

Primary Chondrosarcoma of the Chest Wall Simulation Lung Cancer: A Case Report

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Abstract

Introduction: Chondrosarcoma (CS) is the third most common primary bone malignant tumor; long bones and the pelvis are its usual localization.

Case Report: A 45-year-old man presented to our department with a left lateral thoracic mass. The initial clinical presentation suggested a huge bronchial carcinoma with parietal invasion. Computed tomography showed a dense left anterolateral parietal mass of 19/18/16 cm with areas of necrosis and several calcifications. A CT scan guided biopsy was performed, showing a grade 2 chondrosarcoma. Radiation and chemotherapy with palliative measures were indicated because of the non-operability of the lesion. The patient started the treatment and the immediate follow-up was remarkable by a discreet improvement but then the patient did not complete the rest of his treatment.

Conclusion: Chondrosarcoma is one of the most difficult bone tumors to diagnose and treat. Their management requires a multidisciplinary approach which includes the primary physician, medical oncologist, surgeon, pathologist and radiologist.

Keywords: Chondrosarcoma; Chest Wall Tumors; Diagnosis

Introduction

Chondrosarcoma is a malignant tumor of the bone characterized by a hyaline cartilage differentiation, which can occur either as a primary tumor or a secondary one to an underlying tumor such as enchondroma or osteochondroma. Chondrosarcoma represents 10 to 15% of all primary bone tumors and 20% of all malignant bone tumors [1]. It is the third most common primary malignant tumor of the bone after myeloma and osteosarcoma [2]. The incidence of chondrosarcoma peaks during the sixth and seventh decade of life. Chondrosarcoma has a predilection for trunk bones, which are reportedly involved in nearly 50% of the cases, with rib and pelvis being the most common sites of involvement.

Case Report

A 43 years old male, working as a tiler consulted for a left cervicobrachial neuralgia associated with a left anterolateral chest mass. He had no noticeable medical history but was exposed to passive smoking and silica. The thoracic mass has been increasing in size progressively for the last 6 months. The patient also presented with weight loss, asthenia and anorexia. The physical examination found a huge anterior fixed mass of the upper third of the left hemi-thorax of firm consistency, painless on palpation without any inflammatory signs of the skin, measuring 20 cm on its long axis (Figure 1). A left Horner syndrome was also found and the pleuro-pulmonary examination was normal. There were no palpable peripheral lymph nodes. This clinical presentation suggested an enormous bronchial carcinoma invading the chest wall.

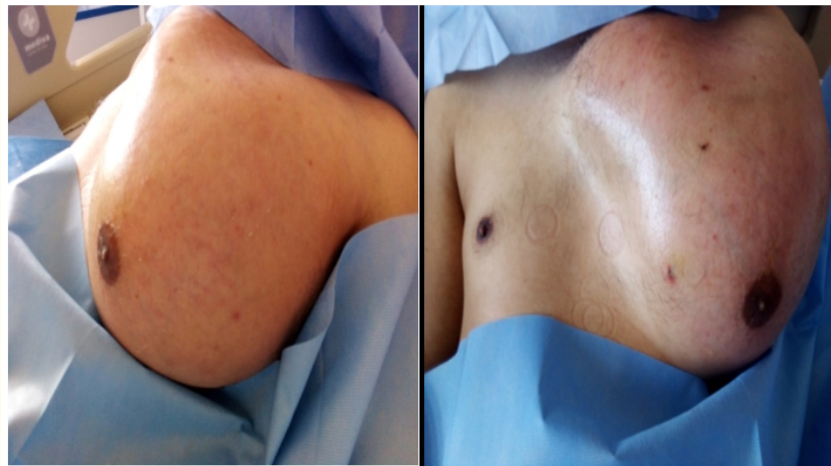


Figure 1: Huge left thoracic mass at the inspection.

The chest X-ray showed a dense, homogeneous triangular opacity extending from the clavicle to the base of the left hemithorax with ascension of the left diaphragmatic dome (Figure 2).

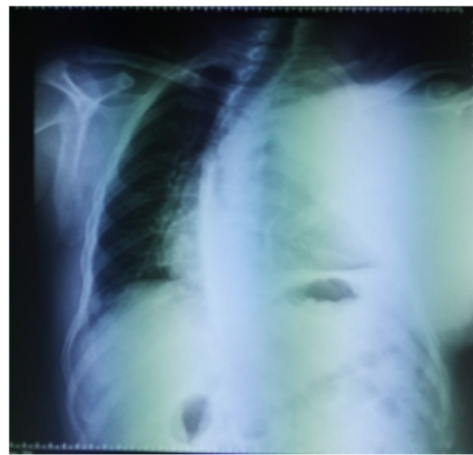


Figure 2: Chest X-ray at the admission.

Thoracic computed tomography showed a hypo dense anterolateral left parietal mass measuring 20/18/16 cm with areas of necrosis and several calcifications with lysis of the first 4 ipsilateral ribs, the sternum and the chondro-sternal junction. The mass invades the subclavian artery and the left carotid. It reaches the pulmonary artery trunk and the left pulmonary artery without a separation interface or mediastinal lymphadenopathy (Figure 3).

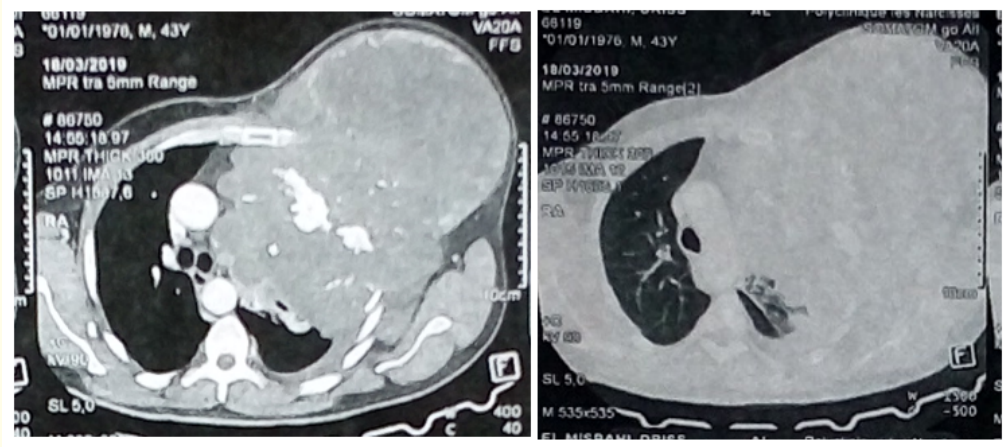


Figure 3: CT scan showing the thoracic mass.

The bronchoscopy found an extrinsic compression of the whole left bronchial tree with narrowed orifices but no visible tumor or granulomas. The pathology of bronchial biopsies and aspirations showed inflammatory bronchial cytology without any signs of malignancy. The Acid fast Bacilli test which was done on bronchial aspirations was negative.

A CT guided transthoracic biopsy was performed without any incidents and its pathology study showed the morphological appearance of a grade 2 chondrosarcoma (Figure 4).

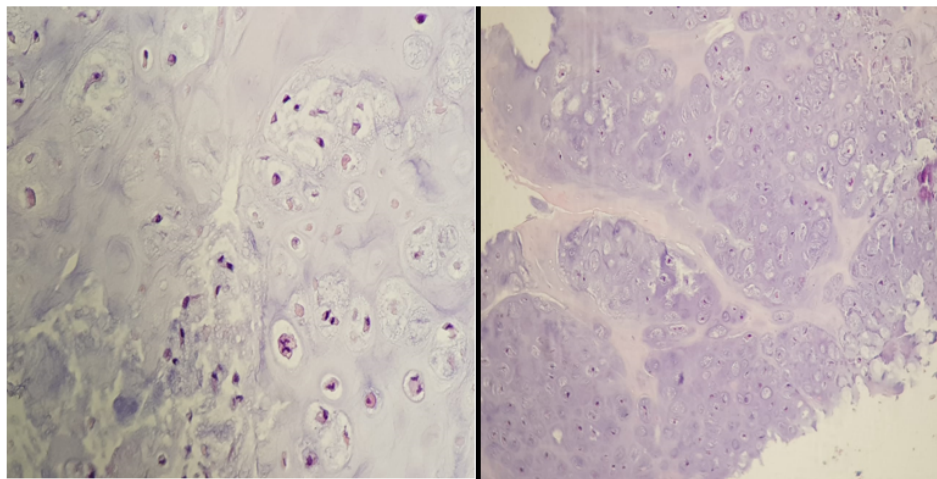


Figure 4: Histological aspect of chondrosarcoma.

In terms of operability, the patient had a WHO Performans Status at 2; the respiratory function exploration showed a small airway involvement with a FEV1 at 1.5 L (48% of the reference value) and a normal cardiac evaluation.

As part of the extension assessment, the medullary Magnetic Resonance Imaging (MRI) found an extension of the tumor to the following medullar nerve roots: C7-D1, D1-D2 and D2-D3, associated with left subclavicular lymphadenopathy, the largest one was 1 cm in diameter. Cerebral and abdominopelvic CT scan did not show any metastasis (Figure 5).

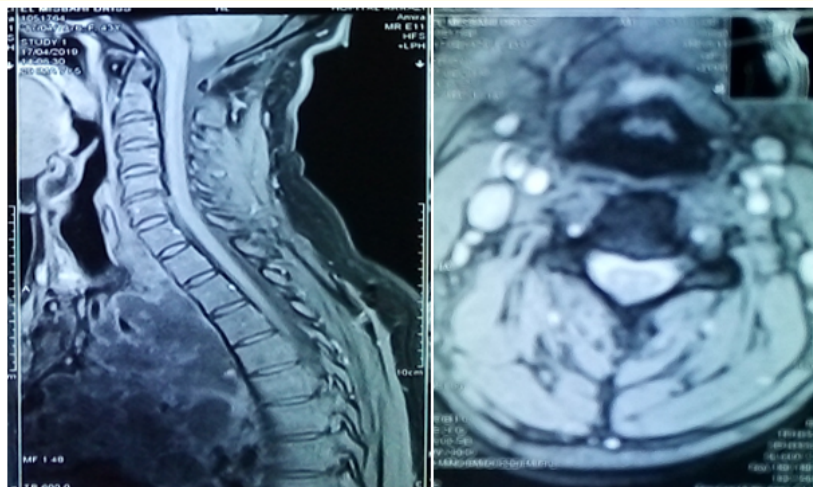


Figure 4: Medullary extension on MRI.

A multidisciplinary consultation meeting was held for a therapeutic decision, the radio-chemotherapy with a palliative care support was chosen because of the non- resectability of the lesion.

Discussion

Primary rib tumors are rare and malignant in 29% of the cases. They are dominated by chondrosarcoma, which represents for 40% of all malignant tumors of ribs [1]. The first five ribs are the more often affected and lateral localization is more frequent than the anterior or posterior one [3].

Rare before the age of 20, chondrosarcoma affects mainly middle-aged men, between the fourth and the fifth decade [4]. Our patient is in his forties which correlates with the epidemiological data. The time between discovery of lesions and consultation is usually long. The slow evolution is not a benignity criterion because this is the usual evolution of these tumors [5]. The CS is most often symptomatic and the main symptom is the pain. Swelling is also a frequent revealing sign which was observed in our patient. Sometimes, a pathological rib fracture, a hemothorax or a metastasis may reveal the diagnosis. The painful and adherent characteristics evoke the malignancy of the tumor. In more than 90% of cases, CSs start spontaneously “de novo” without any pre-existing lesion. They rarely develop on a benign pre-existing lesion such as a chondroma, osteochondroma or Paget’s disease. External thoracic irradiation and trauma might be implicated in the genesis of CS [6].

Imaging plays an important role in the diagnosis and management of thoracic wall tumors. Chest x-rays can guide the diagnosis of ribs CS by showing a parietal opacity, rib swelling, calcifications in “clods” of irregular shapes and distribution as well as rib fractures [7]. The chest CT scan and the MRI provide a better appreciation of the tumor’s localization and extension. The CT scan shows a hypodense, rounded well-defined parietal mass with calcification clusters, which are usually located on the periphery of the tumor forming arcs or rings. The CT scan in our case showed the calcifications at the periphery of the tumor. These calcifications are not constant, rib

fractures and vertebral and soft tissue infiltration can be observed. The thoracic tomography is usually sufficient to decide on a surgical intervention. The MRI, which is less specific than the CT, shows often a parietal mass with well-defined limits which gives an intermediate signal on T1 sequences and a hyper signal on T2 sequences. The tumor appears heterogeneous due to the absence of a signal in the calcifications foci. MRI allows a better assessment of cortical destruction and obsolete reaction [3].

The diagnosis of certainty can only be made on the basis of pathological examination. The removal of the tumor is preferable whenever it is possible. This allows the analysis of the entire lesion and minimizes the risk of recurrence and dissemination [5]. The transthoracic needle biopsy of the tumor often gives a small fragment which is not very representative of the tumor [8]. It is indicated when the removal of the tumor is not immediately feasible or involves a higher risk which was the case for our patient. It allows in these cases the elimination of certain differential diagnosis of which the treatment is not exclusively surgical such as Ewing's sarcoma [5].

Macroscopically, the tumor is greyish, opaque, of soft and gelatinous consistency and sometimes pseudo-cystic. Indeed, cartilaginous plants are often voluminous, coalescent and not very limited with a loss of the lobular aspect characteristic of cartilaginous tumours. Pathologically, CS is a malignant tumour with cartilage differentiation, no bone tumour formation, but foci of reactive or endochondral ossification can be found. According to the World Organization of the health (WHO) classification, chondrosarcomas are subdivided into three grades of increasing aggressiveness based on the cell density, the degree of anisokaryosis and the nuclear hyperchromatism [9]. Grade I has a moderate cell density, the nuclei are hyperchromatic and uniform. The cytological aspect is similar to the one of a benign chondroma and occasionally, some aspects of binucleation are noted. Grade II is characterized by a higher linear cell density. The nuclei are more atypical and hyperchromatic and bi- or multinucleated cells are more frequently observed while mitoses are rare (Figure 2). Grade III is characterized by very dense cells which are pleomorphic and very atypical and the mitoses are frequent.

The spontaneous evolution of the CS leads to local progression, development of mainly pulmonary metastases and ultimately death. Surgery is the only curative treatment [8]. The quality of the surgical procedure is an essential prognosis factor. The resection must be large, taking in mono-blocks the rib carrying the lesion but also the two above and below intercostal spaces, with a safety margin of at least 4 cm. This margin can be reduced in case of a contact with vital organs but should always be bigger than 2 cm. The puncture path of the needle biopsies must also be resected. If closer organs such as the lung and muscles are invaded or adhering to the tumor; then their resection is also necessary [5]. The major resection adds the problem of reconstructive surgery, which must in no way make the surgery palliative and must restore the morphology and physiology of the chest wall. There are many methods to reconstruct the chest wall using autogenous biological material or synthetic material such as polypropylene and polytetrafluoroethylene (PTFE) [6,10].

Radiation and chemotherapy have a limited use in the treatment of Chondrosarcoma. They are indicated in the case of a large tumor which is not immediately resectable in an attempt to reduce the tumor size. In our case, the tumor was very large and extending to the mediastinum as well as the spinal nerve roots making it not resectable. After surgery, these adjuvant treatments have not been shown to be effective [4]. The treatment of recurrences is also surgical [8]. The prognosis of CS is determined by its degree of histological differentiation and the quality of surgical resection. Other prognostic factors are mentioned such as tumor size and the occurrence of metastasis [5]. According to some authors, trunk CSs have a worse prognosis than extremity CSs [11]. This could be explained on one hand, by the larger tumor size of the CS of the trunk at diagnosis, due to their often asymptomatic growth in the rib cage or in the pelvis and on the other hand, by the difficulties of performing a complete surgical resection with safe margins in case of extended CS. Other authors consider that the survival rate and local recurrence rate of thoracic CSs are comparable to those of other CSs localizations, provided that surgical margins are safe [12]. Recurrences and metastases are frequent. Recurrences occur within three years of surgical removal but in 37% of cases, they occur after a longer period of time. Clinical and radiological postoperative monitoring is mandatory every three months for the first three years then every six months for at least ten years, which is the duration for a malignant tumor of the chest wall to be considered as cured [7,10].

Conclusion

The CS is rare tumors, characterized by a high potential for locoregional extension as well as recurrence after treatment and metastatic dissemination. The treatment is based essentially on surgery, which can only be curative in the less extensive and early diagnosed cases. This justifies the monitoring of benign bone lesions and the rigorous follow-up of CSs in order to detect early any recurrence or metastasis.

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