Desmoplastic Fibroma of the Left 6th Rib in a 35-year-old Man: a Case Report and Literature Review

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Case Report

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Abstract

Background: Desmoplastic fibroma (DF) is a rare, benign tumor. The most common sites are the long tubular bones and mandible. Although generally considered a benign tumor, it can invade surrounding tissues and has a high rate of local recurrence after incomplete surgical excision. However, there is currently no standard treatment. Here we present a novel case of DF in the left 6th rib in a 35-year-old man.

Case presentation: The man presented to our center with left chest pain and swelling. Enhanced computed tomography (CT) showed a 4.5×2.0 cm mass on the left 6th rib with pathological fracture. Wide resection was performed. Open biopsy revealed that the tumor destroyed medullary cavity and partially broke through the bone cortex. Pathologically, the tumor was composed of spindle-shaped cells arranging in a woven pattern on a background of abundant collagenous fiber. No β -catenin were detected. Based on the pathological and radiological findings, a final diagnosis of DF was made. No postoperative adjuvant treatments were administered. Fortunately, there was no evidence of recurrence 22 months after surgery.

Conclusions: DF originated from rib is a kind of extremely rare benign tumor but locally aggressive and show unique biological features. Wide resection or total resection can effectively reduce the risk of local recurrence when compared with curettage. Rarity of the tumor favors documentation in literature.

Background

Desmoplastic fibroma (DF) is a rare, locally aggressive, benign bone tumor with an incidence of 0.06% of all bone tumors and 0.3% of all benign bone tumors[1]. It was first reported by Jaffe in 1958[2]. As reported in the literature, the most common sites are the long tubular bones (56%), mandible (26%), and pelvis (14%) [3]. Rare cases of desmoplastic fibroma have been reported in other locations, including the maxilla, skull, scapula, clavicle, sternum, vertebral column, calcaneus, small bones of hands, and other sites. Clinical symptoms of DF are non-specific. Typical radiological features include osteolytic changes with destruction of the cortical bone. However, these findings can be seen in other bone tumors such as fibrous dysplasia (FD), hemangioma, eosinophilic granuloma, and low-grade osteosarcoma. Thus, nonspecific radiographic findings of DF sometimes make the correct diagnosis difficult. Here we present a rare case of DF in 6th rib of a 35-year-old man. After completely excision, there was no recurrence at a follow-up of 22 months. We present the following case in accordance with the CARE-Guideline.

Case Presentation

On April 21st in 2020, a 35-year-old male presented to our hospital, complaining of intermittent left-chest pain for over 8 months. He told me that he had received antibiotic treatment for 10 days in the community hospital, but the chest pain was not relieved notably. He had no tumor family history, tuberculosis and thoracic trauma. The patient reported no fever, cough, expectoration, hemoptysis and hoarseness. Chest enhanced CT revealed a mass (4.2 cm×2.0 cm) of the 6th anterior rib with expanded cystic change and pathological fracture (Fig. 1A&1B). Based on these clinical symptoms and radiological findings, the initial clinical diagnosis was rib chondroma. Preoperative radionuclide bone images revealed bone destruction and active bone metabolism, therefore, we primarily suspected of bone malignancy.

Owing to not completely ruling out malignant or borderline tumor, the patient underwent confine operation in April 24th, 2019. During the surgery, we found that the tumor was expansive growth, so wide resection of the 6th rib

mass and adjacent tissues with 2cm of tumor margin, including costal cartilage of 6th rib, inferior border cortex of 5th rib and superior border cortex of 7th rib, was performed. After resection of the tumor, we used 1 – 0 Prolen suture to bring the 5th rib and 7th ribs closer to furthest reduce the chest wall defect. The surgical specimen was a yellowish tumor with multilocular cystic change containing a viscous liquid (Fig. 2A/2B). The boundary between the tumor and the surrounding intercostal muscles is not clear. An intramedullary tumor located on the proximal portion of left 6th rib destroyed the medullary cavity and penetrated the external cortices, suggesting that the tumor originated from the rib. Pathological features showed that the tumor cells were spindle-shaped against a background of dense collagen fibers (Fig. 2C/2D) and arranged in a woven pattern, lacking atypicality and pleomorphism, and nuclear mitotic figures were rare (Fig. 2E/2F). Immunohistochemistry (IHC) demonstrated focal positivity for smooth muscle actin (SMA), and the staining indices for Ki-67 was approximately 1%. Finally, DF was verified by pathology. After 5 days of postoperative recovery, the patient safely discharged without any complications. There were no signs of recurrence 22 months after surgery by telephone follow-up. A timeline showed the whole medical procedure of the special case (Fig. 3).

Discussion

DF derived from rib is extremely rare and occurs at any age without gender difference. Local pain and swelling are the most common symptoms in DF patients. Up till today, only 7 cases have been published in the English literature[4–8]. Table 1 summarizes all these cases including the present case. The most common clinical symptoms are local chest pain and swelling, and the major imaging features are osteolytic change and cortical bone destruction, as well as invasion into surrounding soft tissues. Pathological fracture, it should be noted, may occur in some cases. Cystic change was found in just two cases. Moreover, total excision and chest wall reconstruction were performed in three cases. However, recurrence occurred in two cases treated with curettage.

Table 1
Summarization of reported cases of desmoplastic fibroma in rib

Author	Age	Sex	Clinical symtoms	Location	Size(cm)	Radiographic findings	Treatment	Follow-up
Butters et al.[1]	17	M	Pain and swelling	Left 6th	-	Osteolytic and non-destruction	Resection	-
Taconis et al.[2]	57	F	Swelling	Right 5th	-	Osteolytic, destruction and soft tissue extension	Curettage	Recurence after 9 year/9 year
Barbashina et al.[3]	19	M	Swelling	Left 10th	11.0	Osteolytic and destruction	Total resection	-
Kaddour et al.[4]	45	M	Pain and hypoesthesia	Right 2nd	7.0	Expensive formation	Resection	NER/4 year
Kaddour et al.[4]	55	F	Swelling	Right 8th or 9th	10.0	Osteolytic and soft tissue extension	Wide resection	Recurence after 2 year/2 year
Taketo Okubo[5]	40	M	Swelling	Right 9th	14.0	Cystic and destruction	Total resection	NER/6 months
Present case	35	M	Pain and swelling	Left 6th	4.2	Cystic and destruction	Total resection	NER/22 months

CT and MRI scan are generally applied to evaluate the invasion degree of tumor for the next planning operation. The rarity and non-specific radiographic findings of DF sometimes make radiological diagnosis rather difficult, because the similar imaging features may be found in other bone lesions such as extra-abdominal desmoid tumor invading bone, eosinophilic granuloma and low-grade osteosarcoma. Radiological findings could confirm whether the tumor is originating from rib or not. In this case, enhanced CT scan showed low-density area with 6th rib destruction, and it revealed that the tumor was aggressive. Moreover, the pathological differential diagnosis is obviously a challenging work, which means that pathologists have to exclude other benign or low-grade malignant bone tumors such as FD, low-grade intraosseous osteosarcoma and low-grade fibrosarcoma[9]. In view of the difficulty in differential diagnosis, making a correct diagnosis is rather essential. Furthermore, immunohistochemical of β -catenin are important for differential diagnosis, since the APC/ β -catenin pathway has been proved to be associated with desmoid-type fibromatosis. Several studies showed that β -catenin plays a vital role in the tumorigenesis of desmoid tumors other than DF[10, 11]. In the present case, IHC of β -catenin is negative, thus further certifying the diagnosis of DF.

The rate of recurrence in DF dealt with local curettage or intralesional resection is almost 40%[1, 12]. Due to the risk of tumor recurrence, the optimal treatment for DF is wide or total resection with partial normal tissue around the tumor. However, there is no uniform treatment guideline for DF currently. Local irradiation is not routinely recommended because of its side-effects. If there was a huge defect in chest wall after surgery, repairment with the tissue-engineered ribs should take into consideration[13]. Postoperative recurrence may occur in those who just underwent curettage or wide resection especially in those large tumors[5, 7]. Therefore, it is a crucial step to choose

the surgical approach. Total resection may reduce the rate of local recurrence but increase the chest surgical trauma and bring patients unsatisfactory chest-wall appearance. Considering that the rib tumor was relatively small, we finally performed wide resection rather than total resection. In addition, another opinion about the tumorous type is that DF is a kind of “border-line” tumor rather than benign tumor[1, 12]. Metastases have never been reported. In my opinion, this view can more accurately describe the biological behavior of the tumor. Fortunately, this case successfully received wide excision with no evidence of recurrence 22 months after surgery.

In summary, this case demonstrates aggressive characteristic of desmoplastic fibroma despite its benign nature and rarity. If the rib mass is relatively small and difficult to exclude malignancy, wide excision should be performed to reduce the rate of recurrence.

Conclusions

DF originated from rib is a kind of extremely rare benign tumor but locally aggressive and show unique biological features. Wide resection or total resection can effectively reduce the risk of local recurrence when compared with curettage. Rarity of the tumor favors documentation in literature.

Abbreviations

DF: desmoplastic fibroma; CT: Computed tomography; FD: fibrous dysplasia; IHC: immunohistochemistry; SMA: smooth muscle actin; MRI: magnetic resonance imaging; APC: antigen-presenting cell.

Declarations

Acknowledgements

Not applicable.

Authors' contributions

Zhang K and Zhang J designed the study; Wu WB and Wu YH acquired clinical data; Guan JX performed the pathological examination; Chen HG and Gu LJ performed the image examination; Zhang Kai wrote the manuscript; Zhang J and Wu WB revised the manuscript. All authors issued final approval for the version to be submitted.

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Ethics approval and consent to participate

The need for ethics approval and consent was waived, since a consent for publication was obtained from the patient.

Consent for publication

Written informed consent for publication of the clinical details and/or clinical images was obtained from the patient. A copy of the consent form is available for review by the Editor of this journal.

Competing interests

The authors declare that they have no competing interests.

References

1. Gebhart. M, Campbell. C, Schiller. A, Mankin. H. Desmoplastic fibroma of bone. A report of eight cases and review of the literature. *The Journal of bone and joint surgery American volume* 1985;67(5):732-747.
2. Jaffe H. Tumors and tumorous conditions of the bones and joints. Philadelphia: Lea & Febiger 1985;p. 298-303.
3. Smith S, Kransdorf M. Primary musculoskeletal tumors of fibrous origin. *Semin Musculoskelet Radiol* 2000;4(1):73-88.
4. M Butters, H Hamann, Mohr. W. Desmoplastic fibroma of the rib. *The Thoracic and cardiovascular surgeon* 1985;33(5):317–318.
5. TakonisWK, SchutteHE, HeulRO. vd. Desmoplastic fibroma of bone: A report of 18 cases. *Skeletal Radiol* 1994;23(4):282-288.
6. Violetta Barbashina, Rouzan Karabakhtsian, Seena Aisner, Paul Bolanowski, Francis Patterson, Hameed. M. Desmoplastic fibroma of the rib. *ArchPathol Lab Med* 2002;126:721-722.
7. Ayadi-Kaddour A, Ben Slama S, Braham E et al. Desmoplastic fibroma of the rib: Two case reports. *Annales de pathologie* 2005;25(5):398-401.
8. Taketo Okubo, Tsuyoshi Saito, Tatsuya Takagi, Yoshiyuki Suehara, Kaneko. K. Desmoplastic fibroma of the rib with cystic change: A case report and literature review. *Skeletal Radiol* 2014;43(5):703-708.
9. L. Perlick, D. Zander, T. Wallny, Zhou H. Desmoplastic fibroma of the fibula. A difficult clinical, radiological and histological diagnosis. *Zentralbl Chir* 200;125(11):895-899.
10. Bo N, Wang D, Wu B, Chen L, Ruixue M. Analysis of beta-catenin expression and exon 3 mutations in pediatric sporadic aggressive fibromatosis. *Pediatric and developmental pathology : the official journal of the Society for Pediatric Pathology and the Paediatric Pathology Society* 2012;15(3):173-178.
11. Colombo C, Bolshakov S, Hajibashi S et al. Difficult to diagnose' desmoid tumours: A potential role for ctnnb1 mutational analysis. *Histopathology* 2011;59(2):336-340.
12. Paul Bohm, Stefan Krober, Annette Greschniok, Michael Laniado, Kaiserling. E. Desmoplastic fibroma of the bone. A report of two patients, review of the literature, and therapeutic implications. *Cancer* 1996;78(5):1011-1023.
13. Hui-Qi Xie, Fu-Guo Huang, Yong-Fan Zhao et al. Tissue-engineered ribs for chest wall reconstruction: A case with 12-year follow-up. *Regen Med* 2014;9(4):431-436.

Figures

Figure 1

A & B Enhanced CT revealed a mass (4.2 cm×2.0 cm) of the 6th anterior rib with destruction of bone cortex and pathological fracture.

Figure 2

A & B The specimen was a irregularly shaped yellowish tumor with multilocular cystic change containing a viscous liquid. C Tumor cells grow invasively and infiltrate the surrounding non-tumor trabecular bone. D The background of tumor cells is plenty of dense collagen fibers. E & F Tumor cells were spindle-shaped and arranged in a woven pattern, lacking atypicality and pleomorphism, and nuclear mitotic figures were rare.

Figure 3

A timeline showing the medical procedure of the case.

Supplementary Files

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