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Primary Pulmonary Undifferentiated Pleomorphic Sarcoma: A Rare Malignant Lung Tumor

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Abstract

We report a case of a 56-year-old man with persistent right upper lobe cavitary mass. A chest computed tomography scan showed about 4-cm-sized mass with internal low attenuation and peripheral enhancement in right upper lobe apical segment. The lesion size increased over 1 month. Right upper lobectomy was performed with the intention to treat the lesion. Pathological examination showed primary pulmonary undifferentiated pleomorphic sarcoma. We describe this rare lung disease to remind that primary pulmonary undifferentiated pleomorphic sarcoma could be the differential diagnosis of pulmonary cavitary mass lesions.

Keywords

malignant fibrous histiocytoma, undifferentiated pleomorphic sarcoma, lobectomy

Introduction

Undifferentiated pleomorphic sarcoma is one of the most common soft tissue sarcomas of late adulthood.[1] Undifferentiated pleomorphic sarcoma is most often found in the extremities and the retroperitoneum.[2] The lungs are the most common sites of metastasis.[3] However, primary pulmonary undifferentiated pleomorphic sarcoma is an extremely rare type of primary lung malignancy, which was first described in 1979. Approximately 36 cases have been identified in the English literature till now.[4] Primary pulmonary undifferentiated pleomorphic sarcoma is very rare but aggressive tumor with a poor prognosis and low survival rate.[5] There is no established treatment recommendation for its paucity. The present study case presents a case of primary pulmonary undifferentiated pleomorphic sarcoma, which was treated by surgical resection.

Case Report

A 56-year-old man presented to our clinic with a history of persistent right upper lobe (RUL) cavitary mass. He was a 30-pack-year current smoker. A chest computed tomography scan revealed about 4-cm-sized mass with internal low attenuation and peripheral enhancement in RUL apical segment. A positron emission tomography-computed tomography scan showed a well-defined mass-like lesion with intense fluorodeoxyglucose uptake in medial portion of RUL (Figure 1). The maximum standardized uptake value was 44.4. Any extrapulmonary involvement was not detected. The patient had been under observation for approximately 1 month in the outpatient pulmonology department. The size of lesion had increased in size, from 4 cm to 7 cm in long diameter. The patient underwent a right upper lobectomy and systemic lymphadenectomy. Wedge resection for intraoperative frozen section was performed to determine the nature of lung mass. Cut section of the wedge resection specimen showed an ill-demarcated yellow-tan solid mass with hemorrhage and necrosis (Figure 2A). After a diagnosis of malignancy was made on frozen section, lobectomy with lymph node dissection was followed. Microscopically, the ill-defined tumor was bordered by thick inflamed tissue containing lymphoid follicles (Figure 2B). On high power microscopic view, the tumor was composed of large, pleomorphic cells showing patternless growth with numerous inflammatory cell infiltrates. Frequent mitotic figures were identified (Figure 2C). Several foci of tumor cell necrosis were also seen (Figure 2D). Histopathologically,

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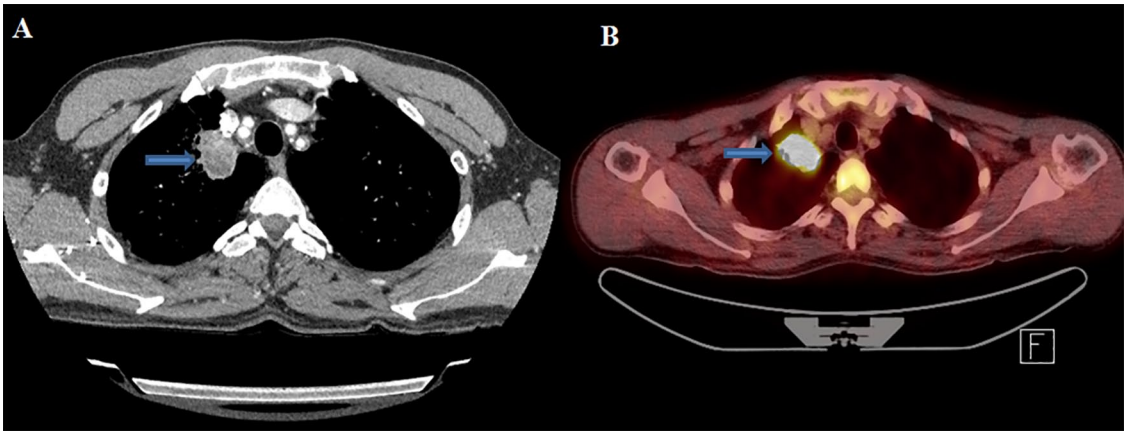

Figure 1. A preoperative chest computed tomography scan shows about 4-cm-sized mass with internal low attenuation and peripheral enhancement in right upper lobe apical segment (A, arrow). A positron emission tomography-computed tomography scan shows a well-defined lesion with intense fluorodeoxyglucose uptake in right upper lobe (B, arrow).

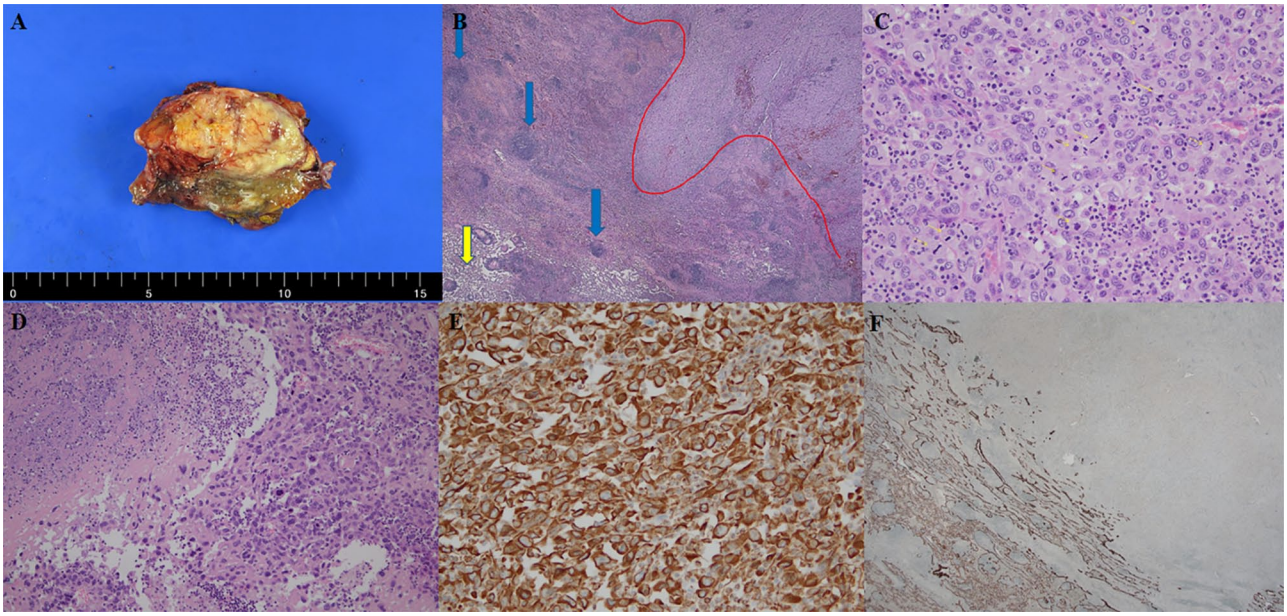

Figure 2. Gross photograph of the wedge resection specimen shows an ill-demarcated yellow-tan solid mass with hemorrhage and necrosis (A). Microscopically, the ill-defined tumor is bordered by thick inflamed tissue containing lymphoid follicles (B, red: tumor, blue: lymphoid follicles, yellow: lung parenchyma). The pleomorphic tumor cells show numerous neutrophils infiltrate. Note the frequent mitotic figures (C, yellow arrows). Tumor cell necrosis is seen, which is a feature of high-grade sarcoma (D). Immunostaining for vimentin reveals diffuse strong cytoplasmic staining (E). Immunohistochemistry for pancytokeratin is negative in tumor cells, while remaining pneumocytes and bronchiolar epithelium show normal expression (F).

no carcinomatous differentiation was identified even after meticulous search with examination of entire tumor. Immunohistochemically, the tumor cells expressed vimentin, but were negative for CK (pan), CK7, CK5/6, EMA, TTF-1, p63, LCA, CD34, CD30, S-100, SMA, and desmin (Figure 2E and F). This histopathologic findings and results of immunohistochemistry supported that the tumor was malignant mesenchymal tumor without identifiable line of differentiation. Therefore, this lung mass was diagnosed as undifferentiated pleomorphic sarcoma. There was no lymph node metastasis. The patient had an uneventful clinical course and discharged without complication.

Discussion

An undifferentiated pleomorphic sarcoma, which was previously named malignant fibrous histiocytoma, was newly named by the World Health Organization in 2012. This new terminology suggests that undifferentiated pleomorphic sarcoma is an aggressive soft tissue sarcoma originating from mesenchymal cells.[1] An undifferentiated pleomorphic sarcoma usually occurs in patients between 32 and 80 years old with a male predominance.[6] Although patients usually present late with metastasis, most frequently to lungs, primary pulmonary undifferentiated pleomorphic sarcoma arising in the lung is rare. Patients may be asymptomatic or are diagnosed with cough, dyspnea, or chest pain. Radiographically, most patients with primary pulmonary undifferentiated pleomorphic sarcoma presented with a solitary pulmonary nodule on imaging, such as chest computed tomography. A positron emission tomography-computed tomography scan could be helpful in ruling out extrathoracic metastasis.[5] Maeda and colleagues[7] reported that preoperative histological diagnosis was rare and only 4% of reported cases were histologically diagnosed before surgery. A primary pulmonary undifferentiated pleomorphic sarcoma was insensitive to both chemotherapy and radiotherapy, and complete surgical resection of tumors with systematic lymph node dissection was the treatment of choice for the survival of patients in the published literatures.[4] The prognosis for patients with primary undifferentiated pleomorphic sarcoma is usually poor. The overall survival of reported cases ranged from 0 to more than 168 months.[4,5]

Our report serves as reminder that that primary pulmonary undifferentiated pleomorphic sarcoma could be the differential diagnosis of pulmonary cavitary mass lesions although it is extremely rare.

Declaration of Conflicting Interests

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Ethics Approval

Our institution does not require ethical approval for reporting individual cases or case series.

Informed consent

Written informed consent was obtained from the patient(s) for their anonymized information to be published in this article.

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References

1. O'Brien JE, Stout AP. Malignant fibrous xanthomas. *Cancer*. 1964;17:1445-1455.
2. Weiss SW, Enzinger FM. Malignant fibrous histiocytoma: an analysis of 200 cases. *Cancer*. 1978;41:2250-2266.
3. Yousem SA, Hochholzer L. Malignant fibrous histiocytoma of the lung. *Cancer*. 1987;60:2532-2541.
4. Li X, Liu R, Shi T, et al. Primary pulmonary malignant fibrous histiocytoma: case report and literature review. *J Thorac Dis*. 2017;9:E702-E708.
5. Patel DP, Gandhi YS, Sommers KE, Mangar D, Camporesi EM. Primary pulmonary malignant fibrous histiocytoma. *Case Rep Pulmonol*. 2015;2015:381276.
6. Seomangal K, Mahmoud N, McGrath JP. Malignant fibrous histiocytoma, now referred to as undifferentiated pleomorphic sarcoma: a case report of an unexpected histology of a subcutaneous lesion. *Int J Surg Case Rep*. 2019;60:299-302.
7. Maeda J, Ohta M, Inoue M, et al. Surgical intervention for malignant fibrous histiocytoma of the lung: report of a case. *Surg Today*. 2007;37:316-319.