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Radiology

Primary Undifferentiated Pulmonary Sarcoma (FNCLCC Grade II) in a young adult: A Rare Case and Review of the Literature

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Abstract | **Case Report**

Primary pulmonary sarcomas are extremely rare malignant tumors of mesenchymal origin, representing less than 0.5% of all primary lung malignancies. Due to their rarity and non-specific clinical and radiologic features, diagnosis is often delayed. We report the case of a 26-year-old patient presenting with a primary undifferentiated pulmonary sarcoma of FNCLCC grade II. Diagnosis was established through imaging and histopathological examination. Despite initiation of chemotherapy, the disease progressed rapidly and the patient died shortly after the third cycle. We discuss the diagnostic challenges and therapeutic considerations in managing such rare and aggressive tumors.

Keywords: Primary pulmonary sarcoma, Undifferentiated sarcoma, Rare lung malignancy, Diagnostic challenge, Aggressive tumor.

INTRODUCTION

Primary sarcomas of the lung are exceedingly rare and heterogeneous neoplasms of mesenchymal origin. They are often misdiagnosed as more common epithelial lung tumors. Among them, undifferentiated sarcomas represent a subgroup characterized by the absence of any identifiable line of differentiation on histology or immunohistochemistry. These tumors are aggressive and associated with poor prognosis. Due to their rarity, there is no standardized treatment, and management often relies on multimodal therapy. This case report highlights the diagnostic and therapeutic challenges associated with this entity.

CASE PRESENTATION

A 26-year-old male with no significant past medical history presented with a 2-month history of chronic cough and dyspnea. Chest radiography revealed a large opacity in the left upper lobe. A chest CT scan confirmed the presence of a large, heterogeneously enhancing mass in the left upper lobe, measuring 83 x 73 x 83 mm. The mass was in close contact with adjacent vascular structures, including the left subclavian artery and vein, as well as the aortic arch, without evidence of invasion. Additionally, subcentimetric mediastinal lymphadenopathy was noted in the anterior mediastinum, pretracheal region, and the aorto-pulmonary window, with the largest node measuring 7 × 14.9 mm.

A thoraco-abdomino-pelvic and brain CT scan showed no evidence of distant metastasis.

Axial (A–B), coronal (C–D), and sagittal (E) contrast-enhanced CT images in mediastinal window, with lung window settings shown in axial images (F–G), demonstrate a large, well-defined heterogeneous mass occupying the left hemithorax. There is a mediastinal shift to the right and compression of the adjacent lung parenchyma. The lesion shows no clear invasion of the chest wall or mediastinum. It rests on the major (oblique) fissure, which appears bulging due to mass effect.

The mass was subsequently biopsied; the final diagnosis was consistent with a poorly differentiated sarcoma of the lung, FNCLCC grade II, with no specific histologic subtype identified on morphology or immunohistochemistry — thus classified as an undifferentiated sarcoma.

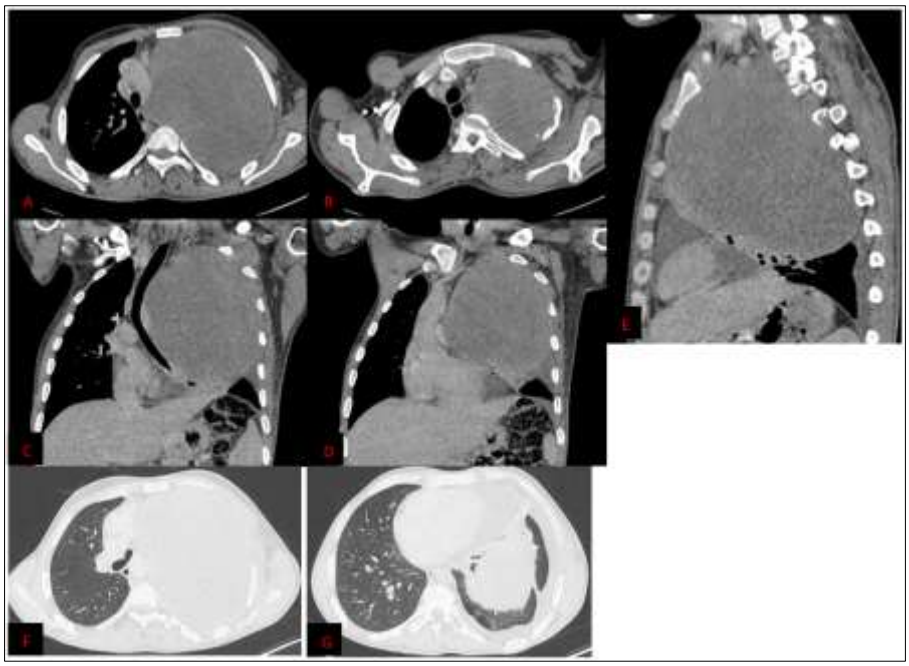

Figure 1: Thoracic contrast-enhanced CT showing a large left pulmonary mass

The case was reviewed in a multidisciplinary tumor board. Due to the large size and proximity to vital mediastinal structures, the tumor was deemed unresectable. The patient was started on systemic chemotherapy and received three cycles. Unfortunately, the clinical course was rapidly progressive, and the patient died shortly after completing the third chemotherapy cycle.

Histopathological examination demonstrated a malignant proliferation of small round to spindle-shaped cells with high mitotic activity and a fibro-inflammatory stroma. Immunohistochemistry revealed heterogeneous positivity for CD34, EMA, TLE1, and focal positivity for smooth muscle actin, while markers such as desmin, pan-cytokeratin, GFAP, and STAT6 were negative. The Ki-67 proliferation index was elevated at 35–40%. These findings are consistent with a poorly differentiated sarcoma, FNCLCC grade II, without a specific histologic subtype, thus classified as an undifferentiated sarcoma (not otherwise specified).

DISCUSSION

Primary pulmonary sarcomas (PPS) are exceedingly rare, accounting for less than 0.5% of all primary lung malignancies [1, 2]. They encompass a heterogeneous group of mesenchymal tumors, including synovial sarcoma, leiomyosarcoma, angiosarcoma, and undifferentiated pleomorphic sarcoma [3, 4]. Due to their rarity and nonspecific clinical presentations, PPS often pose significant diagnostic challenges [1-5]. Radiologically, PPS often present as large, well-circumscribed masses with heterogeneous enhancement on contrast-enhanced CT scans [6]. Calcifications, necrosis, and hemorrhage may be present, and these tumors can exhibit aggressive features, such as invasion into adjacent structures [6, 7].

Differential diagnoses for such pulmonary masses include metastatic lesions, primary lung carcinomas, and other sarcomatoid tumors [3]. The absence of epithelial markers and the specific immunohistochemical profile aid in distinguishing primary pulmonary sarcomas from these entities [5-8]. Moreover, the lack of a primary tumor elsewhere supports the diagnosis of a primary pulmonary origin [5].

Treatment options for PPS are limited and often involve surgical resection when feasible, combined with chemotherapy and/or radiotherapy [2-6].

However, the prognosis remains poor due to the aggressive nature of these tumors and their tendency for early metastasis [1-9]. In this case, the patient underwent three cycles of chemotherapy but unfortunately succumbed shortly thereafter, highlighting the need for early detection and the development of more effective therapeutic strategies [7].

CONCLUSION

Primary undifferentiated sarcoma of the lung is a rare and aggressive malignancy with a high risk of early progression. Timely diagnosis and multidisciplinary evaluation are crucial. Prognosis remains poor in advanced cases despite systemic chemotherapy. This case underlines the need for heightened clinical awareness and more research to establish effective treatment protocols.

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