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Mediastinal Chondrosarcoma: Case Report and Review of the Literature

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Abstract **Case Report**

Chondrosarcomas originating primarily in the mediastinal compartment are extremely rare. Unlike other unusual tumors of the mediastinum, chondrosarcomas have been described in both the anterior and posterior mediastinum. The histopathological spectrum described in these mediastinal tumors is similar to that observed in soft tissues. In our current practice, diagnostic imaging plays an essential role in determining the mediastinal origin of any of these tumours. Therefore, in this review, we will outline more specific criteria for the diagnosis of chondrosarcomas, as well as the essential elements of their histopathological evaluation. In addition, we will discuss current knowledge of these entities and their differential diagnosis, which inevitably depends on the anatomical location of the tumor in the anterior or posterior mediastinum. We report a rare case of primary chondrosarcoma of the posterior mediastinum with unusual pathological features, namely the absence of anatomical relationship between the tumor and cartilage-containing organs, and the presence of parietal and pleural metastases

Keywords: Chondrosarcoma, Mediastinum, Metastasis.

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INTRODUCTION

Primary chondrosarcomas arising in the mediastinal compartment are extremely rare. However, cases have been reported in the literature, mainly as small case series or isolated case reports [1-19]. The question of primary cartilage tumours of the mediastinum must be considered in a context similar to that of extra-skeletal chondrosarcomas or soft-tissue osteosarcomas [20-25]. Thus, it is not surprising that such tumors, like those arising in soft tissues, may also originate in the mediastinal compartment.

The criteria for this designation in the mediastinum, as in other anatomical locations, must include the absence of connection of the tumor to any bony structure in the thoracic cavity. Furthermore, in the mediastinal compartment, these criteria can be slightly expanded to include not only the absence of connection with any bony structure, but also the absence of association with any other tumor condition likely to harbor a malignant cartilaginous component.

Within this framework of strict criteria, and thanks to diagnostic imaging techniques and advances in molecular biology, the number of cases that can be considered as truly originating in the mediastinal compartment or authentically representing chondrosarcomas (whether located in the anterior or posterior compartment) is considerably reduced.

It is now recognized that certain mediastinal chondrosarcomas, notably those known as "mesenchymal" or "myxoid", belong to distinct categories and are considered sarcomas of uncertain differentiation or histogenesis. Indeed, specific molecular alterations have been identified in these entities. Nevertheless, although these tumors may be classified as sarcomas of uncertain histogenesis or as "mesenchymal" and/or "myxoid" chondrosarcomas, they remain exceptional occurrences in the mediastinal compartment and will be discussed in this context.

CASE REPORT

A 71-year-old man was admitted with progressive dyspnea for two months. Blood count and biochemistry were normal.

X-rays and a thoracic CT scan (Figure 1) revealed a well-limited lesional process in the posterior mediastinum, with "popcorn" calcifications and heterogeneous enhancement after injection of contrast medium. This process pushes the esophagus forward and the heart forward, coming into intimate contact with the right atrium. Arriving posteriorly in contact with the

prevertebral spaces from D9 to D11, respecting the separation line and without bone extension or lysis. Associated right pleural and parietal masses with the same characteristics.

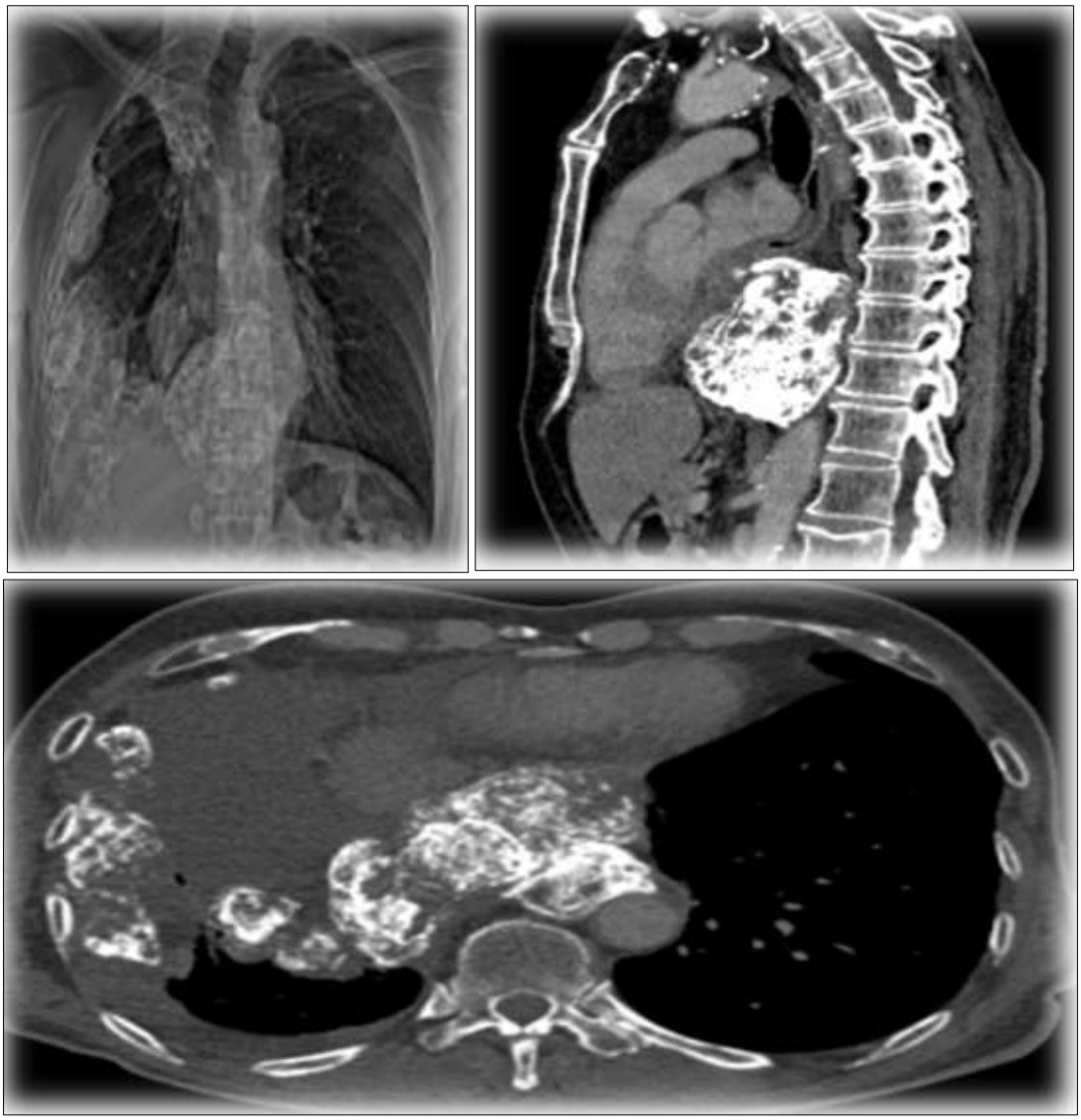

Figure 1: a Front thoracic X-ray, b and c : CT scan in axial and sagittal sections : Heterogeneous posterior mediastinal lesional process with "popcorn" calcifications, associated with right pleural masses with the same characteristics

Histopathological examination showed a bimorphic appearance with basophilic spindle and round cells, as well as a hemangiopericytoma-like vascular pattern and nodular foci of well-differentiated cartilage (Figure 2). The capsule was infiltrated in places. Markers Six months later, a chest CT scan revealed a peripheral pulmonary nodule in the right lung (Figure 1B). Needle biopsy under CT confirmed the diagnosis of chondrosarcoma. The patient refused further treatment. Two years later, a thoracic CT scan showed an increase in nodule volume, prompting a second thoracotomy. The nodule was removed by resection of the middle lobe. Histopathological examination confirmed the chondrosarcomatous nature of the lesion.

Five years after the initial surgery, the patient is healthy and asymptomatic, and the chest CT scan shows no recurrence.

immunohistochemistry for keratins, epithelial membrane antigen, CEA, calretinin, desmin and actin were negative. In contrast, CD99 was multifocally positive, and chondroid foci were marked by S-100 protein. The proliferation index (MIB-1) was 60%.

DISCUSSION

Diagnostic criteria and histopathological features Clinical and radiological features Mediastinal sarcomas are generally rare. Anatomically, the mediastinal compartment contains the heart, large vessels, a portion of the esophagus, the thymus and adipose tissue. It is therefore logical to assume that any

mesenchymal tumor appearing as a mediastinal mass could originate from one of these structures. Thus, the esophageal wall could be the origin of a smooth muscle tumor, while a vessel wall could give rise to a vascular tumor.

However, it is even more pertinent to point out that the abundant presence of adipose tissue in the mediastinum suggests that lipomatous tumours should not only be considered as the only true primitive "mediastinal" tumours, but also as the most frequent mesenchymal neoplasms in this compartment. In fact, numerous cases of mediastinal sarcomas of various types have been described in the literature over the years.

The clinical manifestations of patients with mediastinal masses are usually related to compression of adjacent structures, which may result in symptoms such as dyspnea, cough, chest pain, back pain or other non-specific symptoms. However, some patients may be asymptomatic, and their mediastinal tumor may be discovered incidentally during a routine radiographic examination.

Unlike certain epithelial tumours such as thymomas, there is no specific link between mediastinal chondrosarcomas and autoimmune diseases such as myasthenia gravis, and any association is likely to be fortuitous. It is important to note that cases of chondrosarcoma and osteosarcoma have been reported following treatment for other conditions [18-26].

Mediastinal chondrosarcomas often present as a mediastinal mass. However, the presence of an associated periosteal connection may indicate a bony origin of the tumor, despite its appearance as a "mediastinal tumor". Thus, these tumors should not be considered to be of primary mediastinal origin, underscoring the importance of careful radiological evaluation.

Radiological characteristics

On computed tomography (CT) images, the presence of coarse internal calcifications in a "ring and bow" pattern is typical of a chondroid matrix (Figure 1A, B, C, D). The non-calcified part of the mass shows low attenuation relative to the muscle, suggesting the presence of hyaline (water-rich) cartilage, tumour necrosis or cystic content [3].

On T1-weighted magnetic resonance imaging (MRI) images, the masses show low to intermediate signal intensity, with foci of slight hyperintensity corresponding to areas of mineralization. On T2 sequences, these tumors show mixed heterogeneous signal intensity, with scattered areas of high intensity, reflecting high water content [3].

The presence of fibrovascular septa in chondrosarcomas is thought to be responsible for their lobulated morphology, enabling the growth of a predominantly avascular cartilaginous tumor [3]. After injection of intravenous contrast, the tumor shows heterogeneous enhancement.

Diagnostic Criteria

In view of the restrictions mentioned, we consider that the diagnosis of mediastinal chondrosarcomas should follow the following guidelines:

- **Unequivocal radiographic evidence** that the tumor has no connection with a bone structure.
- **No clinical history of radiotherapy** for mediastinal tumors, particularly in cases where only biopsy material has been evaluated, e.g. mediastinal germ cell tumors treated prior to complete surgical excision.
- **A full histopathological evaluation of** the resected tumour is required to establish the diagnosis.
- **Cases based solely on fine-needle biopsy or cytopuncture** should be regarded as presumptive diagnoses. Definitive interpretation can only be made after complete surgical evaluation of the resected tumor.
- **Extensive sampling of the tumor mass** is necessary to exclude the presence of any other component, whether epithelial or mesenchymal.
- **Diagnostic criteria for mediastinal chondrosarcomas** should be the same as those applied to conventional bone tumours.

Adherence to these guidelines will provide the essential parameters for accurate final interpretation of mediastinal chondrosarcomas.

Histopathological Features

Mediastinal chondrosarcomas can present a spectrum of forms similar to those of bone. There are conventional, myxoid and mesenchymal chondrosarcomas. Chondrosarcomas in the anterior mediastinum tend to be larger than those in the posterior. Anterior tumors are often well circumscribed but not encapsulated, while those in the posterior mediastinum may have less defined boundaries.

Molecular and Immunohistochemical Characteristics

Tumors often show expression of S-100, SOX4 and SOX9 protein. Some subcategories of chondrosarcomas, such as myxoid and mesenchymal chondrosarcomas, show specific molecular alterations, such as HEY1-NCOA2 gene fusion and NR4A3 gene rearrangement. These findings suggest that these tumors could be reclassified as sarcomas of uncertain origin.

DISCUSSION

The presence of malignant cartilage tissue in the mediastinum has been reported in a variety of mediastinal tumours, including germ cell tumours. Germ cell tumors can undergo somatic transformation into chondrosarcoma post-therapeutically, complicating their classification. What's more, some masses diagnosed as mediastinal chondrosarcomas may actually originate in other thoracic structures such as the ribs or spine.

CONCLUSION

With advances in molecular and imaging techniques, it is clear that many cases of mediastinal chondrosarcoma need to be reclassified. The application of strict criteria and the use of new technologies are reducing the number of tumors that can truly be considered primary mediastinal chondrosarcomas. This reappraisal calls for a rethinking of the classification and management of these rare tumors.

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