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CLINICAL CASE

Giant chondrosarcoma of the costal cage in an elderly female: case report

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Abstract

A 75-year-old female presented with progressive increase in volume in the region of the costal cage of right hemithorax. Physical examination: Tumor of 30 × 20 cm depending on the 8th-9th costal cartilages, indurated, adherent to deep planes, slightly mobile. Tomography: Tumor extending to skin, displacing right lung, infiltrating diaphragm and liver. In bloc resection and diaphragm plasty were performed. The post-operative evolution was unfavorable. Chondrosarcoma of rib is a very rare tumor, also this case is important because describes an atypical presentation of the disease and we conclude that chest wall reconstruction is utmost importance for adequate evolution.

Keywords: *Chondrosarcomas. Sarcoma. Bone neoplasm. Surgical mesh. Case report.*

Condrosarcoma gigante de caja costal en una mujer anciana: reporte de caso

Resumen

Mujer de 75 años, presenta aumento progresivo de volumen en región de caja costal, hemitórax derecho. Exploración física: Tumor de 30 × 20 cm dependiente de cartílagos costales 8-9°, indurado, adherente a planos profundos, ligeramente móvil. Tomografía: Tumor que se extiende a piel, desplaza pulmón derecho, infiltra diafragma e hígado. Se realizó resección en bloque y plastia de diafragma. Con evolución postoperatoria desfavorable. El condrosarcoma de costilla es un tumor muy raro, además este caso es importante porque describe una presentación atípica de la enfermedad y concluimos que la reconstrucción de la pared torácica es de suma importancia para una evolución adecuada.

Palabras clave: *Condrosarcomas. Sarcoma. Neoplasia de hueso. Malla quirúrgica. Reporte de caso.*

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Introduction

The term chondrosarcoma (CS) is used to describe a heterogeneous group of tumors with diverse morphological characteristics and clinical behaviors that are characterized by the formation of chondroid matrix; it corresponds to the second most frequent primary malignant bone tumor[1]. It most frequently originates in pelvic bones or long bones; it is relatively rare for it to arise in the ribs[2,3]. Patients with CS usually present with an irregular palpable mass, with increased pain and fractures due to bone weakening[4]. Computed axial tomography (CT) with intravenous contrast is the gold standard radiographic study for diagnosis and surgical planning[5,6]. CS has a significant potential for metastasis and are relatively resistant to chemotherapy and radiotherapy. Surgical excision with negative microscopic margins is the treatment of choice[7]. The prognosis for most patients is favorable and correlates with histologic grade and adequate surgical margins[8]. The 10-year survival rate after wide resection is 96.4% compared to 65.4% for those who had only local excision[2]. This case report describes an atypical presentation of a primary chest wall (rib) mass managed by surgical excision and reconstruction. In addition to the fact that the thoracic location of these tumors is quite uncommon, there are few case reports, in the Latin American scientific literature, of CS with the dimensions presented here.

Case report

This is a 75-year-old female patient with a hereditary family history of sister with breast and ovarian cancer, within chronic degenerative history: Diabetes mellitus being treated with Metformin, systemic arterial hypertension being treated with Enalapril, chronic obstructive pulmonary disease (COPD) being treated with Tiotropium Bromide and Salmeterol/Fluticasone; with no other history of importance for current pathology. She came to the oncological surgery service of the High Specialty Medical Unit No. 25 (IMSS), for presenting progressive increase of volume in the costal grill region of the right hemithorax for 5 years of evolution, accompanied by compressive pain that improved with the intake of analgesics, without other systemic affection. Physical examination revealed the presence of a tumor measuring 30 × 20 cm depending on the right costal cartilages 8-9th, indurated, adherent to deep planes, non-displaceable, without visualization of collateral venous network, the rest of the examination was unaltered (Fig. 1).

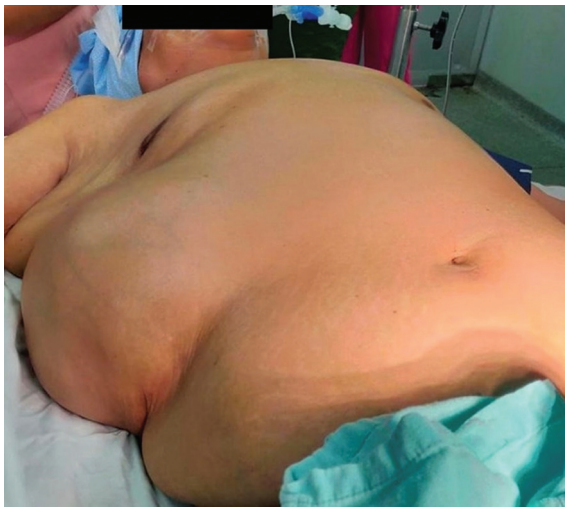

Figure 1. Patient in supine decubitus position. Physical examination revealed a tumor in the right costal region measuring 30 × 20 cm.

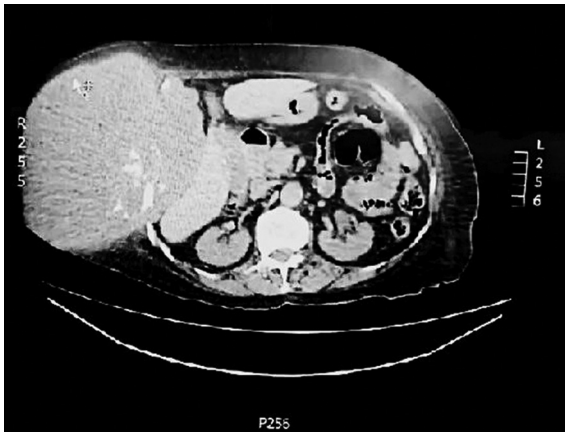

Figure 2. Simple axial computed tomography, axial section. Tumor dependent on the 9th right costal arch of 20 × 20 cm, displacing structures (liver and lung), with areas of calcification in its interior.

Simple CT reported a heterogeneous image of dimensions 14.3 cm × 17.5 cm depending on the right ninth costal arch, producing bone lysis of it, penetrating and displacing the right lower pulmonary lobe and right liver, with apparent loss of the interface, but without infiltrating structures or presenting mediastinal adenopathies (Figs. 2 and 3). The IV contrast CT scan reported a right costal tumor of 15 × 15 × 15 cm extending to the skin, displacing the right lung, infiltrating the diaphragm and suggesting infiltration of hepatic segments

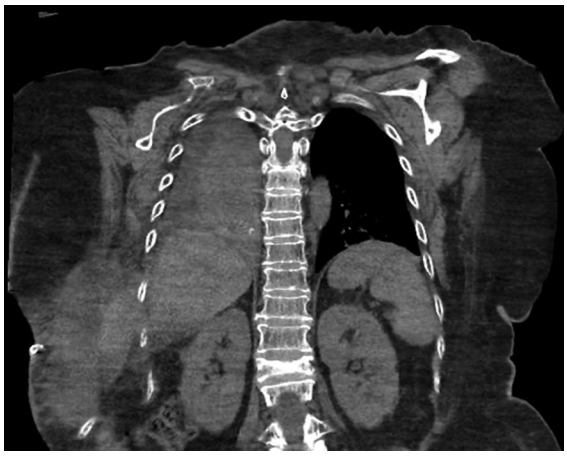

Figure 3. Simple computed axial tomography, coronal section, and thoracic region. Hyperdense area is observed in the right thoracic region, probably due to fluid accumulation.

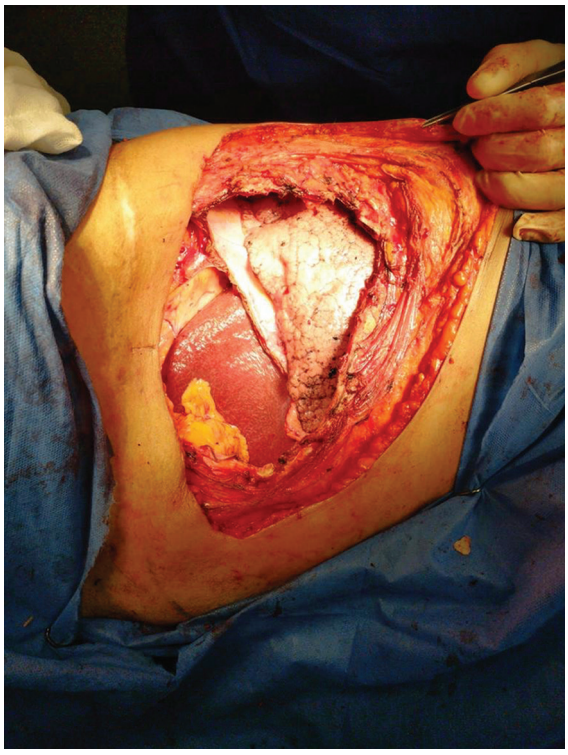

Figure 4. Tissue exposure to the en bloc extraction of the tumor. Absence of 9th and 10th ribs. Lung and liver respected.

5 and 7, encompassing 4 costal arches. Liver and lung without lesions: A percutaneous biopsy was performed, where it was reported: Bone biopsy. Slice 1: CS of the thoracic wall, conventional, central, and primary, grade I, fragments of hyaline cartilaginous tissue, atypical chondrocytes, bone fragment without alterations, no necrosis, and no bone infiltration. Slide 2: Very scanty material and proliferation of atypical chondrocytes.

The following diagnoses were determined: 1. CS grade 2, of the right costal cartilages 8th and 9th; 2. Diabetes mellitus type 2; 3. Systemic arterial hypertension; 4. COPD; and 5. Obesity grade I, the World Health Organization. The differential diagnosis considered was osteosarcoma, which was ruled out by percutaneous biopsy.

The patient underwent wide *en bloc* resection of the tumor of the 8th, 9th, and 10th right ribs, with diaphragm plasty (Fig. 4). The subcutaneous cellular tissue and muscles were dissected circumferentially to the tumor, up to the thoracic wall with visualization of the costal arches and abdominal wall, under direct vision partial resection of the compromised costal arches and diaphragm was performed, a block piece was extracted, an anterior endopleural probe (SEP) 36Fr was placed, and a plasty of the diaphragm insertion was performed (Fig. 5). The diaphragmatic insertion plasty was performed towards the remnant of the costal arches with Vicryl 0, sealing the pleural cavity, pectoralis major, abdominal oblique, and latissimus dorsi muscle flaps were made, two-layer polypropylene mesh was placed in the defect (Sublay), fixed with PDS 1 cardinal mattress stitches and the emergence was anchored on the edge of the defect with PDS 3-0 (Fig. 5). Surgical findings: Tumor 25 cm × 20 cm, exophytic, non-ulcerated, involving abdominal parietal peritoneum, diaphragm, and parietal pleura (Fig. 6). The surgery was successfully completed, and the patient was transferred to the intensive care unit. Scales were used to calculate prognosis, which gave the following results: SOFA: 19pts, estimated mortality 92%; APACHE II: 42pts, estimated mortality 92.2%; and SAPS 3: 142.5pts, estimated mortality 99%. Hence, we knew that the mortality of our patient was very high.

On the fourth postoperative day, hemoglobin levels decreased from 11.9 mg/dL to 9.8 mg/dL and progressive dyspnea appeared, with clinical suspicion of a hemothorax. Therefore, patient was taken to the operating room for thoracotomy. Two-layer mesh was removed in the upper portion, 36-fr PES was placed with an anterior trajectory leading to the posterior and upper pleural cavity, the trajectory of the previous probe was explored, and abundant clots were extracted, packed with gelfoam and mesh was fixed again to the upper

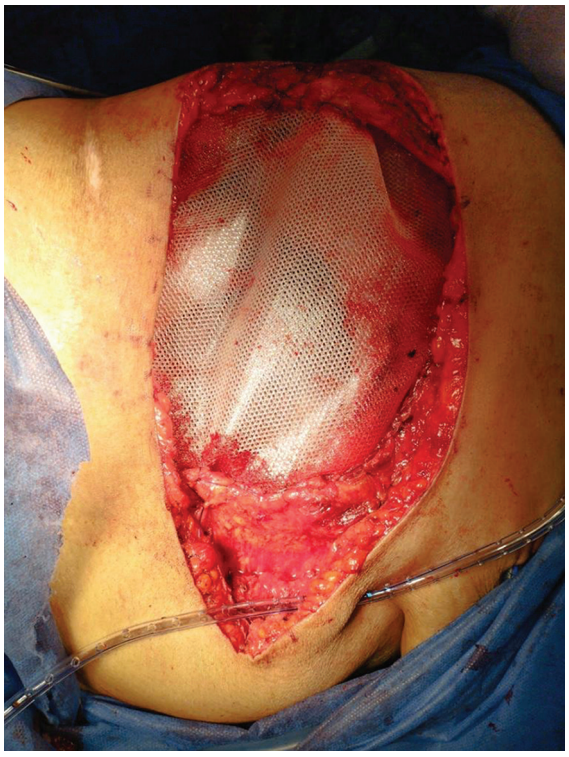

Figure 5. Placement of polypropylene mesh for chest wall reconstruction.

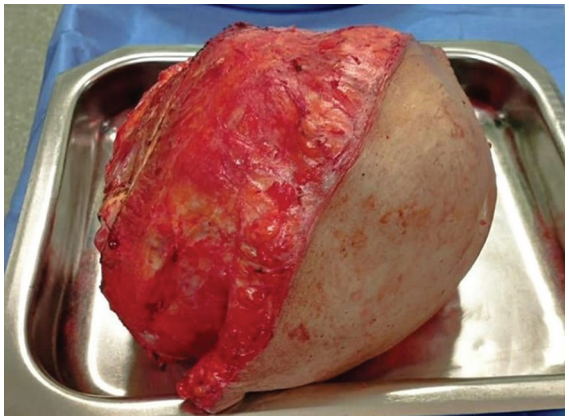

Figure 6. 25 × 20 cm, exophytic, non-ulcerated right costal tumor block.

edge with 2-0 prolene, and ¼ drenovac was placed over the mesh. A total SEP output of 1000 cc is quantified. Subsequently, she was admitted to intensive care, with data of tissue hypoperfusion with lactate >15 mmol/l, delayed capillary filling, under sedation, with vasopressor support to maintain MAP 70, tachycardic (110 bpm), and under programmed mechanical ventilation in CMVS mode PEEP 5, FR19, and Fio2 50%. At 48 h post-surgery she evolved to cardiorespiratory arrest, 4 complete cycles of advanced cardiopulmonary resuscitation were given, with no response. It was concluded that the direct cause of death was hemorrhagic hypovolemic shock.

Finally, the histopathological report of the excised surgical specimen was the following: A whitish tumor of 22.5× 17× 15. 5 cm of multilobulated surface, covered by soft and lobulated light yellow adipose tissue as well as by dark brown and soft muscle bundles, multiple cuts with mucinous orange liquid, heterogeneous parenchyma, whitish areas with calcifications, areas of necrosis and presence of projections of papillary aspect, three ribs are identified which macroscopically are surrounded by the tumor, the cut of the skin spindle macroscopically in contact with subcutaneous cellular tissue. Diagnosis: CS grade 2. Tumor site: Right costal cartilages 8th and 9th (referred in clinical history). Tumor size: 22.5 × 17 cm. Necrosis: Not identified.

The challenges we faced during the evaluation and management of this case were mainly: the search for medical care by the patient, since she came 5 years after the onset of the disease, delaying timely diagnosis and treatment. This could be translated into a high rate of deferral by the first level and poor access to health services. We were able to determine three important challenges for diagnosis and management: (1) lack of primary input (bilayer mesh), at the second level of care, for treatment; (2) delay in the cabinet studies for the complete study protocol, due to lack of supplies for the performance of a CT scan with contrast; and (3) delay in referral to the third level, on the one hand, due to the fact that the patient was a foreigner and on the other hand due to administrative issues.

Discussion

Rib CS is a very rare malignant tumor. It usually presents as a fixed, solid, gradually growing mass, occasionally with concomitant chest pain, which is a poor prognostic sign[9]. The age of presentation is above 50 years of age and predominantly male[10]. The patient in the present case, in comparison, is a woman in her seventh decade of life who presented with mild chest pain. This is an unusual presentation seen in 6.5% of individuals presenting with what appears to be an intrathoracic mass without a pre-existing lesion, suggesting a primary etiology in origin[11,12]. The proliferative behavior is variable, ranging from a slow growing form

with little chance of metastasis, to an aggressive sarcomatous form with a high chance of metastasis. More than 90% of CS are conventional. Approximately 90% of them are low to intermediate grade (grade 1-2) and react indolently and rarely metastasize[8]; only 5-10% are grade 3 and have a high metastatic potential[13]. Chest wall resection and reconstruction procedures require adequate radical resection associated with maintenance of thoracic stability, adequate pulmonary function, and an acceptable cosmetic outcome. Surgical management continues to be accepted as the main treatment for primary malignant tumors of the chest wall, as they are often resistant to chemo- or radiotherapy, that is, the reason why we proceeded to give our patient that management. The goal of appropriate radical surgery is removal of the tumor with a wide disease-free margin along with maintenance of chest wall stability. Inadequate tumor resection is associated with a high incidence of recurrence[14]. Chest wall reconstruction is recommended in case of a full thickness defect of more than 5 cm or anterior resection involving more than three ribs[15], as discussed in the case presentation, the patient underwent tumor resection of the 8th, 9th, and 10th right ribs, making her a candidate for wall reconstruction. Various techniques have been used for chest wall reconstruction, such as mesh, methyl methacrylate cement, flaps, titanium plates, and the creation of a neo-resection. However, in this case, we prefer to use a double-layer polypropylene mesh because of its inert properties, ease of processing, and versatility for conversion into various shapes; it is also inexpensive, readily available, and produces promising results in extensive chest wall reconstruction[16].

The most important complication of wide chest wall resections is chest wall instability and usually the operated patients need post-operative mechanical ventilation, some even warrant prolonged intubation with the risk of nosocomial pneumonia and ventilator-associated pneumonia[17]. Prognosis is rarely described, but we can found as prognostic factors for worse outcome after treatment for primary CS: Tumor size > 5 cm, positive resection margins, and high histological grade of the tumor. Obtaining adequate surgical margins when possible is therefore of vital importance[18]. The 5-year survival rate is 81% for intermediate grade tumors (Grade II)[19]. We can conclude that rib CS is an extremely rare tumor that can also have an unusual presentation, so it is important to have a thorough knowledge of the disease, its prognostic variables such as patients age, location of tumor, histological subtype, grade, and clinical stage to be able to offer the best surgical management in a timely manner. Surgical resection is certainly the current treatment of choice, but chest wall reconstruction is of utmost importance for adequate thoracic stability. Informed consent was obtained for the purpose of this publication.

Conclusion

Rib chondrosarcoma is an extremely rare malignant tumor, often requiring radical surgical resection due to its resistance to chemotherapy and radiotherapy. Adequate tumor removal with wide disease-free margins is essential to reduce recurrence risk, while chest wall reconstruction plays a key role in maintaining thoracic stability and pulmonary function. This case underscores the importance of a thorough understanding of the disease and an individualized surgical approach to achieve optimal oncological and functional result.

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Conflicts of interest

The authors declare that they have no conflicts of interest.

Ethical considerations

Protection of humans and animals. The authors declare that no experiments involving humans or animals were conducted for this research.

Confidentiality, informed consent, and ethical approval. The authors have followed their institution's confidentiality protocols, obtained informed consent from patients, and received approval from the Ethics Committee. The SAGER guidelines were followed according to the nature of the study.

Declaration on the use of artificial intelligence. The authors declare that no generative artificial intelligence was used in the writing of this manuscript.

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