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# Advanced Unilateral Fibrous Dysplasia of the Scapula: A Rare Clinical Entity and Surgical Challenge

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**Learning Point for this Article:**

Fibrous dysplasia tumor of the scapula is very rare, and we have deployed an innovative technique to get rid of the scapular deformity, which improved shoulder functionality and decreased pain scale.


## Abstract

**Introduction:** Fibrous dysplasia (FD) is an uncommon benign tumor of bone. Although FD can affect flat bones, it is rare for the scapula to be involved. In addition, little is known about the management of FD when it involves the scapula. We present possibly the first comprehensive case report of the management of advanced unilateral FD of the scapular region.

**Case Report:** A 47-year-old male presented to us with pain and swelling over the left shoulder. The swelling was 11 cm × 15 cm × 8 cm and was hard and tender with rough texture. Radiograph showed large homogenous lesion with irregular but well-defined margins and a ground glass appearance. Magnetic resonance imaging scans showed well-defined borders with the expansion of the bone, with intact overlying cortices and endosteal scalloping. Biopsy confirmed the lesion to be FD. An innovative application of an existing surgical technique to minimize the impact of the residual deformity and dead space left after curettage of the scapula was done. The patient had good clinical and functional outcome at 6-month follow-up.

**Conclusion:** Surgical exercise in FD is purely on symptomatic basis. In our case, the swelling was causing most discomfort, and we curettaged and compressed the bony swelling which resulted in excellent outcome in this patient.


**Keywords:** Fibrous dysplasia, functional outcome, unilateral fibrous dysplasia, rare scapula lesion, unique surgical technique.

## Introduction:

Tumors affecting the scapula are uncommon. They represent about 3–4% of all bony tumors [1]. Fibrous dysplasia (FD) is a rare, benign tumor of bone. Overall, FD makes up between 5 and 7% of all benign bone tumors [2]. There is no difference in the incidence of FD between males and females [2]. It may be restricted to a single bone (monostotic) or involve many skeletal sites (polyostotic). Monostotic FD maybe diagnosed as an incidental finding, showing most activity before puberty, unlike polyostotic FD, which may continue to grow after puberty and beyond adolescence [3]. FD predominately affects long bones, ribs, and the craniomaxillofacial skeleton. FD of the scapula accounts for <0.5% of these tumors. It is therefore exceedingly rare. Although considered asymptomatic it can be locally aggressive and may cause pain, swelling, deformity, and have a serious impact on a patient's ability to carry out daily activities. The differential diagnosis of FD includes Paget's disease of bone [4], non-ossifying fibroma [5], osteoblastoma [6], low-grade intramedullary osteosarcoma [7], chondroblastoma, fibromyxoma of bone, and adamantinoma [8]. Ultimately, the diagnosis is based on clinical, radiological, and histopathological findings [2, 9, 10, 11]. The pathological process underlying FD is the replacement of bone with fibrous tissue. [12]. A mutation in the Gsα (G-protein α-subunit) gene located on chromosome 20q13.2–13.3 has been implicated as a possible cause of FD [13]. Treatment of FD can either be

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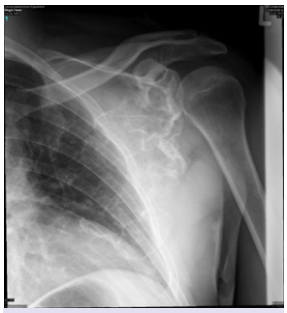

**Figure 1:** Anteroposterior radiograph of the left shoulder showing irregular expansile sclerotic lesions (red arrow) with overlying ground glass haziness (black arrow), involving the scapula, sparing its superior, and medial borders. Features suggest fibrous dysplasia, but chondrosarcoma should be included in the differential diagnosis.

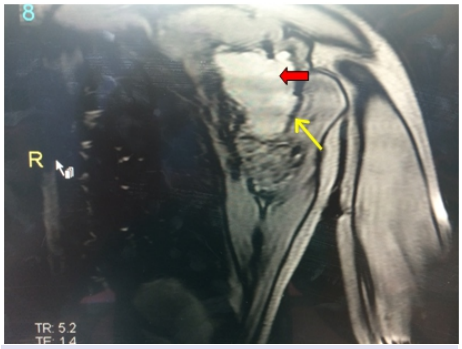


**Figure 2:** Coronal T1-weighted magnetic resonance imaging of the left shoulder showing the expansile scapular lesion (red thick arrow), with high signal intensity surrounded by scalloped low signal intensity rim (yellow thin arrow). The pattern suggests a proteinaceous or hemorrhagic content.

conservative or surgical. Conservative treatment has focused on monitoring and observation of the lesion [14]. Recently, treatment of FD with bisphosphonates has been reported. However, the results have not been conclusive [14, 15]. The surgical management of FD has involved curettage of the affected site and bone grafting when indicated [16]. A literature review has failed to reveal any previous reports on the comprehensive clinical management of these tumors in the scapula. We report the management of advanced FD of the scapula that had given rise to deformity, functional disability, and severe pain of the left arm and shoulder in a 47-year-old male.

## Case Report:

### History of presenting complaint

A 47-year-old male office employee was referred to the orthopedic oncology clinic of the University Hospital Hamburg-Eppendorf by his family physician. On presentation, he had pain and swelling over the posterior aspect of his left shoulder. Both pain and swelling had increased over the previous 10 months. Movement of the left arm made the pain worse, especially by raising the arm over the head. He was otherwise healthy.

### Examination

Clinical examination revealed a large swelling over the posterior aspect of the left shoulder blade. It measured 11 cm × 15 cm × 8 cm. The swelling was hard and tender to palpation. It had a rough texture. The swelling extended entirely over the scapular body and both the supra- and infra-spinous fossae. Sensation over the left suprascapular nerve dermatome was diminished, and the right side was normal. Attempts at flexion and abduction at the left glenohumeral joint were painful, as well as lifting weight in a forward bending position. Distal neurovascularity of the left upper limb was normal. Movement at the right glenohumeral joint was normal. Pain measured by the visual analog scale (VAS) was 8/10. The Revised Musculoskeletal Tumor Society Score was 13/30. There were no respiratory problems.

### Investigations

Serological investigations were within normal limit apart from a raised alkaline phosphatase of 178 IU/L.

Radiological imaging

Conventional radiographs showed a large homogenous lesion with irregular but well-defined margins occupying most of the left scapula. It had a ground glass appearance. Septa appeared to compartmentalize the main body of the lesion. There was an expansion of the involved area of bone dorsolaterally (Fig. 1). Magnetic resonance imaging (MRI) scans showed well-defined borders with expansion of the bone, with intact overlying cortices and endosteal scalloping (Fig. 2 and 3).

### Treatment plan

First, an open biopsy of the lesion under general anesthesia (GA) was taken. Based on the clinical, radiological, and histological findings, the diagnosis of FD was made. The histology is shown in (Fig. 4). In view of the patient's pain (VAS 8/10), which had drastically reduced his capacity to work and subsequently led him to take leave from employment, he was scheduled for exploration and curettage of the lesion.

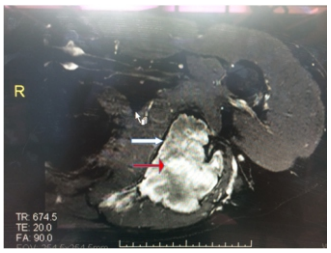


**Figure 3:** Short TI Inversion Recovery (Fig. 3) axial magnetic resonance imaging demonstrating a lesion with high signal intensity (red arrow) surrounded by scalloped margins (white arrow).

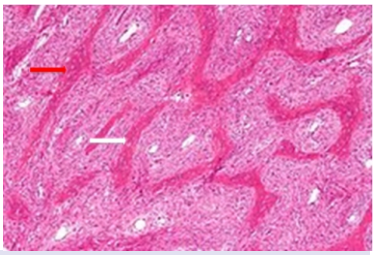

**Figure 4:** Intermediate-power view of typical histology of fibrous dysplasia. Note the bland fibrous stromal tissue (white arrow) with islands of disorganized, immature osteoid (red arrow). A key feature is the absence of rimming osteoblasts around the osteoid.

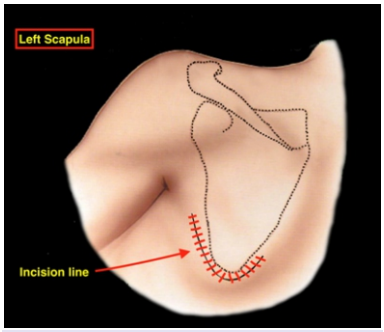


**Figure 5:** Shows pre-operative landmarks of approach and skin incision (red arrow).

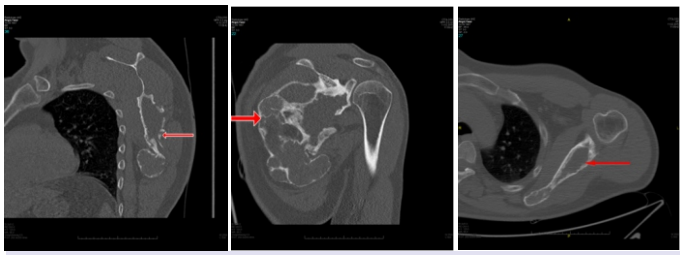

**Figure 6:** (a) Post-operative sagittal computed tomography (CT) scan of the left scapula shows mildly expansile lytic lesion with irregular border (thin arrow). (b) Post-operative coronal CT scan of the left scapula shows some septation of the scapula body with a hazy bone matrix (thick arrow). (c) Post-operative axial CT scan of left scapula shows thinned expansion and deformity of the scapula with surgical defects in the cortex. The dead space has been considerably reduced (red arrow).

### Surgical procedure

The patient was positioned in the prone position. Under GA, a U-shaped incision was made over the inferior angle of the scapula (Fig. 5). Access was achieved by entering between the two cortices of the anteroposterior surface (inferior-medial) of the scapula. A large amount of hard fibrous tissue was removed from the scapular cavity. Curettage proceeded from the inferior angle to the suprascapular area. A large amount of fibrous tissue (430 mg) was obtained from the cavity. The extensive curettage that had been undertaken within the expanded scapula had left a large empty cavity inside the scapula. To reduce the dead space between the two walls of the cavity and the two opposing bony cortices, the sides of the lateral cortices were osteotomised, to flatten the scapula and bring the two opposite cortices together. A green stick fracture was produced at the medial cortices to collapse the two cortical plates. Thus, in one surgical movement, both the dead spaces in the residual surgical cavity and the deformity were reduced. The wound was closed with interrupted sutures. A redivac drain was kept in for 3 days. Post-operative multiplanar computed tomography (CT) scans of the left shoulder were taken at 2 weeks. They showed multiseptated expansile residual lytic lesions in the body of the left scapula. Patchy areas of osteosclerosis were seen in the expanded bone. The sagittal CT scan (Fig. 6a) shows mildly expansile lytic lesion involving the body of the scapula, showing a chondroid/ground glass such as matrix, with some cortical breaks. A coronal CT (Fig. 6b) shows some septation of the scapula body with a hazy bone matrix with probable cortical breaks, and axial CT image (Fig. 6c) shows thinned expansion and deformity of the scapula with surgical defects in the cortex.

### Post-surgical course

The immediate post-operative period was uneventful, apart from mild-to-moderate pain (VAS - 4/10). Before discharge, his left scapula was immobilized with a Gilchrist Bandage. This bandage remained for 4 weeks. At 2 weeks post-surgery review, his sutures were removed. The wound had healed.

### Physiotherapy regimen

Physiotherapy was started in the 2nd post-operative week. Maximum left shoulder abduction and flexion to 90° were encouraged. It was noticed that by the 2nd week, the patients VAS score for pain had improved considerably. It had reduced from the pre-surgery score of 8/10 to a VAS of 3/10. From the 5th week, post-surgery, full exercise of the left shoulder was initiated. The Revised Musculoskeletal Tumor Society score was now 25/30 compared to a score at presentation of 13/30.

### 6-month follow-up

At 6-month follow-up, the patient still had some residual pain (VAS 4/10) in the left scapular region, especially on movement of his left shoulder and lying on it. However, the pain was much less than before surgery and was controlled with ibuprofen. His range of arm movement at his left shoulder was now: 90-0-20° for anteversion/retroversion, 100-0-20° for abduction/adduction, and 50-0-60° for internal rotation/external rotation. The CT scans showed a good surgical result with a collapse of the left scapular cavity. The patient went back to partial employment 4 months after surgery and resumed full-time duties after 6th month.

## Discussion:

FD of scapula is an extremely rare disease. It can behave as a benign silent tumor growing to a considerable size before producing symptoms. That is why, patients with FD, usually complain of swelling (94%) first rather than pain (15%) [17]. However, as our patient attended the oncology clinic with his condition in an advanced state, pain was an important presenting feature. Swelling and deformity were also prominent, as the FD had spread extensively through the left scapula. There was also a reduced range of movements at his left shoulder as well as severe pain and reduced function of his left arm, which had resulted in an inability to work and a constant need for analgesia. Sleep deprivation was a major problem both due to pain and also discomfort in lying on the bed. Imaging and clinical findings may provide sufficient information to make a diagnosis without a biopsy [18]. Radiographically, FD appears as a well-circumscribed lesion with a ground glass or hazy appearance of the matrix. There is sometimes focal thinning of the overlying cortex, called "scalloping from within." The radiological appearance can also be cystic, "Paget like," or dense and sclerotic [19]. CT scan often shows ground glass opacities in about 56% of lesions [20]. It can also have a homogeneously sclerotic appearance in 23% of patients and show cystic changes with well-defined borders in about 21% of cases [21]. Expansion of the bone, with intact overlying bone and endosteal scalloping as with our case report, is quite

characteristic [21]. MRI alone cannot be relied on to make a diagnosis of FD, with lesions appearing more aggressive than on CT scan. Most of the times, the MRI appearance of FD resembles that of tumours [20]. Although bone scans of patients with FD show increased tracer uptake on Tc99, this imaging modality is not specific enough to make a diagnosis [22, 23]. In our case, the ground glass appearance on radiograph and MRI findings was unique of FD. Since the swelling in our case was quite large, a biopy was essential to rule out any malignant changes. Histology of FD is again quite typical. Microscopically, in FD, there are irregular islands of woven bone within a fibrous cellular stroma [21], and this was seen in our case. Macroscopically, the tumor may appear solid white or a tan color [24]. When cut, the exposed surface has a gravel texture because of the fine bone spicules it contains [2]. The management of FD still remains controversial because not all lesions produce pain and functional impairment. However, pain may arise from mechanical pressure as FD grows, fracture of the bony stalk of the tumor, or even nerve impingement syndromes [25]. There was considerable pain, impairment of function, and deformity in our patient to warrant surgical intervention. Indeed without surgical intervention, our patient would not have been able to lead a normal life. Quite often, orthopedic surgeons choose the modified Judet approach for the operative treatment of scapular fractures/small tumor excisions. The modified Judet approach [26] involves preservation of the infraspinatus muscle, which is not dissected out of the scapular fossa. This approach has all the advantages of the classic Judet [27] approach with visualization of almost the entire scapular body but without the morbidity of extensive dissection. However, in our case, we choose a different approach to gain access to the entire scapular cavity, which would not have been possible through a modified Judet approach, as this would have entailed extensive muscle and soft tissue stripping and a risk of injury to the suprascapular nerve. Therefore, a U-shaped incision was made at the inferior angle of the scapula. Access to the scapula was achieved from the inferior angle of the flat bone. The surgical procedure consisted of meticulous curettage followed by drilling of the residual cavities with a large surgical burr. It was then decided to osteotomize the ends of the lateral cortices to flatten the walls of the scapula and close the large residual cavity. The conservative surgical approach that we have outlined for FD of the scapula as seen in Fig. 5 and the method of closing the dead space appears to be unique and not previously cited in the literature. The post-surgical FD patient should be followed up for a long time as the recurrence rate of FD has been reported close to 100% when patients are monitored for many years [28]. FD treated with intravenous pamidronate can lead to reduced bone turnover, less pain, and improvement of radiological lesions [29]. Pamidronate is a potent inhibitor of bone resorption. Like other bisphosphonates, the effects on bone turnover can be present for a long time [29, 30, 31].

## Conclusion:

FD affecting the scapula is exceedingly rare. The presented case outlines the management of FD of the scapula and deploys an innovative surgical technique to minimize the surgical deformity and improve clinical outcome (pain and activities).

### Clinical Message

FD of scapula is an extremely rare problem and not much has been highlighted about it in the literature. Although FD is vastly discussed and explained in the literature but there is not even a single comprehensive paper explaining this problem in scapula and how to deal with its enlarging dimensions which disturbs the patient's daily activity of his shoulder joint. For this, we brought a unique innovative approach in reducing and collapsing the expanded scapular cortices, which resulted in pain relief, improved cosmesis and improved the patient's shoulder activity.

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