Respiratory Medicine Case Reports 33 (2021) 101419

Contents lists available at ScienceDirect

Respiratory Medicine Case Reports

journal homepage: www.elsevier.com/locate/rmcr

Case report

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

Ellen Yang [a,1,*], Tianyang Li [b,1], Ariel Katz [c]

[a] Chicago Medical School, Rosalind Franklin University of Medicine and Science, North Chicago, 60064, IL, USA
[b] Department of Thoracic Surgery, Jinzhou Central Hospital, Liaoning, 121000, China
[c] Clinical Sciences Department, Chicago Medical School, Rosalind Franklin University of Medicine and Science, North Chicago, 60064, IL, USA

ARTICLE INFO

Keywords:
Giant cell tumors
Bone tumors
Rib
Anterior arc
Osteoclastoma

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)[2] 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 x 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.

* Corresponding author. 1414 N. Noble St. Unit CH2, Chicago, IL 60642, USA.
E-mail address: ellen.yang@my.rfums.org (E. Yang).
[1] Authors contributed equally to the manuscript.
[2] Abbreviations: Giant cell tumor of the bone (GCTB); giant cell tumor (GCT); computed tomography (CT).

https://doi.org/10.1016/j.rmcr.2021.101419
Received 8 December 2020; Received in revised form 21 February 2021; Accepted 27 April 2021
Available online 1 May 2021

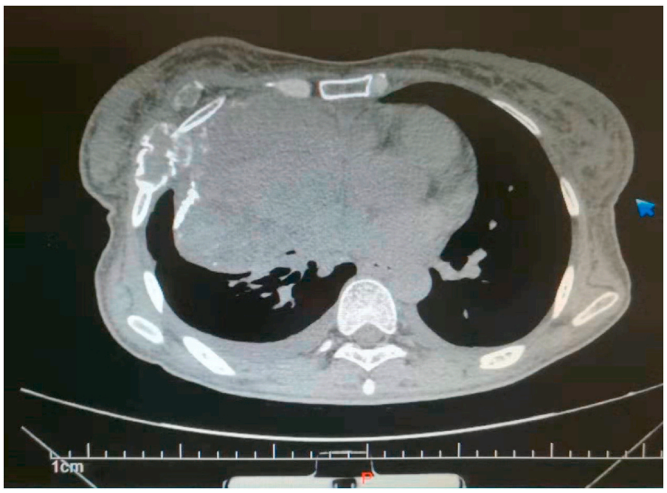

Fig. 1. Computed Tomography image of mass in the right thorax, with invasion of the rib and involvement of thoracic structures.

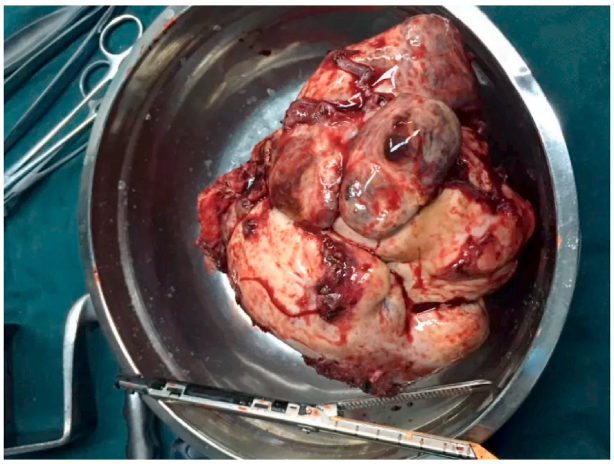

Fig. 2. Resected surgical specimen measuring 16 cm × 12 cm x 12cm.

3. Discussion

GCTB is a rare condition that comprises about 5% of primary bone tumors in adults [1]. The most common locations for giant cell tumors are 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 motion 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 account 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 significantly higher than in curettage [12]. Adjuvants to curettage therapy have helped decrease recurrence rates, and modalities include cryosurgery, high-speed burring, phenol, and more [12]. Though surgery remains 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 bisphosphonates 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 neoadjuvant 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 residents 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.

Funding

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

Declaration of competing interest

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

Acknowledgements

Thank you to Dr. Ligaya Marasigan for her assistance in proofreading the article.

References

[1] M. Balke, L. Schremper, C. Gebert, et al., Giant cell tumor of bone: treatment and outcome of 214 cases, J. Canc. Res. Clin. Oncol. 134 (9) (2008) 969–978, https://doi.org/10.1007/s00432-008-0370-x.

[2] V. Gupta, R. Mittal, Giant cell tumor of rib–rare location on the anterior aspect, Arch. Orthop. Trauma Surg. 120 (3–4) (2000) 231–232, https://doi.org/10.1007/s004020050053.

[3] A. Sharma, A.E. Armstrong, Giant cell tumor of rib arising anteriorly as a large inframammary mass: a case report and review of the literature, Case Rep Med 2012 (2012) 850509, https://doi.org/10.1155/2012/850509.

[4] W. Heo, D.K. Kang, H.K. Min, H.J. Jun, Y.H. Hwang, Giant cell tumor arising from anterior arc of the rib, Korean J Thorac Cardiovasc Surg 46 (5) (2013) 377–379, https://doi.org/10.5090/kjtcs.2013.46.5.377.

[5] G.D. Kalra, A. Agarwal, Rare gigantic giant cell tumor of anterior ribs in a young lady, Indian J Surg Oncol 5 (4) (2014) 300–302, https://doi.org/10.1007/s13193-014-0352-1.

[6] J.S. Shin, I.S. Lee, A. Kim, B.H. Kim, Giant cell tumor originating from the anterior arc of the rib, J. Kor. Med. Sci. 17 (6) (2002) 849–851, https://doi.org/10.3346/jkms.2002.17.6.849.

[7] Heng Du, Huan Xu, Guowei Che, Malignant giant cell tumor of the rib with lung metastasis in a man, J. Thorac. Dis. 6 (9) (2014) 1307–1310, https://doi.org/10.3978/j.issn.2072-1439.2014.07.43.

[8] K.E. Volmar, T.A. Sporn, E.M. Toloza, S. Martinez, L.G. Dodd, Xie Hb, Giant cell tumor of rib masquerading as thymoma: a diagnostic pitfall in needle core biopsy of the mediastinum, Arch. Pathol. Lab Med. 128 (4) (2004) 452–455, https://doi.org/10.1043/1543-2165(2004)128<452:GCTORM>2.0.CO;2.

[9] A.F. Mavrogenis, V.G. Igoumenou, P.D. Megaloikonomos, G.N. Panagopoulos, P. J. Papagelopoulos, P.N. Soucacos, Giant cell tumor of bone revisited, SICOT J 3 (2017) 54, https://doi.org/10.1051/sicotj/2017041.

[10] M. Campanacci, N. Baldini, S. Boriani, A. Sudanese, Giant-cell tumor of bone, J Bone Joint Surg Am 69 (1) (1987) 106–114.

[11] H. Wang, N. Wan, Y. Hu, Giant cell tumour of bone: a new evaluating system is necessary, Int. Orthop. 36 (12) (2012) 2521–2527, https://doi.org/10.1007/s00264-012-1664-9.

[12] J. Ke, S. Cheng, M.Y. Yao, et al., Novel strategy of curettage and adjuvant microwave therapy for the treatment of giant cell tumor of bone in extremities: a preliminary study, Orthop. Surg. 13 (1) (2021) 185–195, https://doi.org/10.1111/os.12865.

[13] D.T. Miles, R.T. Voskuil, W. Dale, J.L. Mayerson, T.J. Scharschmidt, Integration of denosumab therapy in the management of giant cell tumors of bone, J. Orthop. 22 (2020) 38–47, https://doi.org/10.1016/j.jor.2020.03.020. Published 2020 Mar 28.

[14] S. Boriani, S. Bandiera, R. Casadei, et al., Giant cell tumor of the mobile spine: a review of 49 cases, Spine (Phila Pa 1976 37 (1) (2012) E37–E45, https://doi.org/10.1097/BRS.0b013e3182233ccd.

[15] W. Guo, W. Xu, A.G. Huvos, J.H. Healey, C. Feng, Comparative frequency of bone sarcomas among different racial groups, Chin Med J (Engl). 112 (12) (1999) 1101–1104.

[16] H.W. Sung, D.P. Kuo, W.P. Shu, Y.B. Chai, C.C. Liu, S.M. Li, Giant-cell tumor of bone: analysis of two hundred and eight cases in Chinese patients, J Bone Joint Surg Am 64 (5) (1982) 755–761.

[17] J. Rockberg, B.A. Bach, J. Amelio, et al., Incidence trends in the diagnosis of giant cell tumor of bone in Sweden since 1958, J Bone Joint Surg Am 97 (21) (2015) 1756–1766, https://doi.org/10.2106/JBJS.O.00156.