Case Report

Chondromyxoid Fibroma of Ribcage: Case Report

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Abstract

Background: Chondromyxoid Fibroma (CMF) is a slow growing benign tumor of chondroblastic origin. It is more frequent in the metaphysis of long bones that represent between 0.5% to 1% of all primary bone tumors. Its costal location is not frequent and the diagnosis is difficult prior to resection.

Case presentation: We report the diagnosis and management of a 68-year-old male with a chondromyxoid fibroma originating in the fifth rib of who consulted for a palpable chest wall tumor. Magnetic resonance imaging demonstrated a rounded lesion with expansive aspect and well-defined lobe borders in the fifth rib. We perform a complete surgical resection of these tumor and reconstruction of chest wall. No recurrence of the tumor at 12 months of follow up was demonstrated.

Objectives: Demonstrate the management of a patient with a rare tumor of the chest wall.

Conclusion: CMF is an exceedingly rare tumor and its costal location in possible. The definite diagnostic only can be made after histopathologic examination. Complete surgical resection of these lesions with tumor-free margins is recommended.

Keywords: Chest wall reconstruction; Chondromyxoid fibroma; STRATOS fixation System; Chest wall tumors; Case report

Introduction

Chondromyxoid fibroma (CMF) is a slow-growing benign tumor of chondroblastic origin. It represents between 0.5% to 1% of all primary bone tumors. The tumor is slightly more common in men, usually in the second decade of life. Almost half of the tumors involve the long bones, tibia principally. Its costal location is unusual [1]. Oftentimes, they are misdiagnosed as chondrosarcomas and are excised due to concern for malignancy. Metastases do not occur from these benign tumors but recurrence can occur after incomplete resection [2,3]. We introduce the case of the resection of a CMF originated in the fifth rib. This case report is reported in line with the SCARE Guidelines [4].

Case Presentation

We introduce the case of a 68-year-old man with no significant medical history that consulted us for a chest wall palpable tumor. It was located above the fifth anterior left costal arch, in the chondrosternal joint. It presented well-defined borders, smooth surface, hard consistency, of about 6 cm in diameter, which caused a visible chest wall deformity. The patient manifests that he has had the tumor since adolescence, and it had increased the size over the last years; at the same time, it had become slightly painful. A chest X-ray revealed a costal tumor on the left rib cage. Subsequently, a Magnetic Resonance Imaging (MRI) was made, which showed the existence of a rounded lesion with expansive aspect and well-defined lobe borders, of a heterogeneous intensity, which shows fibrous linear opacities with central predominance, of hypointense development predominantly on T1 sequences, hyperintense, on T2 sequences, without restricted molecular motion of water on diffusion sequences, estimated in 49 mm × 43 mm × 48 mm. It is located at the level of the anterior arch of the fifth rib, and generates mass effect on the surrounding structures, without invading them. It presents spared plane separation with surrounding soft tissues (Figure 1 and 2). Additional imaging studies were not performed. We decided to perform a surgical resection owing to the slow growth and the absence of malignant signs, both in the clinical practice as well as the images.

We performed en block resection of the tumor through safety margin; therefore, it was required a partial resection of the fifth rib and cartilage (Figure 3 and 4). We reconstruct the defect through the placement of a titanium costal osteosynthesis system affixed sideways to the rib and medially to the sternum, through a bridge between them, previously moulded to achieve a suitable thoracic reconstruction (STRATOS System) (Figure 5). We also placed a polypropylene mesh underneath the titanium bridge to avoid lung herniation. During postoperative treatment, the patient manifested mild subcutaneous emphysema and mild contralateral pneumothorax that was monitored expectantly. Subsequently, the patient evolves favorably, with optimal pain management and is discharged on the fifth postoperative day. In the long-term follow-up (12 months later), the patient does not present imaging or clinical signs of relapse.


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Figure 1: MRI T2. Space-occupying lesion, compatible with Chondromyxoid fibroma. Axial cut.
Chondromyxoid fibroma is a rare but benign primary bone neoplasm, accounting for less than 1% of all tumors of this type [1]. This tumor was first described in 1948 by Jaffe and Lichtenstein as a separated neoplasm from chondrosarcoma, characterized by slow-growing neoplasms of chondroblastic origin [5].

All bone sites may be involved at the onset of these lesions, but they predominantly arise from long bone metaphysis, particularly the distal femur, proximal tibia or proximal fibula. Costal affection is rare and just represented 22 of 278 cases described by Wu et al. [1,6,7].

The nodular lesion from the surgical sample measured 5.5 cm × 4 cm × 5 cm (Figure 6). The histological sections reveal a benign tumoral lesion, conformed by hypocellular areas and other slightly hypercellular areas that surround bone trabeculae and hyaline cartilage. The cells are fused to stellate, of dispersed chromatin in the nuclei, without mitosis. Which are located in the middle of a fibromyxoid stroma? Neither necrosis nor cytologic atypia are observed (Figure 7). Findings suggestive of chondromyxoid fibroma.

**Figure 2**: MRI T2. Space-occupying lesion, compatible with Chondromyxoid fibroma Coronal cut.

**Figure 3**: Intraoperative picture, Chondromyxoid fibroma before resection.

**Figure 4**: Intraoperative picture, Partial resection of the fifth rib and costal cartilage.

**Figure 5**: Intraoperative photo, Reconstruction of chest wall using STRATOS fixing system.

**Figure 6**: Chondromyxoid fibroma with rib fragment.

**Figure 7**: Photomicrograph shows that cells are fused to stellate, of dispersed chromatin in the nuclei, without mitosis. There are located in the middle of a fibromyxoid stroma.

**Discussion**

Chondromyxoid fibroma is a rare but benign primary bone neoplasm, accounting for less than 1% of all tumors of this type [1]. This tumor was first described in 1948 by Jaffe and Lichtenstein as a separated neoplasm from chondrosarcoma, characterized by slow-growing neoplasms of chondroblastic origin [5].

All bone sites may be involved at the onset of these lesions, but they predominantly arise from long bone metaphysis, particularly the distal femur, proximal tibia or proximal fibula. Costal affection is rare and just represented 22 of 278 cases described by Wu et al. [1,6,7].
There is no racial or sexual predilection although males might found in patients by the second or third decades of life, [8]. It has been described a second peak of incidence from the fifth to the seventh decades approximately, in these patients, lesions are observed more frequently in unusual locations [9-15], as in the case of our patient.

About 70% of patients are symptomatic at the time of diagnosis. CMF usually cause progressive pain, swelling or cough [2,16,17] and, occasionally, can be completely asymptomatic or may present with a pathological fracture [18]. Our patient presents chest wall palpable tumor.

The definitive diagnosis of this lesion could be exceptionally difficult before surgical procedure [19], initially considering as differential pathologies the presence of fibrous dysplasia, aneurysmal bone cyst, chondroblastoma, chondrosarcoma, periosteal chondroma, intraosseous schwannoma, and periosteal hemangiomata [2,16,18].

These injuries are also difficult to differentiate through imaging test. Conventional radiography could frequently show an expansive ovoid or lobulated osteolysis lesion, with radiolucent pattern and well defined sclerotic margins, septa and overlying cortical thinning [18-20]. A Computed Tomography (CT) or MRI is preferable for diagnosis. Cortical changes are better evaluated in CT and range between cortical thinning, making progress until cortical destruction [20]. Although some lesions contain calcification foci, internal mineralization is rare, and just appears in 2 to 13% of the lesions [21]. MRI is the preferred radiology modality to restrict the extension of the tumor, since it provides information regarding soft tissues affection. In this case, all the lesions are isointensive uniformly to the muscle in T1WI and mostly present heterogeneous signal intensity in images T2-STIR o T2-FS due to the different composition of fibrous constituents, chondromyxoid and myxoid [20]. The CMF often appear hyperintensive in PET, what creates concern owing to the probability of malignancy.

A definitive diagnosis can only be carried out after a biopsy and histopathological analysis. Histologically, the CMF show a unique relative appearance, with a specific lobulation; and such lobes host cells on a myxoid or chondroid matrix. Lobes periphery tends to be more cellular than the center [22].

Surgical exeresis of these lesions with tumor-free margins to avoid local relapses recommended, which has a rate of 4% to 20% subsequent to surgical procedure [23,24]. Chest wall reconstruction with rigid material is necessary in cases of 5 cm or larger wall resections. In our patient, we applied STRATOS system made up of malleable titanium bars and rib and sternal clips. We consider that this system adapts to the required chest wall shape, due to titanium bars flexibility and the absence of parietal dead space. We incorporated a polypropylene mesh to avoid lung herniation in these patients. This combination provides as a result, a minimal morbidity, it is easy to use; it prevents paradoxical breathing and also provides aesthetic appearance in the repair of large chest wall defects.

Conclusion
CMF is an exceedingly rare tumor and its costal location in possible. The definite diagnostic only can be made after surgical resection and histopathologic examination. Complete surgical resection of these lesions with tumor-free margins to reduce the risk of tumor recurrence is recommended.

References