Case Report

Giant chondrosarcoma of the sternal body: case report

Condrossarcoma gigante de corpo esternal: relato de caso

JOSÉ DALVO MAIA NETO[1]*
ANTERO GOMES NETO[1]
RYAN NOGUEIRA LIMA[1]
MARIA DE FATIMA DE BRITO COUTINHO NOGUEIRA LIMA[1]

■ ABSTRACT

Chest wall tumors are relatively uncommon, representing 1 to 2% of all neoplasms, and approximately 5% of thoracic neoplasms. Sarcomas are rare tumors with heterogeneous presentation. They are divided into two large groups: bone and soft tissue tumors. Occurrence in the chest wall is rare, representing 8% of all sarcomas. The treatment of choice is resection with wide margins and, when invasion of deep structures occurs, thoracotomy is indicated. The reconstruction of the bone framework is carried out with synthetic materials, providing stability, and is followed by the reconstruction of soft tissues with muscular, musculocutaneous, or random local flaps. Primary malignant tumors of the chest wall account for less than 1% of all neoplasms and include a wide variety of bone and soft tissue lesions. Chondrosarcomas represent 20% of primary tumors of the chest wall, with 80% originating in the ribs and 20% in the sternum.

Keywords: Chondrosarcoma; Sternum; Thoracic wall; Thoracic neoplasms. Myocutaneous flap.

■ RESUMO

Os tumores de parede torácicas são relativamente inusitados, representando 1 a 2% de todas as neoplasias, e cerca de 5% das neoplasias torácicas. Sarcomas são tumores raros e com apresentação heterogênea. São divididos em dois grandes grupos: tumores ósseos e de partes moles. A ocorrência na parede torácica é rara, representando 8% de todos os sarcomas. O tratamento de escolha é a ressecção com margens ampliadas e, quando ocorre a invasão de estruturas profundas, a toracectomia é indicada. A reconstrução do arcabouço ósseo é realizada com materiais de síntese, proporcionando estabilidade, e é seguida da reconstrução de partes moles com retalhos musculares, musculocutâneos ou locais ao acaso. Os tumores malignos primários da parede torácica correspondem a menos de 1% de todas as neoplasias e incluem grande variedade de lesões ósseas e de tecidos moles. Os condrossarcomas representam 20% dos tumores primários da parede torácica, sendo que 80% têm origem nas costelas e 20% no esterno.

Descritores: Condrossarcoma; Esterno; Parede torácica; Neoplasias torácicas; Retalho miocutâneo.

Institution: Hospital de Messejana, Fortaleza, CE, Brazil.

Article received: October 16, 2023.
Article accepted: April 30, 2024.

Conflicts of interest: none.

DOI: 10.5935/2177-1235.2024RBCP0886-EN

INTRODUCTION

Chest wall tumors are relatively uncommon, representing 1 to 2% of all neoplasms and approximately 5% of thoracic neoplasms. Primary tumors of the chest wall are rare and include a large group of neoplasms that can arise not only from bones or cartilage of the chest wall but also from associated subcutaneous tissue of muscles and blood vessels[1].

Primary malignant tumors of the chest wall account for less than 1% of all neoplasms and include a wide variety of bone and soft tissue lesions. Chondrosarcomas represent 20% of primary tumors of the chest wall, with 80% originating in the ribs and 20% in the sternum. Occurrence in the chest wall is rare, representing 8% of all sarcomas[1]. The treatment of choice is resection with wide margins and, when

[1] Hospital de Messejana, Fortaleza, CE, Brazil.

invasion of deep structures occurs, thoracotomy is indicated[2-4].

This study aimed to describe the clinical presentation, method of preoperative planning and surgery, and perioperative management of giant sternal chondrosarcoma. Sternectomy to treat malignant tumors results in large defects in the bone framework and soft tissues, causing deformity and paradoxical movement of the chest wall and making thoracic reconstruction very important[5]. The reconstruction of the bone framework is performed with synthetic materials providing stability and is followed by the reconstruction of soft tissues with well-vascularized muscular, musculocutaneous, or random local flaps[5-11].

CASE REPORT

Male, 53 years old, born and living in Fortaleza (CE), admitted to the thoracic surgery service at Hospital de Messejana, in 2021. Six months ago he presented with a bulging sternum (Figure 1) with mild local pain and a progressive increase in the lesion. He developed dyspnea on exertion over the last month. He reported weight loss of around 10 kg in the last six months.

On physical examination, he was in good general condition, anicteric, acyanotic, afebrile, eucardial, and eupneic. Pulmonary and cardiac auscultation was normal, and abdominal palpation was innocent. From complementary exams, the chest tomography (Figure 2) showed an expansive and septate formation compromising the thoracic wall and with significant involvement of the sternal body, with foci of osteolysis, measuring 15.3 x 13.8cm, insinuating itself into the region intrathoracic and determining significant posterior rejection of the heart, without signs of pericardial invasion.

A bone scintigraphy was performed, which demonstrated diffuse irregularity in the uptake of the drug in the sternum. The echocardiogram demonstrated a mediastinal mass compressing the right heart cavities, mild pericardial effusion, and an ejection fraction of 70%. An incisional biopsy of the lesion was performed, the anatomopathological examination of which revealed a low-grade cartilaginous neoplasm, suggestive of osteochondroma, which could not be completely distinguished from grade 1 chondrosarcoma.

The patient underwent surgery, with complete resection of the tumor (Figure 3) by partial sternectomy en bloc with cartilage from the 3rd to 10th costal arches, and preservation of the manubrium (Figure 4). Reconstruction of the chest wall was performed with double polypropylene mesh (Figure 5) and bilateral

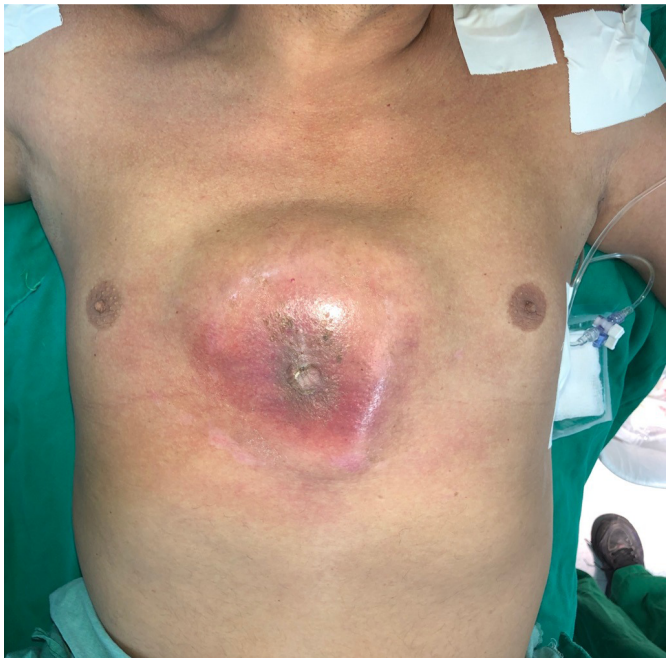

Figure 1. Initial presentation of the patient.

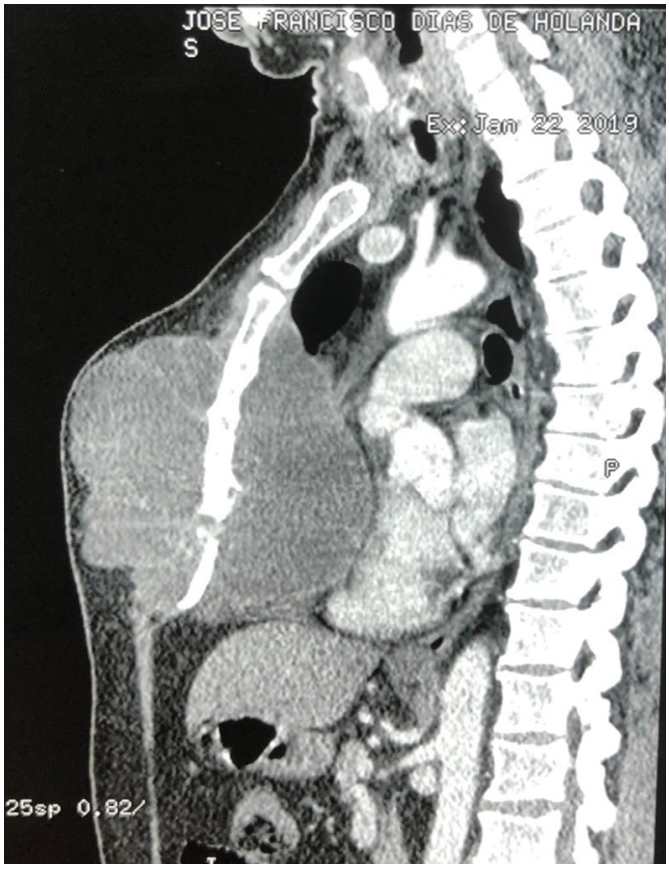

Figure 2. Tomography of the tumor lesion with mediastinal invasion preoperatively.

pectoral flaps as described by Starzynski for the correction of large defects in the chest wall, with the advent of the plastic surgery service of the reference hospital. The final histopathological diagnosis was grade II chondrosarcoma, with a microscopically compromised margin.

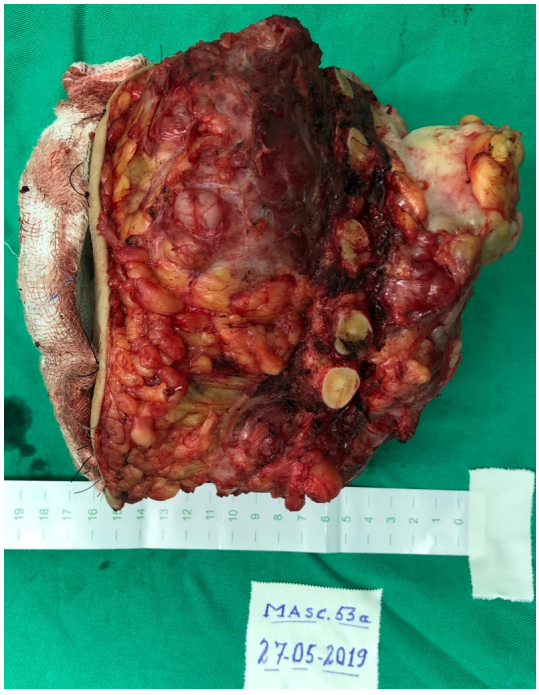

Figure 3. Resected tumor lesion, aspect upon referral to histopathology.

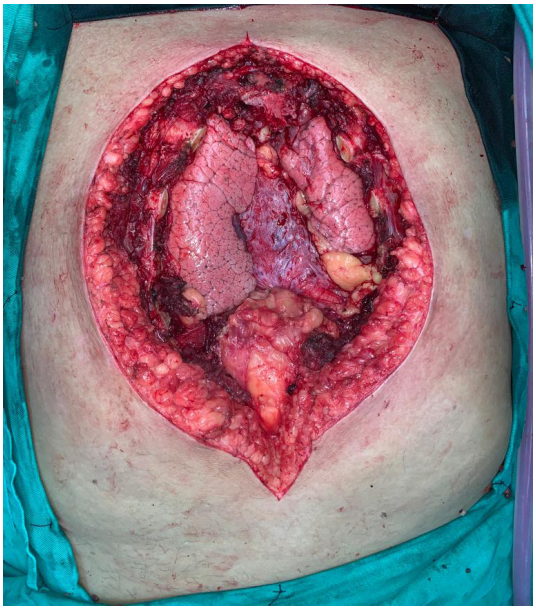

Figure 4. Intraoperative image after removal of tumor mass showing exposed mediastinum.

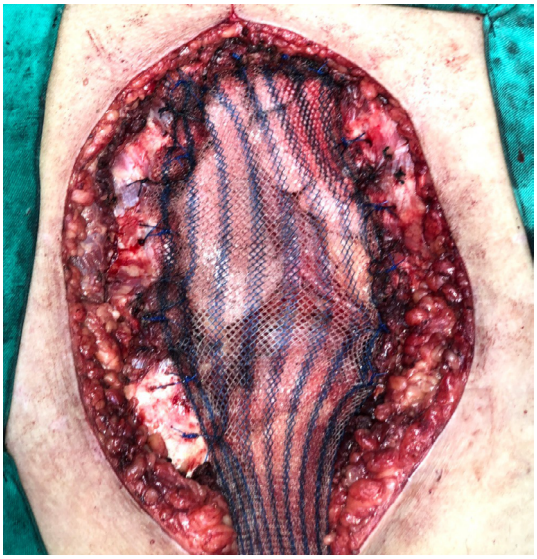

Figure 5. Marlex mesh used as a form of additional protection to the mediastinum.

The patient underwent adjuvant chemotherapy and radiotherapy and evolved without signs of tumor recurrence until the 9th-month post-surgery, when he underwent a chest computed tomography scan which showed only findings compatible with surgical manipulation, thickening of the subcutaneous tissue, and skin retraction. In this case, reconstruction of the chest wall without the use of bone cement or metallic prostheses is noteworthy, with good oncological, aesthetic, and functional results (Figure 5). However, it is known that primary bone chondrosarcomas, hyaline cartilage neoplasms, tend to show slow progression and high recurrence, especially when surgical resection margins are not adequate, measuring at least four centimeters on all sides.

DISCUSSION

Chondrosarcomas represent approximately 30% of primary malignant bone neoplasms, the most common being those of the anterior chest wall. This tumor most commonly occurs between the third and fourth decades of life and is relatively uncommon in people under 20 years of age. Males are more affected[3]. Chondrosarcomas are lobulated neoplasms that can grow to massive proportions and, consequently, can extend internally into the pleural space or externally, invading muscle and adipose tissue of the chest wall. Microscopically, findings range from normal cartilage to obvious malignant changes.

Differentiating between chondroma and chondrosarcoma can be extremely difficult[3]. A palpable

mass in the chest is the main symptom in approximately 80% of patients with a chest wall tumor. Of these, 60% have associated pain[2]. Respiratory failure and hemothorax are rarer findings and are only present when the tumors are very extensive[4].

Imaging exams can be useful in suggesting the condition; however, a definitive diagnosis requires a correlation between histology and radiology. Computed tomography (CT) and magnetic resonance imaging (MRI) are good tests to characterize the tumor and its extent. CT is superior to MRI for demonstrating calcifications, while MRI is the exam of choice for evaluating the extent of the tumor and its relationships with adjacent structures[4].

Chest wall chondrosarcomas typically grow slowly and recur locally. If left untreated, late metastases will occur. Complete control of the primary neoplasia is the main determinant of survival. The objective of the first surgery should be a resection wide enough to prevent local recurrence. This means getting a 4cm margin on all sides. This approach results in the cure of approximately all patients, resulting in a 10-year survival rate of 97%[3].

Some authors propose that, as preoperative histological diagnosis is difficult, wide resection should be performed in all cases of chest wall neoplasia[5]. Reconstruction of defects in the thoracic costal framework and preservation of the manubrium. The patient presented intraoperatively with the following defect in the rib cage (Figure 4). The reconstruction of the chest wall was performed with double polypropylene mesh (Figure 5).

The reconstruction was defined with the plastic surgery service of the Hospital de Messejana, where the following therapeutic options were postulated using the algorithm proposed by the service concerning the reconstruction of the thoracic framework:

1) To reconstruct the chest wall, we perform a muscle flap associated with skin grafting (pectoralis major muscle); 2) Myocutaneous flaps: transverse rectus abdominis muscle (TRAM) 3) vertical rectus abdominis muscle (VRAM); 4) Association of TRAM with VRAM; 5) Latissimus dorsi muscle; 6) Fasciocutaneous flaps from the region and bilateral pectoral flaps as described by Starzynski to correct large defects in the chest wall with the advent of the plastic surgery service of the reference hospital; 7) Free flaps.

Due to the degree of vascular compromise in the surgical resection, which required the interruption of bilateral blood flow from the internal mammary arteries, an arc of rotation with the latissimus dorsi muscle was not achieved. The fasciocutaneous flap proposed by Starzynski (RASP- Rotation-advancement split pectoralis) was suggested, a musculocutaneous flap based on the pectoralis major muscle (type 5 Mates-Nahai), with bilateral ligation of the dominant pedicles (thoracoacromial) and using the somersault flap. based on its vascularization with intercostal accessory pedicles to cover the thoracic framework defect at the sternal level (external absent and its topography covered with polypropylene mesh).

Coverage of the skin defect was based on a wide sliding fasciocutaneous flap. The fasciocutaneous flap was anchored with adhesion points described by Baroudi (Figure 6) and the patient developed mild epidermolysis in the part downstream of the bilateral blood supply (edges of the flap in the central region), which was treated conservatively, excellent evolution was achieved (Figure 7).

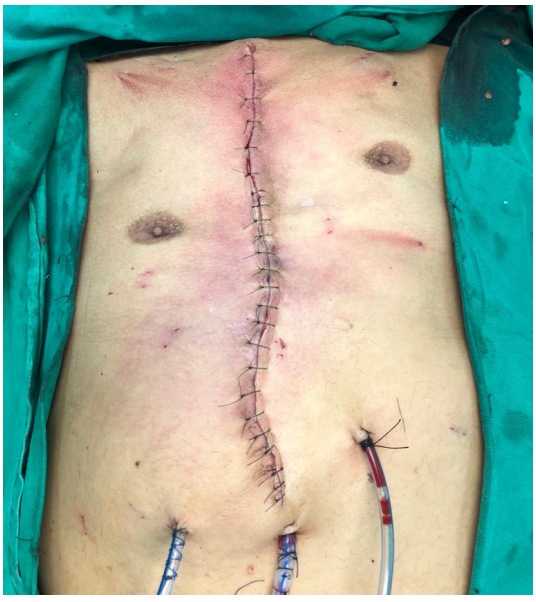

Figure 6. Final appearance after reconstruction with Marlex mesh plus muscle flaps and skin flaps anchored by Baroudi stitches.

The final histopathological diagnosis was grade II chondrosarcoma, with a microscopically compromised margin. The patient underwent adjuvant chemotherapy and radiotherapy and evolved without signs of tumor recurrence until the 9th month post-surgery, when he underwent a chest computed tomography scan which only showed findings compatible with surgical manipulation, and thickening of the subcutaneous tissue.

CONCLUSION

Through the progressive presentation of the case when he sought the thoracic surgery service in Messejana, Fortaleza-CE, the necessary work-up was performed to stage the tumor lesion following

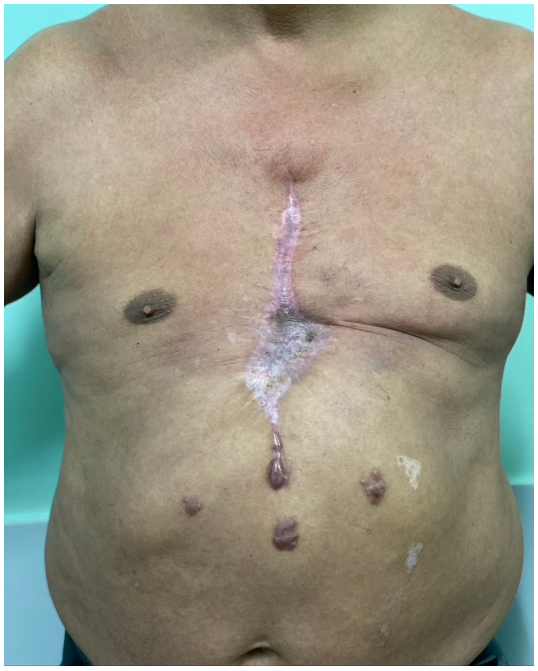

Figure 7. Final appearance after surgical procedure.

the precepts of the existing literature. The patient's evolution was satisfactory and we obtained a survival result with an important quality of life, according to the international literature consulted. It was possible to return the patient to his home activities with stipulated monitoring to detect possible tumor recurrences through imaging exams on predetermined dates.

The result obtained, as well as the surgical proposal given the clinical picture presented by the patient, corresponds to a challenge given the size of the lesion, the need for free margins, and the topography of the lesion, but achieved success despite adversities.

COLLABORATIONS

JDMN Data Curation.

AGN Realization of operations and/or trials.

RNL Realization of operations and/or trials.

MFBCNL Realization of operations and/or trials.

REFERENCES

1. Gonfiotti A, Salvicchi A, Voltolini L. Chest-Wall Tumors and Surgical Techniques: State-of-the-Art and Our Institutional Experience. J Clin Med. 2022;11(19):5516. DOI: 10.3390/jcm11195516
2. Walsh GL, Davis BM, Swisher SG, Vaporciyan AA, Smythe WR, Willis-Merriman K, et al. A single-institutional, multidisciplinary approach to primary sarcomas involving the chest wall requiring full-thickness resections. J Thorac Cardiovasc Surg. 2001;121(1):48-60. DOI: 10.1067/mtc.2001.111381
3. Rascoe PA, Reznik SI, Smythe WR. Chondrosarcoma of the thorax. Sarcoma. 2011;2011:342879. DOI: 10.1155/2011/342879
4. Wang L, Yan X, Zhao J, Chen C, Chen C, Chen J, et al. Expert consensus on resection of chest wall tumors and chest wall reconstruction. Transl Lung Cancer Res. 2021;10(11):4057-83. DOI: 10.21037/tlcr-21-935
5. Dai Z, Maihemuti M, Sun Y, Jiang R. Resection and reconstruction of huge tumors in the chest wall. J Cardiothorac Surg. 2022;17(1):116. DOI: 10.1186/s13019-022-01877-9
6. Isaac KV, Elzinga K, Buchel EW. The Best of Chest Wall Reconstruction: Principles and Clinical Application for Complex Oncologic and Sternal Defects. Plast Reconstr Surg. 2022;149(3):547e-62e. DOI: 10.1097/PRS.0000000000008882
7. Sebayang ANO. Management of Chest Wall Tumors with Tahalele's Method: Review Article. J Heal Study Med. 2020;2:109-16. DOI: 10.36145/JHSM2020.14
8. Crowley TP, Atkinson K, Bayliss CD, Barnard S, Milner RH, Ragbir M. The surgical management of sarcomas of the chest wall: A 13-year single institution experience. J Plast Reconstr Aesthet Surg. 2020;73(8):1448-55. DOI: 10.1016/j.bjps.2020.02.036
9. Çitak N, Çelikten A, Metin M, Pekçolaklar A, Gürses A. Radical resection of a giant recurrent chondrosarcoma of the anterior chest wall. Gen Thorac Cardiovasc Surg. 2011;59(7):512-4.
10. Stanić V, Vulović T, Novaković M, Ristanović A, Stamenović D, Cvijanović V, et al. Radical resection of giant chondrosarcoma of the anterior chest wall. Vojnosanit Pregl. 2008;65(1):64-8.
11. Arnold PG, Pairolero PC. Chest-wall reconstruction: an account of 500 consecutive patients. Plast Reconstr Surg. 1996;98(5):804-10. DOI: 10.1097/00006534-199610000-00008

***Corresponding author:** **José Dalvo Maia Neto**
Avenida Pintor Antonio Bandeira, 1500, ap 1001, Vicente Pizon, Fortaleza, CE, Brazil
Zip Code: 60182-292
E-mail: dalvomaia@hotmail.com