



Desmoplastic fibroma of the bone: Report of an unusual location with literature review

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ABSTRACT

Desmoplastic fibroma (DF) is an exceedingly rare benign primary tumor of bone, first described in 1958, which can be locally aggressive. It is considered to be the bony equivalent of the desmoid-type fibromatosis (aggressive fibromatosis of soft tissue). This tumor affects usually long bones and bones of the jaw; rib involvement is extremely uncommon. Its diagnosis is often challenging for the pathologist. Recognition of this entity is important to ensure proper surgical treatment. We report a case of DF of the rib in a 46-year-old woman, with lytic costal lesion and chest wall extension and we present a literature review of this rare disease.

KEY WORDS: Bone, desmoid, fibroma, rib

INTRODUCTION

Desmoplastic fibroma (DF) of bone is a rare, slow growing and locally invasive, benign fibrous tumor. It was first described by Jaffe in 1958 [1]. The reported incidences of DF among all primary and benign bone tumors are 0.06-0.11% and 0.3%, respectively [2]. The tumor occurs usually in young adults without sex predilection [3]. It affects mostly the metaphysis of long bones (femur, tibia) or flat bones (pelvis, facial bones, and spine) [4]. To our knowledge, rib involvement by DF is extremely rare, with only 6 cases reported in the literature to date [5]. Herein, we present the 7th case of DF in the rib of a 46-year-old woman.

CASE REPORT

A 46-year-old woman without a medical history presented with a painful swelling of the left second rib. She had no history of trauma. Chest computed tomography (CT) and magnetic resonance imaging (MRI) found an expansive mass blowing the left second rib with areas of osteocondensation and without soft tissue extension. The surgical procedure consisted of wide resection of the rib. Grossly, the mass was white to yellowish, firm, showing hemorrhagic areas. It measured 4.5 cm × 2 cm × 1.5 cm. On histologic examination, the tumor was moderately cellular and composed of typical spindle cells in a collagenous background with focal hemorrhagic changes. Fibroblasts didn't show atypical features and mitotic figures were absent [Figures 1 and 2]. Some areas revealed entrapped bone within tumoral proliferation [Figure 3]. The tumor was extending focally to

the soft tissue. There was not pleural extension or invasion of surgical limits.

DISCUSSION

DF was first described by Jaffe in 1958 as a rare primary bone tumor that can affect any bone [2]. It is a benign fibrous lesion made up of wavy fibroblasts and abundant collagenous tissue, bearing a close resemblance to the desmoid tumor of soft tissue. Till date, around 200 cases of DF of bone have been reported in the world literature with an incidence of 0.11-0.13% among primary bone tumors [1]. Any bone can be affected, and the most common sites are the long tubular bones (56%), mandible (26%), and pelvis (14%) [2]. Although DF has been reported in rarer locations, rib involvement is extremely rare. To our knowledge, only 6 cases have been published in the English literature, Table 1 summarizes all cases including this case.

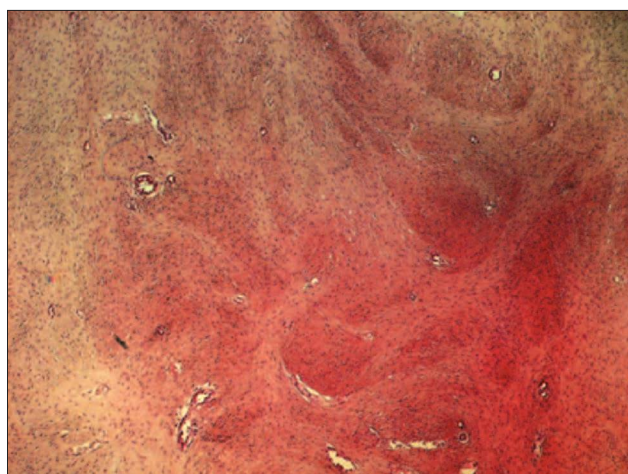
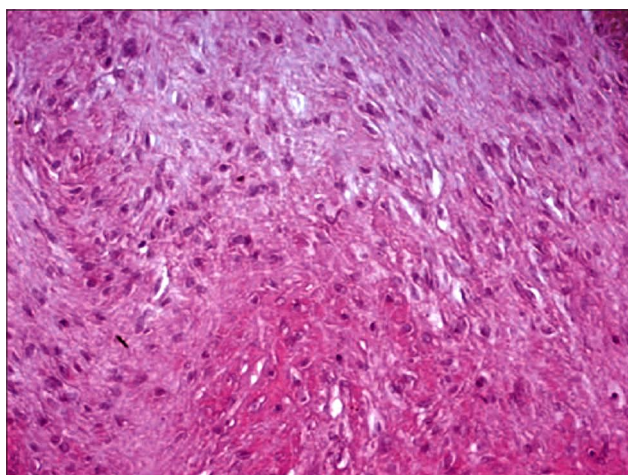
DF of the bone may occur at any age, approximately 75% of patients are younger than 30 years and only 6% are older than 50 years without sex predilection [5]. For the rib location, the mean age for the 7 reported cases is 40-year-old, with a sex-ratio of 5/2.

Clinically, pain or swelling for long duration precedes functional disability; pathological fracture is an uncommon initial presenting symptom. The rarity of the tumor and nonspecific radiographic findings of DF sometimes make radiological diagnosis difficult. Typically, DF are expansile, lytic lesions, often with internal trabeculation, and soap bubble appearance.

Table 1: Summary of reported cases of DF of the rib

Authors	Age	Sex	Clinical symptoms	Location	Size (cm)	Treatment	Follow-up
Butters <i>et al.</i> [6]	17	M	Pain and swelling	L 6 th	-	Resection	-
Takonis <i>et al.</i> [7]	57	F	Swelling	R 5 th	-	Curettage	Recurrence/9 years
Barbashina <i>et al.</i> [5]	19	M	Swelling	L 10 th	11	Total excision	-
Ayadi-Kaddour <i>et al.</i> [3]	45	M	Pain paresthesia	R 2 nd	7	Resection	NED/4 years
Ayadi-Kaddour <i>et al.</i> [3]	55	F	Swelling	R 8+9 th	10	Wide resection	Recurrence/2 years
Okubo <i>et al.</i> [8]	40	M	Swelling	R 9 th	14×12×8	Total excision	NED/6 months
Present case	46	M	Pain swelling	L 2 nd	4.5×2 × 1.5	Wide resection	NED/1 year

M: Male; F: Female; L: Left; R: Right; NED: No evidence of disease, DF: Desmoplastic fibroma

**Figure 1:** Hypocellular proliferation in an abundant collagenous matrix (H and E, ×100)**Figure 2:** Spindle-shaped cells in a background of thick collagen fibers (H and E, ×200)

Cortical thinning and disruption are common findings, often associated with a soft tissue mass. Periosteal reaction is not seen. CT is superior to standard radiography in assessment of the cortical continuity. MRI best demonstrates the extent of soft tissue involvement [5]. However, these findings may be seen in other bone tumors such as fibrous dysplasia, hemangioma, eosinophilic granuloma, and low-grade osteosarcoma [2].

As many tumors have similar radiological appearances to DF, the diagnosis is very difficult on the basis of radiological finding

alone. Thus, biopsy is the gold standard method of determining the exact nature of any bony lesion [9].

Macroscopic features are not specific. The rib is usually expanded by a well-defined, elongated, white-gray, rubbery mass. The tumor has firm texture. Multilocular cystic changes containing a viscous liquid are described in the literature [2,8]. Histologically, DF is characterized by bundles of abundant collagen fibers often forming thickened, hyalinized bands, evenly separated by scarce spindle-shaped fibroblasts of small size, with elongated or ovoid nuclei and without evident mitotic activity. Trabeculae of the lamellar bone and reactive bone formation are focally present within the tumor usually [5,10]. In all reported cases, the tumor was locally aggressive, infiltrating soft tissue; in fact, it was proliferating intermingled with the surrounding skeletal muscles, though the periosteum. In one case, cystic change was observed in the extraosseous lesion, and there were no cells lining cystic spaces.

Immunohistochemistry is not very useful in the diagnosis of DF. The spindle cells usually have a myofibroblastic phenotype and express the alpha-smooth muscle actin and vimentin [11]. They express PS100, CD34 and focally desmin. In DF, there was no nuclear immunoreactivity for estrogen and progesterone receptor and no immunoreactivity for CD117. Hauben *et al.* [10] conducted a study to investigate the immunohistochemical profile and the involvement of the b-catenin pathway in DF as it is known in desmoid-type fibromatosis. According to their results, the b-catenin pathway does not seem to have the same essential role in the tumorigenesis of DF, as it has in desmoid-type fibromatosis.

The differential diagnosis includes fibrous dysplasia and low-grade fibrosarcoma. The hallmark of fibrous dysplasia is formation of immature woven bone, a bony matrix that characteristically lacks the osteoblastic rim. A low-grade fibrosarcoma is ruled out based on low cellularity and the absence of herring bone pattern, mitotic activity, nuclear enlargement, and hyperchromasia.

DF is generally considered as the bone counterpart of aggressive fibromatosis of soft tissue (desmoid tumor) [1]. Results of a recent cytogenetic study on benign fibrous lesions of soft tissue and bone reported by Barbashina *et al.* [5] showed that DF, soft tissue fibromatosis, and fibrous dysplasia share the same nonrandom aberrations (trisomy 8 and trisomy 20), suggestive of a common pathogenesis. There are also rare reports of DF arising in fibrous dysplasia. Although a genetic linkage for desmoid tumors in familial

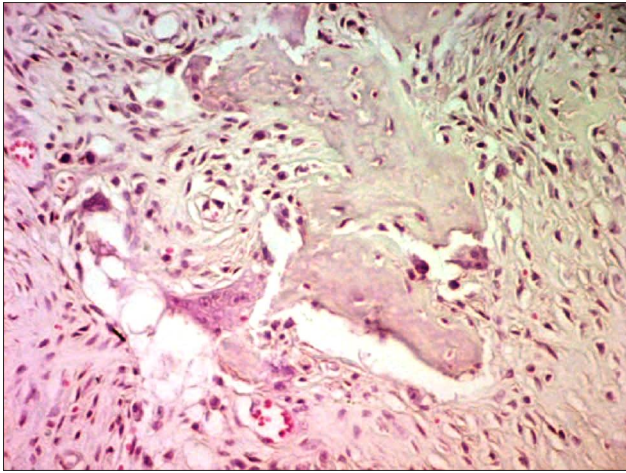


Figure 3: Entrapped bone in the tumoral proliferation (H and E, ×400)

syndromes has been shown, the cause of sporadic desmoid tumors remains to be determined. Local inflammatory change involved in the healing response after trauma has often been postulated as stimulating the development of DF. Even localized trauma such as a surgical incision has been thought to stimulate this development [4]. DF of the chest wall has also been described within thoracotomy scars and after the placement of silicone breast implants. Like desmoid tumors, DF has also been thought to be stimulated by increased levels of estrogen [11].

Management consists of wide surgical excision with bone grafting or prosthetic replacements in appropriate regions. DF does not undergo metastasis; it may, however, recur locally if incompletely removed. Patients treated simply by curettage have a high rate of recurrence which may be related to the cellularity of the lesion [8]. Local recurrence occurred in 15.4% of patients in published series and was statistically significantly linked to DF with associated soft tissue components [9]. This suggests that DF with extraosseous extension area represents the extreme end of the locally aggressive spectrum and, therefore, requires more radical surgery than curettage or marginal excision. Our patient was treated by a wide excision of the rib, without post-operative complications. There were no signs of recurrence 1 year after surgery.

CONCLUSION

In conclusion, a rare tumor like DF of bone needs strong clinical suspicion and an ingenious pathological examination with good supportive radiological investigation for the diagnosis and the management of the lesion. Wide resection is currently the preferred modality of treatment leading to acceptable cure rates and functional outcome.

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