Posterior Mediastinal Chondrosarcoma Presenting as a Dumbbell-Shaped Mass Mimicking a Cystic Schwannoma: An Unusual Presentation

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ABSTRACT

Mediastinal chondrosarcoma is exceedingly rare. Moreover, it is particularly unusual in the posterior mediastinum. Actually, very few cases of chondrosarcoma of the posterior mediastinum are gleaned from the literature. On the other hand, neurogenic tumors make even more than 75% of all tumors in the posterior mediastinum, and they are most often schwannomas that account for about 90% of all dumbbell-shaped tumors. Exceptionally, a dumbbell-shaped mass may correspond to a chondrosarcoma. We report a case of a 48-year-old Moroccan woman who presented with a complaint of intermittent electric shock type pain in the right side of chest radiating to the ipsilateral arm over six months period. No apparent distress or mass was found during the physical examination. Chest X-ray showed well-circumscribed opacity located in the upper right part of the posterior mediastinum. Neck and chest computed tomography (CT) scans followed by magnetic resonance imaging (MRI) were carried out and revealed a dumbbell-shaped mass of the posterior mediastinum. Such radiologic findings have oriented towards the diagnosis of cystic schwannoma. As pathologic results were inconclusive after a CT-guided fine needle biopsy, the patient underwent tumor resection via right posterolateral thoracotomy. Histopathological examination of surgical resection specimen revealed a grade II chondrosarcoma with the invasion of peripheral margins. Therefore, the patient received adjuvant radiation therapy to the tumor bed. Our case suggests that in the case of a dumbbell-shaped or thoracic paravertebral tumor a diagnosis other than nerve sheath tumor should be considered, including chondrosarcoma.

Keywords: Chondrosarcoma, Posterior mediastinum, Schwannoma, Dumbbell-shaped

1. INTRODUCTION

Chondrosarcomas account for 20-27% of primary malignant bone tumors and commonly affect the pelvis and upper and lower extremities(1). Chondrosarcoma of the thorax is scarce; nevertheless, it is the most common malignant primary tumor of the bony thorax and the entire chest wall. Actually, it accounts for nearly one-third of all primary chest wall tumors(2). Furthermore, chondrosarcoma in any part of the mediastinum is exceedingly rare. It is particularly unusual in the posterior mediastinum(3).

The posterior mediastinum is the potential space along each side of the vertebral column and adjacent proximal portion of the ribs. Neurogenic tumors are considered the commonest posterior mediastinal tumors are accounting for 19%-39% of all mediastinal tumors and 75% of all posterior mediastinal tumors(4).They are presented by schwannomas which account for about 90% of all dumbbell tumors. Rarely, dumbbell mass is made by chondrosarcoma(3,5).

In some cases, based only on clinical and radiologic features, posterior mediastinal chondrosarcoma could be misdiagnosed as a nerve sheath tumor(3). Such a case that we report.

2. CASE REPORT

A 48-year-old Moroccan woman who has no past personal history of disease or surgery and no contributory family history, presented with a complaint of intermittent electric shock type pain in the right side of the chest, radiating to the ipsilateral arm, over six months period. Physical examination revealed no apparent distress. Actually, body temperature was 37.2 degrees, respiration rate was 15/min, pulse rate was 80/min, and blood pressure was 130/70. Examination of the thorax showed good expansion; there was no mass detected. Cardiac examination revealed normal. Neurological examination showed no deficit. Conventional chest radiography was performed and revealed an oval-shaped well-circumscribed opacity located in the upper right part of the posterior mediastinum. Chest CT scan was therefore carried out and showed a paravertebral tumor mass measuring 41mm, filling the right costovertebral gutter with no associated bone lysis. Such findings have oriented towards the nerve sheath tumor as a first differential diagnosis. Computed Tomography-guided fine needle biopsy of the mass was therefore performed. However, biopsy results were inconclusive. Furthermore, magnetic resonance imaging (MRI) confirmed the findings of a well-circumscribed tumor measuring 40mm x45mm, occupying the right paravertebral space at the level of the second, third and fourth dorsal vertebrae, and centered on the right T3–T4 foramen. The tumor comprised an enhanced peripheral crown and a large central non-enhancing area which appeared hypointense on T1-weighted images and hyperintense on T2-weighted images, corresponding to a cystic degeneration (Figure 1).

In addition, the tumor mass presented a dumbbell-shaped extension to the T3-T4 foramen, without reaching the dorsal spinal canal (Figure 2). Such images were suggestive of cystic schwannoma. The patient underwent gross total tumor resection via right posterolateral thoracotomy. Histopathological examination of surgical resection specimen showed tumor proliferation composed of lobules of variable size, characterized by a blue chondroid background. There were numerous spindle cells with moderate nuclear irregularities present within the chondroid matrix. Outside the lobules, there were areas with higher cellular density comprising small cells with hyperchromatic and dense nuclei, and scarce cytoplasm. In the immunohistochemical examination, tumor cells did not express S100 protein or CD99, whereas they showed positive immunostaining for CD45, CD20, and CD3, confirming thus their reactive nature. Furthermore, cartilaginous structures showed strong diffuse positivity for S100. Neither EMA nor cytokeratin was expressed. Based on pathological and immunohistochemical results the diagnosis of grade II, well-differentiated chondrosarcoma was established.

Fig.1: Spinal MRI: axial T1-weighted sequence (A) and axial T2-weighted sequence (B) showing posterior mediastinal tumor with a large central area which revealed low signal intensity on T1 sequence and high signal intensity on T2 sequence.
As peripheral margins were invaded, the patient received adjuvant radiation therapy to the tumor bed with 18 MV photons to a dose of 70 Gy in 35 fractions (Figure 3). At eight-month follow-up, no local or distant tumor recurrence was detected.

Fig. 3: CT-scan dosimetry imaging is showing dose distribution of adjuvant external beam radiation to the tumor bed

3. DISCUSSION

Chondrosarcoma is a malignant tumor with cartilaginous differentiation, with no bone or osteoid production. However, it can contain foci of tumor-induced or enchondral ossification\(^6\). Despite its rarity in the thorax, this tumor is the most common malignant primary tumor of both the bony thorax and the entire chest wall, whereas mediastinal chondrosarcoma remains exceedingly scarce. Actually, less than ten cases of mediastinal chondrosarcoma have been reported\(^2,7\). Moreover, this malignancy is particularly unusual in the posterior mediastinum\(^3\).

There is a slight male predominance in most series, with a median age near 50 years\(^2\). Clinical features of chondrosarcoma tend to be nonspecific. Pain is the most frequent symptom, occurring in 95% of patients. Neurologic symptoms are apparent in 45% of patients with spinal chondrosarcoma\(^8\). Moreover, a pathological rib fracture, hemothorax, or metastasis can bring out the diagnosis\(^6\).

Diagnostic imaging techniques play an important role in the diagnosis and management of mediastinal tumors. Actually, chest X-ray can orient the diagnosis whereas thoracic CT and MRI provide a better definition of the tumor’s seat and extension. The characteristic CT appearance of chondrosarcoma consists of a well-defined, lobulated soft tissue mass with foci of chondroid matrix calcification. Bone destruction and invasion of overlying soft tissue may also exist. Magnetic resonance imaging (MRI) is particularly useful for defining vascular or neural involvement and therefore provides complementary

information to the CT scan for tumors with mediastinal, paravertebral, or thoracic outlet involvement\cite{2,6,8}. In addition, on MRI chondrosarcoma presents a heterogeneous internal architecture and is of intermediate or low signal intensity on T1-weighted MR images and are heterogeneous on T2-weighted ones, with characteristically scattered areas of high signal intensity. Furthermore, chondrosarcoma may appear as a dumbbell shaped mass on MRI. However, very rare cases of dumbbell shaped chondrosarcoma of the posterior mediastinum are reported in the literature\cite{8,9}. The term “dumbbell tumors” means separate tumors that connect and comprise two or more separate regions, such as locations outside the paravertebral space, the intradural space and epidural space\cite{10}. Schwannoma which is a nerve sheath tumor accounts for about 90% of all dumbbell-shaped tumors. Thus, the dumbbell shaped tumor is considered as a typical shape for spinal schwannoma\cite{8,10}. In our case, based on these findings as well as on the presence of cystic degeneration image on MRI, we initially assumed the presented posterior mediastinal tumor was a cystic schwannoma, whereas it was actually a chondrosarcoma. Thus, regardless clinical and radiological presumptions, only histopathological examination could establish the definitive diagnosis. As fine-needle biopsy of the tumor usually gives only poorly abundant material that is not highly representative of the tumor, immediate wide resection of the tumor is the best course whenever possible\cite{6}. Macroscopically, chondrosarcomas ranged in size from 5 to 15 cm and were mostly encapsulated. They are grayish in color, opaque, soft in consistency, with a gelatinous aspect, sometimes pseudocystic. The cartilaginous islets are often voluminous, coalescent, and poorly limited, with at most the loss of the lobulated aspect of cartilaginous tumors\cite{6,7}. Histologically, based on cellular density, the degree of anisokaryosis, and nuclear hyperchromatism, chondrosarcomas are divided into three grades of increasing aggressivity according to the World Health Organization (WHO) classification (Table 1)\cite{6}.

\begin{table}[h]
\centering
\begin{tabular}{|c|c|c|c|}
\hline
\textbf{cellular density} & grade I & grade II & grade III \\
\hline
hyperchromatic and uniform & hyperchromatic and atypical & Hyperchromatic with prominent nuclear atypia \\
\hline
nuclear hyperchromatism & lack of pleomorphism & binucleate or multinucleate cells & pleomorphic and highly atypical \\
anisokaryosis & Absent & Rare & Frequent \\
mitoses & & & \\
\hline
\end{tabular}
\caption{Histological grading of chondrosarcoma according to the World Health Organization}
\end{table}

In terms of treatment, aggressive surgical resection remains the only curative treatment\cite{6,9}. In cases of unresectability or if a wider surgical margin cannot be achieved after incomplete resection, radiation therapy should be administered to achieve maximum local control\cite{2,8}.

Regarding prognosis, it has been reported that local recurrence occurred in up to 50% of patients with thoracic and mediastinal chondrosarcoma, despite wide local excision. About 40% of patients with local recurrences experienced subsequently metastatic disease. High tumor grade, inadequate margins at initial resection, and resection at a non-specialty center have been reported to predict local recurrence, whereas prognostic factors for metastases included the histological grade, tumor size, and local recurrence\cite{1,2}. Furthermore, it has been noticed that survival was influenced by tumor grade, tumor diameter, tumor location, and date of operation\cite{1}.

Owing to the possibility of late local recurrence and distant metastasis, patients should undergo lifelong follow up with a follow-up schedule including physical examination and thoracic imaging every 3–6 months for the first 5 years and annually after that for a minimum of 10 years\cite{2}.

\section{4. CONCLUSION}

Chondrosarcoma of the posterior mediastinum is exceedingly rare. Diagnostic imaging techniques...
including chest CT scan and MRI play an important role in the diagnosis and management of such tumors. However, the only histopathological examination can establish the definitive diagnosis. Actually, sometimes there is some trouble in diagnosing this type of tumors by imaging, especially when they are presenting as dumbbell-shaped tumors. Such was our case. Therefore, we suggest that in the case of a spinal dumbbell-shaped or thoracic paravertebral tumor a diagnosis other than nerve sheath tumor should be considered, including chondrosarcoma.

REFERENCES