

Rib Osteoblastoma: A Rare Benign Primary Bone Tumor - Clinical Case Report

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Abstract: Objective: To present a clinical case of a rare pathology, to know its aggressive behavior, its limited ability to metastasize, and its therapeutic management, as well as to review the current literature. Clinical Case: A 22-year-old male patient, with no personal or family history of the disease, was found to have a radiating mass at the left posterior 10th rib arch, with a rounded appearance, low-density areas inside, rounded, and well-defined microspheres at the margin, measuring 4.3 cm × 4.3 cm. The surgery was performed through a left posterolateral thoracotomy, which removed nearly two-thirds of the 10th posterior arch of the left rib and with its costal-vertebral disarticulation. Anatomic and pathological studies reported a histological diagnosis of rib osteoblastoma or giant cell bone tumor. Conclusion: This is uncommon and difficult to predict bone tumor in behavior, as almost all bone tumors contain giant cells. It is a primary bone tumor with benign features that invades the local area in an aggressive manner, with a biphasic component of giant cells and a highly vascularized spindle cell component. It often relapses and has limited metastatic ability. Osteoblastoma or giant cell tumor of the bone is a rare tumor located in the ribs, making it even more unusual. However, its prognosis is good, although it can recur and metastasize.

Key words: osteoclastoma; bone tumor; giant cells; neoplasia; rib

1. Introduction

Benign bone tumors represent a diverse set of pathological and clinical entities. They differ greatly in terms of aggressiveness and clinical management, requiring extensive treatment management. Many of these lesions can be treated through pure observation or follow-up without any type of intervention, while others require surgical treatment, which involves complete resection followed by reconstruction. It should be noted that some of these tumor lesions have the potential for malignant transformation, followed by metastasis. These metastases, although seemingly benign histologically, can be fatal [1, 2].

The most common tumors of this type are: chondroblastoma, chondromyxoid fibroma and the aforementioned giant cell tumor where malignant behavior has been described, which are generally intra-compartmental lesions at the beginning, but if allowed to evolve, they quickly invade the soft tissues and become extra-compartmental. However, this type of primary tumor lesion of the thoracic wall represents about 4 - 5% of all primary bone tumors, while benign tumors account for 18.2% [1]. The management is complex and requires detailed research and comprehensive treatment. They are a heterogeneous group of proliferative neo-formations (expansive or not), implanted in the tissues that make up the wall of the thoracic cage, originating both from the osteo-chondro-periosteal tissues and from the skin tissue [1, 2].

The average age of onset for benign chest wall tumors is between the second and third decades of life, while malignant behavior usually occurs in patients after the fourth decade, but rib tumors are usually due to metastatic lesions [3, 4].

Giant cell tumor (GCT) or osteoclast tumor of bone is a rare mesenchymal tumor, accounting for 4% to 5% of primary bone tumors. It is characterized by that bone marrow supports the abnormal growth and proliferation of connective tissue and is composed of mononuclear stromal cells, which are considered as tumor quotas. Mononuclear cells are the precursor somatic cell of osteoclast, and multinucleated cells are osteoclast-like cells, responsible for bone destruction [5].

Giant cell tumors account for 18.2% of benign bone tumors, with 75% of cases affecting people between the ages of 20 and 40. It is rare for people under 10 and over 50 years old. Its incidence rate is higher among women, which is 2:1. It almost always appears in the epiphyseal or metaphyseal region of long bones, damaging the epiphyseal and metaphyseal regions in an eccentric position.

The three most seriously affected parts are: the lower end of the femur, the upper end of the tibia and the distal end of the radius, accounting for 60% - 80% of the published cases, followed by the long bone, rarely in the pelvis, patella, vertebra and bones of the skull. The rib is a rare location, with a incidence rate of less than 1% [6]. In these cases, tumors usually originate from the posterior arch. Injuries are usually isolated and symptoms occur when most of the bones are destroyed, as bone fragility can cause pain, usually indicating injury and exposure to the risk of pathological fractures.

In GCT, characteristic cells are giant multinucleated cells located on the matrix of highly vascularized oval and spindle shaped mesenchymal cells, with thin-walled capillaries and small bleeding areas. It is these mesenchymal elements that seal the histopathological diagnosis of GCT and may exhibit minimal mitosis. This tumor has multiple biological activities, but due to its histological appearance, its invasiveness cannot be predicted [6, 7].

At present, genetically, 80% of individuals with GCT present a cytogenetic abnormality of telomeric association, while half of the cells in the tumor present this abnormality. An increased expression of p53 and alterations in different oncogenes have also been described [7].

The clinical, radiological, and histological characteristics of GCT are similar to other tumors and pseudotumors. The histopathological diagnosis of rib wall tumors should be made through open biopsy to avoid confusion with another similar lesion or giant cell tumor. The best treatment option is tumor mass resection with free edges and then reconstruct the ribs. Although it is a tumor with low malignancy, the likelihood of local recurrence is high (20% - 40%) and the incidence of metastasis is low (2%). Radiologically, it appears as a multicellular radiative transparent "soap bubble" image, with fewer single cell images [7].

CT and MRI can better define the actual range of tumors and help distinguish them from other bone injuries [7, 8].

We present a giant cell tumor in the posterior rib arch in a patient. We show the radiologic findings in radiographs, computed tomography, bone survey, the therapeutic conduct practiced, as well as the histopathologic results, and the updated literature was reviewed.

2. Clinical Case

A 22-year-old male patient from Cabimas Municipality, Zulia State, with no personal or family pathological history, who was mentioned, because in an anterior chest radiographic examination and pre-employment assessments conducted in the workplace, they found that the radius of his left posterior tenth rib was opaque. Therefore, he was sent to the outpatient department of thoracic surgery in our hospital center, where he was evaluated.

The physical examination is conducted within the normal range, therefore supplementary laboratory and imaging examinations, chest CT scan with bone window, bone chest wall reconstruction, and bone examination are required.

Results of complementary tests: hematology, biochemical profile, alkaline phosphatases and alpha fetoprotein within normal ranges. Immunological tests: normal. VDRL and HIV negative.

A simple X-ray image of the anterior and lateral chest, with a circular appearance and a diameter of approximately 5 cm, located at the left posterior tenth rib arch (Figure 1).

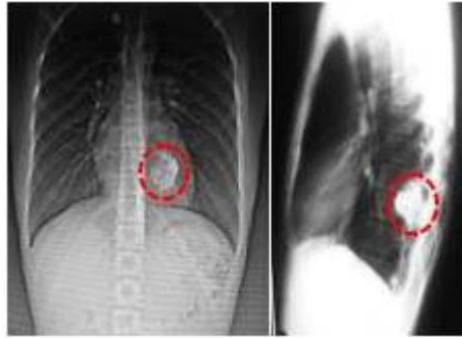


Figure 1. Chest X-ray image: The defect is located in the 10th inner corner of the left rib arch, with a cellular appearance, obvious protrusion of the periosteum, and bone loss (soap bubbles).

3D reconstructed chest CT shows a high-density image at the proximal one-third of the posterior arch of the tenth rib, with a low-density area inside, circular, with clear microsphere like boundaries, and a size of 4.3 cm × 4.3 cm, the lesion protrudes towards the lung parenchyma and dorsal soft tissue, consistent with benign tumor lesions. There is no evidence of lesions in the remaining bone and soft tissue planes (Figure 2).

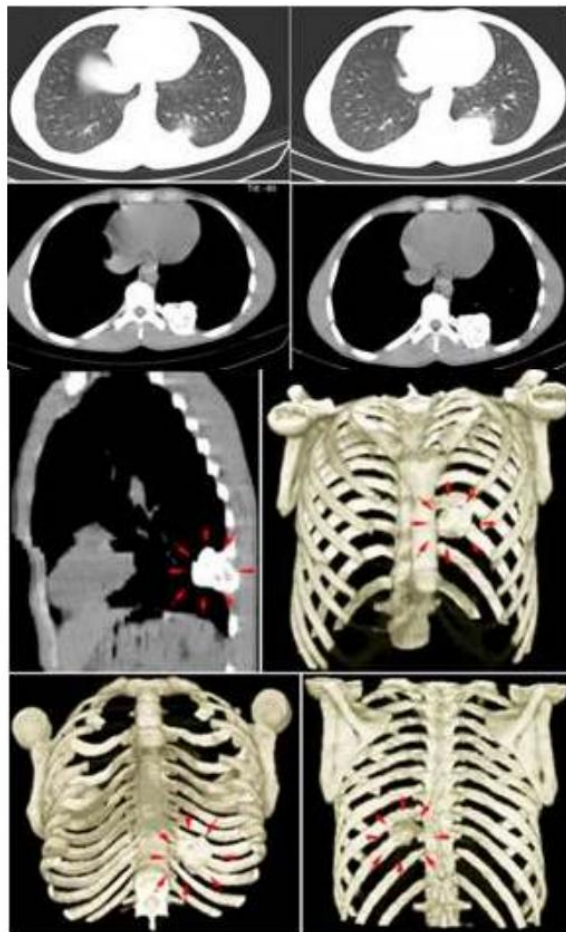


Figure 2. Multi-detector chest CT scan axial section and with 3D reconstruction

Multi-detector chest CT scan axial section and with 3D reconstruction: single space-occupying lesion affecting the 10th left posterior rib arch, high-density with low density areas inside, rounded with well-defined micro-lobulated edges measuring 4.3 cm × 4.3 cm at the level of the proximal one-third, expansive, low-density central portion with osteolytic behavior, with remodeling, thinning and expansion of the cortex (red arrows).

A whole body bone survey and a static chest scan were performed. The body scan showed a space-occupying lesion with an important increase in the concentration activity of the radiotracer projected in the left hemithorax at the level of the left posterior tenth rib arch with significant expansive and deforming activity (Figure 3).

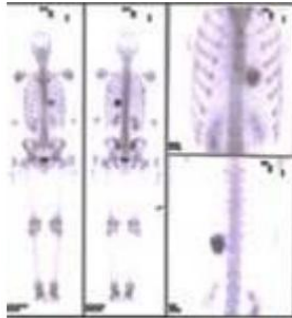


Figure 3. A whole body bone survey and a static chest scan

Bone survey: space-occupying lesion with a significant increase in the concentration activity of the radius-tracer projected in the left hemithorax, at the 10th rib arch left posterior rib arch, with significant expansive and deforming activity.

Due to these findings, it is planned to surgically remove this lesion: by opening the chest on the left side of the plane, entering the chest at the 9th interrib space, surgically removing two-thirds of the proximal end of the 10th left posterior rib arch, and removing its vertebrae. Reconstruction of the chest wall defect was performed with placement of polypropylene monofilament mesh (Marlex®), and mobilization of muscle flap for complete closure of the defect (Figure 4), leaving a chest tube for counter opening through a minimal thoracotomy at the 10th interrib space, posterior axillary line for restitution of the pleural cavity pressure (Figure 5).



Figure 4. Surgical procedure

Left posterolateral thoracotomy was performed at the 9th interrib level, and the proximal two-thirds of the 10th posterior arch was removed and the ribs were stripped. The chest wall defect was reconstructed by placing Marlex® polypropylene monofilament mesh.

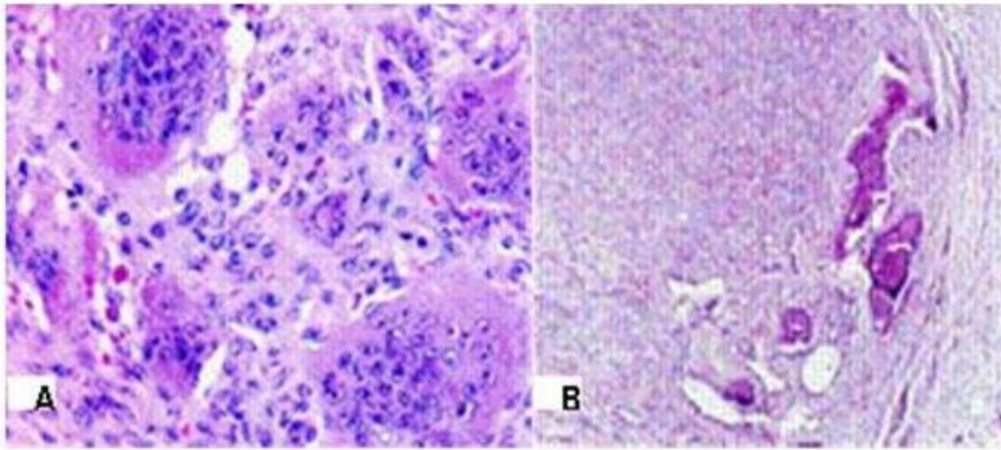


Figure 5. Microphotography

Microphotography. A. Benign giant cell tumor, with evenly distributed giant cells and surrounding mono-nucleated cells. Giant cells are large and have many nuclei, exhibiting the same characteristics as monocytes. B. Dilated growth with cortical endplate erosion of the cortex.

Macroscopic analysis: surgical specimen conformed by the 10th left posterior rib arch, there is evidence of tumor-like formation measuring 4.5 cm × 3.5 cm × 3 cm of circumscribed nodular appearance, grayish with blackish areas to the cut is yellowish gray. Microscopic study shows evidence of neoplasia consisting of two cell types, mononuclear cells with round to oval nuclei, without the presence of mitotic activity and multi-nuclear giant cells with more than fifty nuclei with irregular borders, also, accumulations of foamy macrophages. At the edges of the lesion there is neo-formed bone tissue with osteonecrosis and destruction of the cortical cortex, which corresponds with the diagnosis of giant cell bone tumor of the rib.

Postoperative evolution was satisfactory, for which reason the chest tube was removed and discharge was decided after 72 hours. The patient was cured with the removal of the lesion; however, the patient is monitored by an outpatient clinic to rule out recurrent pathology or the development of multi-systemic forms of the disease.



Figure 6. Postero-anterior chest X-ray: three months after surgery, absence of the 10th left rib arch is observed.

3. Discussion

GCT is one of the rare and less predictable bone neoplasms in terms of its behavior, because virtually all bone tumors contain giant cells.

It is a primary bone neoplasm with benign characteristics, which invades locally in an aggressive manner, with a biphasic component of giant cells and another fusocellular component which is very vascularized, with frequent recurrences and a limited capacity to metastasize [4, 5, 6]. It was first described by Sir Astley Cooper in 1818 [6], which represents 5% - 15% of all benign bone tumors [6, 7]. It occurs in young adults, with a peak incidence between the second and third decades of life, and is slightly more frequent in women [7]. It is typically a monostotic lesion with a predilection for the ends of the bones in the epiphysis and metaphysis of the long bones (knee, femur, tibia, radius), however, almost all the bones of the body have been involved, but localized in the rib area they are very rare [8, 9, 10]. A polyostotic or multicentric form of cases has been observed in less than 1% of cases [11, 12].

There are several theories about its etiopathogenesis: inflammatory, angiogenic and osteoclastic, although none of them has been clearly demonstrated. Recently, the role played by the p53 suppressor gene in its genesis has been confirmed [13].

It usually manifests as a single, asymptomatic or painful tumor with rapid expansive growth, and therefore represents a clinicopathologic and therapeutic challenge due to its variable and unpredictable behavior. The diagnosis is made on the basis of clinical and radiological data, confirmed by histological study. In radiographs, we find a cortical expansion of eccentric localization, with clearly circumscribed margins and without reactive sclerosis. Computed tomography details these features and magnetic resonance imaging is particularly useful to know the integrity of the surrounding soft tissues. This is why Campanacci et al [14] divided these tumors into three stages: stage I: intra-osseous lesion with indolent histology and radiology; stage II: intra-osseous lesion with expansion and cortical thinning but with intact periosteum and benign histology; stage III: extra-osseous lesion with aggressive character but with benign histology. The great majority (70 - 80 %) correspond to stage II, as was our case.

Other studies such as angiography are of great help, showing a hypervascular lesion in most cases (60% - 65%), although there are hypovascular (26% - 30%) or even avascular (10%) cases [5].

It is a neoplasm with malignant potential, especially when it affects older men; however, no clinical, histological and radiographic parameters have been found that can predict its tendency to recur or metastasize, and when it does occur, it occurs in the lung (1% - 10%) [6].

With all these methods, biopsy must be obtained before or within the surgery, and can be guided through a radiation microscope to obtain more accurate samples, which in our case were obtained within the surgery. Under the microscope, GCT is a red tumor of fragile tissue, with a thinning but undamaged cortex. The tumor component is composed of monocytes with osteogenic progenitor cell characteristics and no matrix.

Finally, neither clinical findings nor anatomical and pathological appearance can serve as predictive factors for the behavior of these tumors, as their evolution is variable and unpredictable. As mentioned above, this may lead to local recurrence, distant metastasis, or spontaneous malignant transformation.

In this sense, metastases are more frequent in those patients who have previously suffered local recurrences, and in general, they occur in a period of less than two years.

In terms of treatment, surgery is the preferred treatment method for GCT. There are two types of treatment that can be performed: intratraumatic treatment, including local curettage with or without bone transplantation. Cement is a suitable element for use as a graft because when filling the cavity, it produces a thermal reaction that damages potential residual tumor cells, thereby reducing the risk of recurrence. A more aggressive treatment approach involves marginal excision, enlarged excision, and massive excision with or without reconstruction, as in our case.

Osteoclast tumor or giant cell tumor of bone is a rare tumor. Its position in the rib makes it more unusual, but its prognosis is good. Although it may recur and metastasize when malignant, and may transform into sarcoma, it needs regular research and observation.

In conclusion, we emphasize the importance of good differential diagnosis, enabling us to distinguish this benign pathology from other malignant pathologies, and thus determine the most suitable treatment method based on the patient's symptoms. We must rule out that this is a form of poliomyelitis or a broader syndrome, and consider different treatment options before choosing the most aggressive syndrome.

In many occasions the first diagnostic study corresponds to the simple radiography of the affected area and its findings can be difficult to interpret.

Conflicts of interest

The author declares no conflicts of interest regarding the publication of this paper.

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