Giant Cell Tumor in anterolateral rib portion

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ABSTRACT

Giant Cell Tumor is a benign but aggressive osteolytic neoplasm, richly vascularized and with numerous giant cells with osteoclastic activity. It often occurs in long bone epiphyses and rarely affects the ribs (about 1% of cases). The presentation of this tumor in the anterolateral portion of the costal arch is extremely rare compared to the posterior region. The present report describes the case of a patient who had a painful tumor on the left anterolateral chest wall, of insidious growth. Imaging exams revealed an insufflating lesion in the 10th left costal arch (4.2 x 3.5 cm), with areas of interior calcification. The patient underwent surgical treatment, with wide resection of the tumor, comprising 9th, 10th and 11th ribs and a portion of the diaphragm. The definitive diagnosis of giant cell tumor was made by immunohistochemistry of the surgical specimen. This report provides a basis for studying the therapeutic possibilities of this pathology in atypical locations, reinforcing that the block excision with wide margins of these tumors promotes a lower chance of local recurrence.

Keywords: Giant cells, Ribs, Bone neoplasms, Thoracic wall, Thoracic surgery.
INTRODUCTION

Giant Cell Tumor (GCT) is a benign neoplasm of mesenchymal origin representing 20% of benign bone tumors and 5% of all primary bone tumors.\(^1\text{-}^4\) The incidence is higher in young adults, especially in the third and fourth decades of life, with prevalence in females.\(^5\) It occurs more frequently in the epiphyses of long bones and rarely affects the ribs (approximately 1% of cases).\(^6\text{-}^7\) The presentation of GCT in the anterolateral portion of the rib is even rarer compared to the posterior arch and the costal tubercle.\(^2\text{-}^3\text{,}^4\text{-}^7\)

Although considered a benign neoplasm, giant cell tumors may present locally aggressive behavior, with the invasion of adjacent soft tissues, high recurrence rates after treatment, and, in rare cases (2%), distant metastases, usually pulmonary.\(^1\text{-}^2\text{,}^5\text{-}^7\) When present, lung metastasis from the GCT presents indolent histological characteristics, similar to those of the primary GCT, and may even regress spontaneously.\(^1\text{-}^3\text{,}^5\text{-}^7\) Although it is often confused with malignant neoplasms due to its aggressiveness local, the GCT rarely undergoes malignant transformation.\(^1\text{-}^3\text{,}^5\text{-}^6\)

In this report, we describe a rare case of GCT in the anterolateral portion of the rib, approved by the Ethics Committee for Research with Human Beings HU-UFJF under Opinion nº. 4,341,895, whose diagnosis was obtained through anatomopathological and immunohistochemical analysis after resection tumor surgical procedure with oncological safety margins. This study adds to the literature information about the presentation of the GCT in the anterolateral portion of the rib, considering the rarity of the case and the small number of publications on the subject. The aim of this study is to provide a basis for the study of therapeutic possibilities for aggressive rib GCT, reinforcing surgical resection in a bloc with wide margins as a safer approach to prevent recurrence of neoplasia, distant metastases and malignant transformation.\(^3\text{-}^6\text{,}^7\)

CASE REPORT

Female patient, 38 years old, smoker (approximately 10 pack-years), seen at the Thoracic Surgery Clinic of the University Hospital of the Federal University of Juiz de Fora, with a report of bulging in the left chest wall, with growth for approximately 2 years, associated with pain, which limited some postures. Denied fever or weight loss. On physical examination, he presented a tumor of fibroelastic consistency and adhered to deep planes in the lateral wall of the left hemithorax, painful on palpation.

A chest X-ray (X-ray) showed a lytic and eccentric bone lesion in the 10th left costal arch. Chest Computed Tomography (CT) (Figure 1) revealed the presence of a lateral insufflative lesion in the 10th left costal arch, measuring 4.2 x 3.5 cm, with areas of interior calcification. Bone scintigraphy (Figure 2) showed osteogenic activity in the 10th left costal arch, suggesting a primary lesion. Laboratory tests unchanged.

In view of the findings, we opted for surgical treatment, with wide resection of the tumor, encompassing the 9th, 10th and 11th left ribs and part of the diaphragm (Figure 3.A), which was invaded by the lesion. Chest wall repair was performed with muscle suture, without the need for a prosthesis (Figure 3.B). The postoperative period was uneventful, and the patient was discharged within 4 days.

The Surgical Piece (Figure 4) was sent for the Anatomopathological examination, which revealed a proliferation of ovoid cells with minimal atypia, permeated by numerous osteoclast-like multinucleated giant cells, characteristics suggestive of Giant Cell Tumor (GCT) Bone. The diagnostic confirmation of GCT was given by Immunohistochemistry, by evidencing the presence of Histone H3.3 G34W and SATB2 Antibodies in Stromal cells and CD68 Antibody in multinucleated cells. The patient remains under outpatient follow-up, with no evidence of disease recurrence after 12 months.
Figure 2. Bone scintigraphy revealing osteogenic activity in the 10th left costal arch (yellow arrows).

Figure 3. A: Thoracotomy showing part of the diaphragm resected (green empty arrows), after wide resection of the tumor (solid green arrows). B: Chest wall repair, with diaphragm reconstruction and muscle suturing (yellow arrows).

Figure 4. Surgical specimen. A: Portion outside the rib cage (yellow arrows). B: Intrathoracic portion (green arrows).
DISCUSSION

Giant Cell Tumor (GCT) is a benign but aggressive osteolytic skeletal neoplasm, richly vascularized and with numerous giant cells with osteoclastic activity. In 90% of cases, it has a typical epiphyseal location in long bones, mainly, distal femur, proximal tibia and distal radius. Due to the rarity of GCT in atypical topography, its definitive diagnosis becomes extremely difficult to be performed before a therapeutic approach.

According to the literature, complementary exams such as Simple Radiography (RX), Computed Tomography (CT) and Magnetic Resonance (MRI) show a lytic bone lesion, eccentric, with cortical thinning or even cortical rupture with extension to the adjacent soft tissues. A presence of matrix calcification is rare. The chest X-ray and chest CT of this patient showed an insufflative lateral bone lesion in the 10th left costal arch with areas of calcification in the matrix. The bone scintigraphy finding was compatible with reports in the literature, in which the GCT presents local hyperuptake due to the increase in the reactive osteoblastic activity.

Recent studies have suggested the use of serum acid phosphatase (AcP) as a useful marker for diagnosis of Giant Cell Tumor (GCT) and patient follow-up. According to Goto et al., its elevation is due to secretion or release from the giant cells of the tumor.

Differential diagnosis should include Aneurysmal Bone Cysts, Non-exudative Fibroma, Osteosarcoma, Chondroblastoma, Brown Tumors associated with Hyperparathyroidism and Metastatic Cancer. The differential diagnosis was not obtained preoperatively due to the low specificity presented in aspiration punctures in bone wall tumors thoracic.

Giant cell tumors should be classified according to the Campanacci and Enneking classifications. Our patient was classified as Campanacci Grade III (When it courses with extraosseous lesions that rupture the cortex and extend to the adjacent soft tissue) and Enneking Stage III (When it progresses with the presence of highly vascularized and aggressive tumors), due to cortical rupture and extension to adjacent soft tissues. The gold standard of treatment for patients with this classification is aggressive surgical resection (en bloc) of the lesion with wide margins of oncological safety, involving the bone and adjacent soft tissue, as performed. Several studies suggest that this procedure considerably reduces the risk of local recurrence compared to intralesional curettage associated with cementation, treatment of GCTs in typical topographies and classifications I or II by Campanacci and by Enneking. In this case, a portion of the diaphragm that presented tumor invasion was resected. The diaphragm was reconstructed by suturing with PDS 0 thread, without the need for prostheses.

Radiotherapy treatment for GCT is still very controversial. There are reports that most of the malignant transformations existing in the GCT are associated with previous radiotherapy. For patients with unresectable and locally recurrent disease, options include Denosumab, radiotherapy or arterial embolization. It is noteworthy that Denosumab as an adjuvant in other cases is still debatable, deserving further confirmatory studies.

CONCLUSION

This report allows us to broaden the literary bases about the main therapeutic modality applied to the Bone Giant Cell Tumor, by reinforcing that block excision with wide margins, considered the gold standard in Giant Cell Tumors in typical locations, also promotes a lower chance of local recurrence when in atypical locations, evidenced by the absence of recurrence in the patient’s clinical follow-up.

BIBLIOGRAPHIC REFERENCES


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