Case report

A giant cell tumor of the bone in the rib cage left to proliferate unfettered for seven years to an extensive size

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ABSTRACT

Giant cell tumors of the bone are generally benign tumors of the bone, though they can be locally invasive in nature. They are also known as “osteoclastomas,” and patients are typically between 20 and 40 years of age, who present with pain and swelling of the joints. Though the tumor is benign, malignant degeneration, metastasis, and other complications of tumor growth are possible. Here we present a case where a delay in treatment led to a significant tumor burden. This tumor’s unique location in the anterior arc of the rib, as well as its growth to a size that has rarely been reported, ultimately caused major compressive effects that significantly impacted our patient’s quality of life.

1. Introduction

Giant cell tumors of the bone (GCTB) are benign, but highly invasive tumors that usually affect the epiphysis of long bones. They consist of neoplastic stromal cells, mononuclear cells and reactive multinucleated giant cells [1]. Giant cell tumors (GCTs) located in the anterior ribs are rare. In this report, we present the case of a GCTB that was untreated at diagnosis and left to grow until its massive size spanning the right anterior 4th – 6th ribs significantly impacted the patient’s activities of daily living.

2. Case presentation

A 41-year-old female presented with extreme shortness of breath and fatigue for the past half year. The patient reports that a lump was discovered in her chest about 7 years ago, but she did not seek care at the time. Initially, it was the size of a coin but increased to the size of a grapefruit. She reports that the shortness of breath has been present for a long time, but that over the past few months, she has experienced dyspnea even on light exertion, which severely hinders her daily life. On exam, she was afebrile, normotensive with tachycardia (108 beats per minute). Her respiratory rate was 18 breaths per minute and oxygen saturation was 93%. Cardiac exam revealed tachycardia without rubs or murmurs. Lung exam revealed normal chest excursion with clear lung sounds bilaterally without rhonchi, rales or wheezes. Chest exam revealed a hard, non-mobile non-tender nodule located at the lower aspect of her right rib.

Upon admission to the hospital, a contrast-enhanced computed tomography (CT) scan revealed a mass occupying the 5th rib, with destruction of the normal bone structure of the rib (Fig. 1) and visible compression on the right heart border and deviation of the mediastinal structures. The mass was non-enhancing. Based on these findings, a primary bone tumor with low likelihood of metastasis was suspected. The tumor was removed by osteotomy of nearly the entire 5th rib, and the anterior parts of the 4th and 6th ribs. The diaphragm was also partially resected, and parts of the upper and lower lobes of the right lung were removed. The resected specimen measured 16 cm × 12 cm × 12 cm (Fig. 2). Biopsy revealed a background of stromal cells with round and ovoid shapes, cells with foamy cytoplasm, areas of necrosis, as well as the presence of multinucleated giant cells. A diagnosis of GCTB was made.

Chest wall reconstruction was performed, and in a follow up visit three months later, there was no evidence of recurrence. The patient’s chest CT appeared normal at that time.

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Abbreviations: Giant cell tumor of the bone (GCTB); giant cell tumor (GCT); computed tomography (CT).
often present with symptoms such as feelings of pain or heaviness in the
region when the tumor occupies joint spaces [3]. GCTB in the ribs most-
commonly arises in the distal femur and proximal tibia [1], with a rare location in the
ribs occurring in only about 1% of cases [2,3]. Of those that do arise
from the ribs, the anterior aspect is a relatively rare location [3]. In
addition, GCTB does not usually grow to very large sizes. A review of
literature by Sharma and Armstrong [3] of 13 cases revealed that only
one consisted of a tumor greater than 15cm in diameter, like the one in
our report.

General symptoms include pain, swelling, and limited range of mo-
tion when the tumor occupies joint spaces [3]. GCTB in the ribs most
often present with symptoms such as feelings of pain or heaviness in the
chest that can radiate, or no symptoms that affect activities of daily life
[2–7]. Our patient’s presentation of severe shortness of breath has not
been reported in other cases.

GCTB is usually not suspected in patients who present with a chest
mass, with conditions such as metastatic tumor [6], lymphoma [6],
chondrosarcoma [6], thymoma [8], and breast mass [5] being more
heavily considered as differential diagnoses. The diagnosis of GCTB can
be done based on history and physical, and radiographic and histologic
findings.

Various attempts have been made to grade and classify GCTB based
on radiographic and histologic characteristics. The Jaffe histologic
grading system classified tumors as benign, aggressive, or malignant [9],
but the system was found to be an unreliable prognostic factor [10].
Campanacci et al. [10] used a radiographic-based approach, with grades 1
through 3, based on tumor margins and cortical involvement as seen
on radiographic imaging. The Enneking grading system is similar to
Campanacci’s, and considers radiographic as well as clinical findings
[9]. Between the two, the Campanacci grading system is more widely
used, though, ultimately, neither of the proposed grading systems have
significant value in predicting prognosis, recurrence, nor in taking ac-
count various risk factors to help guide intervention [11]. Standard of
care at our patient’s hospital applies the Campanacci scale only to GCTB
found in the long bones of the extremities, the most typical locations of
GCTB. Additionally, our patient’s tumor size and location necessitated
wide resection, a decision unlikely to be changed based on tumor staging.

GCTB is usually treated with curettage followed by bone filling [1].
Larger tumors can be resected using wide resection [1], as in our case, or
amputation if necessary [1]. Radiotherapy is not recommended due to
risk of malignant transformation [6]. Curettage has been linked to up to
40% rates of recurrence. Wide resection has been found to have little to
no recurrence, though rates of post-operative complications are signif-
ically higher than in curettage [12]. Adjuvants to curettage therapy
have helped decrease recurrence rates, and modalities include cryosur-
gery, high-speed burring, phenol, and more [12]. Though surgery re-
ains the mainstay of treatment, chemotherapy options are available as
well. Bisphosphonates are one of the most favored agents due to their
anti-osteoclastic action; in particular, nitrogen containing bisphospho-
nates such as Zoledronic acid are especially cytotoxic to osteoclasts [9].
Denosumab, a relatively newer anti-osteoclastic agent that acts via the
RANK-L pathway, may also be a good option, especially as a neo-
adjuvant to surgical intervention or in unresectable tumors. It has been
shown to reduce morbidity and improve outcomes in such settings [13].

Seeing as recurrence is not uncommon, clinicians should be diligent
in monitoring for it. There are no official guidelines, though Boriani
et al. [14] has suggested to monitor regularly with CT or MRI in the first
5 years post-surgery - every 3 months for the first 2 years and then every
6 months for the following 3 years.

It is worthwhile to consider the context in which this case took place
in China. China and Japan have significantly higher incidences of GCTB
than the United States [15]. In the United States, GCTB only makes up
3–5% of primary bone tumors, whereas in China, the prevalence reaches
over 20% [9,16]. A relatively higher prevalence of GCTB among primary
bone tumors has also been reported in the Swedish population, though
those findings were attributed to more advanced diagnostics and
comprehensive reporting [17]. Currently, a review of the literature has
not offered any explanations as to why GCTB is more prevalent in Asia,
and the need for more genetics-based studies have been proposed [15].
Furthermore, this patient is from a remote area in China, where resi-
dents are generally of low socioeconomic backgrounds, which make it
difficult to access education on wellness and health maintenance. In this
patient, a mass was discovered over seven years ago, but because her
symptoms were only mild, she felt it wouldn’t be worth the amount of
money it would take to seek treatment. This case then, is an important
reminder of the importance of providing adequate patient education and
promoting access to affordable healthcare.

4. Conclusion

We report a case of a large GCTB on the anterior aspect of the ribs.
This case is unique in its location, size, and severe presentation. It shows
a case where the tumor was left to progress for a long span of time (seven
years), at which time it caused significant impairment to the patient’s
life. Earlier intervention for such tumors greatly reduces the burden of
disease. The tumor was resected and the chest wall was reconstructed,
and the patient has not shown recurrence thus far.
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Declaration of competing interest

The authors have no conflict of interest, financial or otherwise.

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