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Case Report

Primary Pleural Low-Grade Fibromyxoid Sarcoma with Pulmonary Metastasis: A Case Report with a Literature Review

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Abstract

Low-grade fibromyxoid sarcoma also known as Evans tumor, is a rare soft tissue tumor that can occur in all parts of the soft tissues, especially in the deep soft tissues, generally localizes to the extremities and trunk, the primary intra thoracic location is uncommon.

Although the histological appearance is benign, the risk of recurrence and metastases is high, requiring long-term monitoring.

We report the case of a 29-year-old woman presenting with pleural fibromyxoid sarcoma, which is an exceptional primary site of development, associated with pulmonary metastases.

Keywords: *Evans Tumor; Immunohistochemistry; CT Scan*

Introduction

Low-grade fibromyxoid sarcoma, also known as Evans tumor, is a rare soft tissue tumor first described by Evans in 1987. It can develop in any part of the soft tissues, particularly in the deep soft tissues of the extremities and trunk, though it can also occur unusually in visceral areas. Despite its benign histological appearance, it has a high potential for metastasis.

We report the case of a 29-year-old woman diagnosed with a pleural Evans tumour with pulmonary metastases.

Case Report

A 29 year old women with no past medical history presented with a three month persistent cough not improved by symptomatic treatment.

A chest radiography revealed the presence of a large right pleural opacity, with bilateral pulmonary opacity, and a right pleural effusion.

A CT scan was then ordered to characterise the lesion and to perform a scan-guided biopsy to establish a histological diagnosis.

The CT scan showed an extensive basal right heterogeneous enhancing mass arising from the parietal pleura measured 150 x 154 x 130 mm, containing a large hypodense patches of necrosis and amorphous calcifications, with attachment to the right diaphragm which is lowered, compressing the inferior vena cava, with no extension to adjacent parietal subcutaneous soft tissue. We also noted well-defined bilateral lung nodules and masses ranging from 3 mm to 8 cm in size, along with a large left pleural effusion, and no evidence of abdominopelvic metastasis.

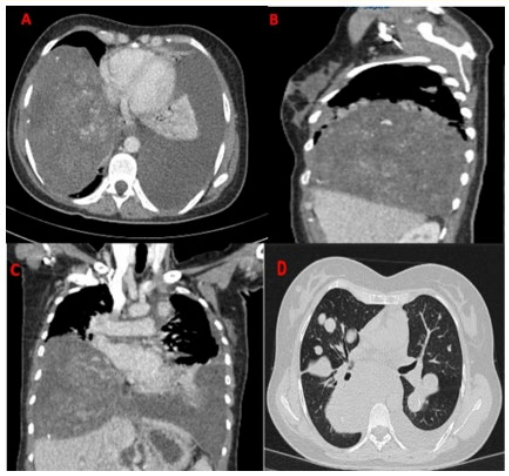

Figure 1: *The contrast-enhanced computed tomography images in the mediastinal window: axial view (A), sagittal view (B), and coronal view (C) show a large lobulated, hypodense mass in the right hemithorax, containing some calcification and heterogeneously enhanced, associated with a moderate left pleural effusion. Multiple bilateral pulmonary masses are observed on the axial section in the parenchymal window (D).*

Light microscopic examination showed a mesenchymal tumor characterized by fibroblastic proliferation made of spindle cells with slightly eosinophilic cytoplasm and round or oval, slightly irregular and hyperchromatic nuclei without visible nucleoli, accompanied by a few myxoid plaques with variable cell density, immunohistochemistry, the tumor was massively positive for MUC4 figure 2, negative for S-100 protein, and CD34, Ki67 index was 3% of tumor cells.

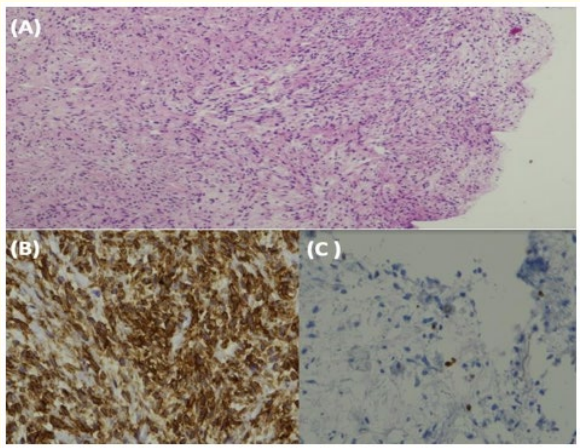

Figure 2: *Pathological examinations of the biopsied pleural mass, demonstrating hyalinized spindle cell rosettes and cellular spindle cell component with hypocellular myxoid areas (A), the immunohistochemistry of the tumor was massively positive for MUC4 (B), and low Ki-67 labeling index (10×) (C).*

Discussion

Low-grade fibromyxoid sarcoma (LGFMS) is a specific variant of fibrosarcoma, first described by Evans in 1987 [1], it's a rare soft tissue tumor, represent less than 5% of soft tissue sarcomas.

Patients of all ages can be affected, though the incidence is higher in young adults, with about 20% of patients being under 18 years old [2]. The tumor can occur in any part of the soft tissues, especially in the deep tissues of the proximal trunk and extremities. Less commonly, it can also be found in the intrathoracic region, abdominal cavity, and retroperitoneum [3-5].

Pleural LGFMS are exceedingly uncommon as a primary location, and metastasis from an extra-thoracic site needs to be excluded [6]. Only six cases were reported in the literature, including 4 cases with no metastases at diagnosis, originally reported in 2005 in a 37-year-old man by Kim., *et al*. [7] the second case was described in a 42-year-old woman by Liang., *et al*. in 2016 [8], a third case was reported in a 32- year-old patient by Perez., *et al.* in 2020 [9] and finally a case in the pediatric population involving a 4-year-old child was documented by Xiangni., *et al* [10].

Furthermore, only one case of LGFM pleural with subsequent peritoneal metastases development a few months after surgical excision of the tumor has been reported [11].

While in our case, the patient already had pulmonary metastases at the time of diagnosis.

The symptoms associated with an intrathoracic mass can differ based on its specific size and location. The patient may experience a chronic cough, chest pain, shortness of breath during physical activity, and accumulation of fluid in the pleural cavity. However, it's not uncommon for such masses to present without symptoms and to be detected incidentally during imaging as a slow-growing mass [9].

LGFMS is distinguished by certain imaging traits observed in MRI and CT scans, although such features are not exclusive to this condition. On non-contrast-enhanced CT scan, the fibrous elements of these tumors present with a density similar to the muscle, and the myxoid regions appear less dense. Additionally, calcifications may be present.

On MRI, the fibrous areas typically demonstrate a low signal intensity on both T1 and T2-weighted images, with a modest enhancement on T1 after contrast administration, while the myxoid areas are low in signal on T1-weighted images, highly intense on T2-weighted images markedly enhanced after contrast administration [7,12].

Histologically, low-grade fibromyxoid sarcoma presents an appearance that can be deceptively benign, with spindle-shaped cells arranged in a whorling pattern as well as regions of dense collagen and myxoid tissue. On immunohistochemistry, these tumour are often positive for mucin 4, negative for a range of other antibodies such as S100, desmin and keratin [13,14].

Although its histological appearance is benign, it has a high recurrence and metastatic potential, different rates of recurrence and metastasis are observed in various studies, and it is commonly acknowledged that tumor morphology does not necessarily indicate a worse prognosis.

However, tumor dedifferentiation can result in a shorter survival duration. On average, the time to metastasis is around 5 years, while the time to local recurrence is approximately 3.5 years [1,3,15]. Therefore, it is advisable to closely monitor these patients after diagnosis and surgery, especially in the long term [16].

Surgical treatment alone is impracticable in LGFMS with metastases. Chemotherapy and radiotherapy are still required, even though there aren't established treatment guidelines for the use of chemoradiotherapy due to the tumor's low malignancy [17].

Conclusion

Pleural low-grade fibromyxoid sarcoma is a rare entity, and metastases to extra-thoracic sites must be excluded. Although the histological appearance is benign, the potential for recurrence and metastasis is high. Therefore, close surveillance of these patients after diagnosis and surgery, especially long-term monitoring is highly recommended.

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