroblastoma usually occur in epiphysis of long bones in the age group of 10 to 25 years, it also can affect the flat bones¹⁸. It is documented in the literature that epiphyseal equivalent areas exist adjacent to the articular cartilages in the pelvis and shoulder girdles. Lesions that commonly develop in these areas are those that show a predilection for the epiphyseal regions of the tubular bones¹⁹. Chondromyxoid fibroma was considered because of age of the patient and its radiographic and MRI appearance. 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 shoulder¹⁰. ABCs may occur in any bone and radiograph usually shows a characteristic expanded bone by a radiolucent lesion that appears to have a honey comb shaped. Fluid–fluid levels are seen with MRI^{10,18}. Chondrosarcoma is the most common primary malignant neoplasm of the scapula. Radiographs may show cortical destruction by a lesion containing a calcified cartilaginous matrix¹⁰.

Incisional biopsy under general anaesthesia was done and histopathology suggested the diagnosis of chondromyxoid fibroma (Fig. 4). Surgical resection of the tumour en mass along with a rim of macroscopically normal appearing bone with preservation of glenoid was planned under general anaesthesia. As the surgical team had no previous experience of tumour excision in scapula, a cadaveric dissection at anatomy department of the institute was done for familiarization.

Under general anaesthesia and in prone position, the Das Gupta's approach¹⁸ was employed using an elliptical incision incorporating the biopsy scar. The lesion was excised with a margin of rim of normal bone and preserving the unaffected part of

scapula including the glenoid (as shown in schematic diagram below).

Schematic diagram showing scapula from dorsal aspect. Yellow coloured continuous mark shows the lesion and red coloured dotted line shows the margin of surgical resection of scapula.

Haemostasis was achieved and wound closed in layers over a drain. Post operatively the left upper limb was supported in an arm sling. Gross examination of the excised lesion revealed a lobulated, scapula expanding neoplasm with greyish white and flesh coloured surface and it was sent for biopsy (Fig. 5).

With subsidence of pain by 7 post-operative days, she was encouraged to remove the arm sling support and undertake gradually upgraded rehabilitation under the guidance of

Fig. 2. Pre-operative X ray showing an eccentric lytic expansile lesion arising from left scapula with distended, thinned and partially destroyed cortex.

physiotherapist. Post-operative radiograph revealed radiological clearance of tumour mass (Fig. 8). The excisional biopsy confirmed the diagnosis of Chondromyxoid fibroma with clear margin (Fig. 6). Clinico radiological follow up at 1, 3, 5 months post-operative period reveals a stable shoulder joint with no loss of preoperative range of movement & no recurrence (Fig. 7).

3. Discussion

Scapula plays an essential role in elevation of arm through 180° from the dependent position to vertical position in abduction or in flexion²⁰. Out of total 180° elevation, humerus moves 120° at the shoulder joint and the scapula contributes the remaining 60° at both acromioclavicular and sternoclavicular joints²⁰. In every 15°

Fig. 3. Pre-operative MRI.

Fig. 4. Microscopic picture from incisional biopsy showing features suggestive of CMF.

elevation, shoulder joint contributes 10° and girdle joints 5° , in the ratio of 2:1. According to Inman et al., the 'scapulo-humeral rhythm' starts concurrently, except in the initial $25\text{--}30^\circ$ when humerus moves at the shoulder joint²⁰. As observed by Kapandji (1974), scapular rotation consists of initial elevation of acromial end of clavicle followed by axial rotation of clavicle²⁰.

Elevation of the clavicle ranges about 30° . Acromioclavicular joint permits slight elevation but the main movement takes place by downward swing of the sternal end of clavicle at the meniscoclavicular compartment of sternoclavicular joint. This elevation is performed by the upper fibres of trapezius and levator scapulae. Further elevation is checked by the tension of

coracoclavicular ligament. Eventually, the scapular rotation is associated with axial rotation of clavicle at the sternoclavicular joint permitting further 30° elevation. This allows the glenoid cavity to face vertically upward. The scapular rotation is done by the upper and lower fibres of trapezius along with the lower five digitations of serratus anterior. The middle fibres of trapezius stabilize the scapula during abduction²⁰.

Chondromyxoid Fibroma is a cartilaginous tumour of bone with varying amounts of myxoid and fibrous tissue elements, with giant cells in some cases and occasional cellular atypia¹¹. It is a benign, although potentially aggressive tumour¹⁷. Eighty percent of patients are younger than 36 years of age²¹. Most series have

Fig. 5. Resected Specimen.

Fig. 6. Clinical photographs at six weeks showing normal range of motion.

demonstrated a definite male predilection^{2–4,8}. Approximately two thirds of the recorded cases of this tumour have been in the long tubular bones and one third in the proximal tibia^{2–4}. This finding is consistent with the hypothesis that the tumour arises from remnants of cartilage at this site^{8,9}.

In our review of the literature, we found only few reports of Chondromyxoid Fibroma of the scapula. Prichard and colleagues¹⁵ reported a case of Chondromyxoid Fibroma of the scapula in a 38-year-old man in 1964 (their case report can be considered the first on this location of this tumour). Uematsu and colleagues²² reported a case of “malignant” Chondromyxoid Fibroma in a 39-year-old woman based on histologic characteristics and tumour invasion to the skeletal muscles. Baklouti and colleagues⁵ noticed, in a 23-year-old woman, extensive invasion of the soft tissues—a feature seldom observed in Chondromyxoid Fibroma. Mizuno and colleagues¹³ reported ABC as a secondary component to Chondromyxoid Fibroma and stated that, though uncommon, Chondromyxoid Fibroma should be considered in the differential diagnosis of a radiolucent, expansile lesion of the scapula in a young patient. Jay B. Jani et al. reported a case of Chondromyxoid Fibroma of scapula in a 13 year old girl & recommended that Chondromyxoid Fibroma of the scapula be treated with wide surgical excision when possible and that the patient be regularly followed up for recurrence¹⁰.

Clinically, the main symptoms are pain, swelling or tenderness, and pressure. Duration of symptoms may range from weeks to even years in adults^{1,23}. An associated pathologic fracture occurs in approximately 5% of cases²⁰. Local swelling may be noticed by a patient whose tumour is not camouflaged by a thick layer of overlying tissue. Occasionally, these tumours are asymptomatic, incidental findings on radiographs^{4,14,15}.

Typical radiographic features of Chondromyxoid Fibroma are a single, lobular, eccentric, radiolucent lesion with expansion of the affected bone⁹. The endosteal margin is usually well demarcated, with a sclerotic rim. Although the tumour is benign, the aforementioned radiographic features confirm its potentially aggressive behaviour¹⁰. Geographic bone destruction in conjunction with clinical findings should raise suspicion of Chondromyxoid Fibroma^{24,25}. Distinguishing the lesion from an aneurysmal bone cyst (ABC) may be difficult. In rare cases, Chondromyxoid Fibroma does not produce bone expansion but may cause an unusually thick, dense rim of reactive bone around the central lucency^{26,27}. Radiographic evidence of calcification is rare in Chondromyxoid Fibroma^{2,13}.

Calcified Chondromyxoid Fibromas are reported more commonly in unusual locations, particularly flat bones^{28,29}.

On MRI, depending on the varying amounts of myxoid and cartilage tissue, the centre of the tumour is hyper intense on T2-weighted spin-echo images and STIR sequences. Along with the typical lobulated pattern, the images are distinctive of a tumour of cartilaginous origin. The highly vascularized connective tissue at the border of the lesion accounts for a rim of moderate to high signal enhancement on T1-weighted images after gadolinium-diethylene triamine penta-acetic acid (Gd-DTPA). Planning the extent of resection with MRI demonstrating soft tissue or bone marrow not seen on the plain radiographs may result in a reduction of the recurrence rate^{30–32}.

Treatment of Chondromyxoid Fibroma has changed since this entity was recognized in 1948. Curettage is associated with a 12.5% to 25% risk for recurrence even with the use of local adjuvants⁶. The recurrence rate is lower with resection using wide margins, although this may not always be possible in certain anatomical

Fig. 7. Follow up X ray at 6 months post op showing no evidence of recurrence.

Fig. 8. Microscopic picture from excised specimen showing features of CMF.

regions^{11,33–35}. Therefore, the preferred treatment, where possible, is an en bloc excision with tumour free margins^{6,10}. Radiation therapy has been used only in exceptional cases for local tumour control. It is however not recommended for tumours amenable to limb-sparing surgical procedures, because of reports of radiation-induced malignancies^{1,2,6,13}. In exceptionally rare cases, especially recurring tumours, an amputation was found to be necessary despite the tumour's benign nature³⁶.

Age seems to have a significant influence on recurrence rate. In younger patients, the tumour 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^{3,6}. In cases of recurrence, soft-tissue extension of the tumour is not rare, presumably because of the displacement of tumour cells during surgery^{5,37}.

Few authors have described Chondromyxoid Fibroma as having undergone malignant transformation (overall incidence, 1%–2%)⁸. Some cases of the malignant behaviour of this tumour may be attributed to erroneous classification of low-grade chondrosarcoma as Chondromyxoid Fibroma⁹.

4. Conclusion

Chondromyxoid fibroma is characterised by local aggressiveness and is associated with as high as 25% recurrence rate even after extended curettage. Therefore, the preferred treatment is an en bloc excision with a rim of normal bone all around to achieve wide marginal resection unless there is constrain due to its anatomical location or proximity to vital structures. When it occurs in scapula, partial resection of the scapula with preservation of glenoid (whenever possible) allows having a stable functioning shoulder joint as well as reducing the risk of recurrence.

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