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Case Report

Thoracoscopic Wedge Resection for Low-Grade Fibromyxoid Sarcoma (Evans Tumor) with Massive Calcification and Originating from the Lung: A Rare Case in an Unexpected Location

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We encountered a rare case of low-grade fibromyxoid sarcoma, which is generally known as Evans tumor, with massive calcification originating from the lung. The patient was a 22-year-old man with Duchenne muscular dystrophy who was referred for a detailed investigation of an intrathoracic tumor with massive calcification. Although our preoperative diagnosis was a solitary fibrous tumor originating from the mediastinum or diaphragm, intraoperative thoracoscopy revealed the tumor arising from the left lower lobe without adhesion to the other organs. Considering his medical history, we aimed to preserve lung function and chose wedge resection, which completely removed the tumor. Based on the pathological findings, the tumor was diagnosed as low-grade fibromyxoid sarcoma with massive calcification originating from the lung. Although extremely rare, this tumor should be considered as a differential diagnosis for a solitary lung mass with massive calcification in young adults.

Keywords: calcification, immunohistochemistry, low-grade fibromyxoid sarcoma (Evans tumor), surgical treatment

Introduction

Low-grade fibromyxoid sarcoma (LGFMS) is a rare tumor that was first described by Evans in 1987[1)] and had been generally known as Evans tumor. It usually presents in young adults as a painless mass, mainly on the proximal extremities and trunk.[2)] The incidence of LGFMS was estimated to be 0.18 per million.[3)] LGFMS originating from the lung is extremely rare and has been previously reported in only five cases.[4–8)] Therefore, the clinical characteristics of this tumor have not been well known and preoperative diagnosis based on clinical findings, including computed tomography (CT) scan, has been difficult. In addition, careful pathological assessment is required to correctly diagnose LGFMS, which has a benign-like histology. Because of its low malignancy grade and low mitotic rate, LGFMS appears to be resistant to systemic chemotherapy or radiation. Therefore, surgical resection with clear margins remains the mainstay of treatment for LGFMS.[9)] In this report, we presented a case of successful thoracoscopic lung wedge resection of LGFMS with massive calcification originating from the lung.

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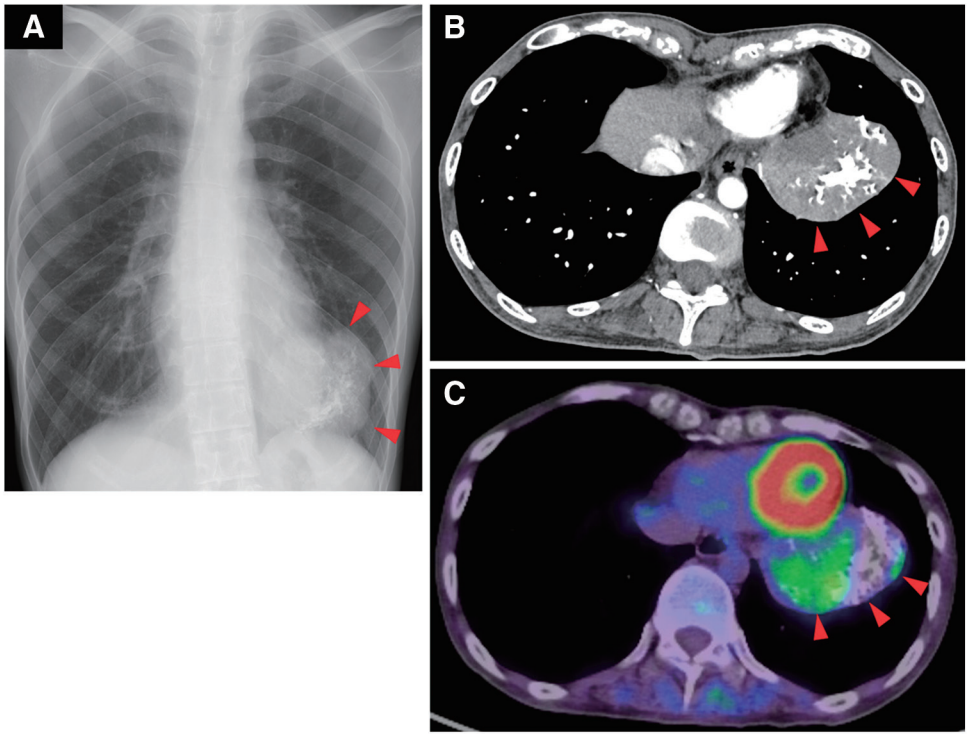

Fig. 1 Imaging findings in this case. (**A**) Chest X-ray examination shows a circular heterogenous mass occupying the lower part of the left thorax (red arrowheads). (**B**) Computed tomography shows an 8.0 cm circular hypodense mass with a well-defined margin and massive calcification (red arrowheads). (**C**) Positron emission tomography demonstrates the mass with partial uptake of ^{18}F-fluorodeoxyglucose with a maximum standardized uptake value of 2.9 (red arrowheads).

Case Report

A 22-year-old asymptomatic man was referred to our institution for detailed investigation of an enlarged left intrathoracic mass, which had been noted on chest X-ray for the past eight years. He was a never-smoker, and his medical history was Duchenne muscular dystrophy, which was diagnosed and treated at another hospital. **Figure 1** shows the imaging findings that were done on the patient. Chest X-ray revealed a circular heterogenous mass occupying the lower part of the left thorax. CT showed an 8.0 cm circular hypodense mass with a well-defined margin and massive calcification and was surrounded by the left ventricle, diaphragm, and lungs. However, the origin of the mass was not evident from the CT images. Preoperative ^{18}F-fluorodeoxyglucose (^{18}F-FDG) positron emission tomography (PET) demonstrated the mass with partial uptake of FDG with a maximum standardized uptake value of 2.9. These examinations excluded extrathoracic malignancy. Our preoperative diagnosis was a solitary fibrous tumor (SFT), commonly encountered in clinical practice, suspected to originate from the mediastinum or diaphragm due to its extensive contact with these organs. Therefore, thoracoscopic surgery was performed for treatment and definitive diagnosis. Intraoperative thoracoscopy revealed the tumor arising from the left lower lobe without adhesion to the other organs (**Fig. 2**). Using two-port thoracoscopic surgery, wedge resection was performed on the affected left lower lobe and enabled complete removal of the tumor. The operative time was 97 min, and the blood loss was 13 mL. A surgical margin greater than 3 cm grossly was secured. Postoperatively, the patient developed aspiration pneumonia, which was likely associated with Duchenne muscular dystrophy, but was discharged from the hospital after a short course of antibiotic therapy. On follow-up after six months, the patient was disease-free and had no symptoms.

On pathology (**Fig. 3**), the tumor had a smooth surface, was white, and measured 8.0 cm in its largest axis on gross examination. Histological examination revealed uniform and bland appearing spindle-shaped cells with a fibrous area; hyalinized rosettes (i.e., giant rosettes); coarse calcification; and vitreous components. The tumor demonstrated no mitosis and necrosis. The tumor was observed to arise continuously from the lung. Microscopically, the surgical margins were free of

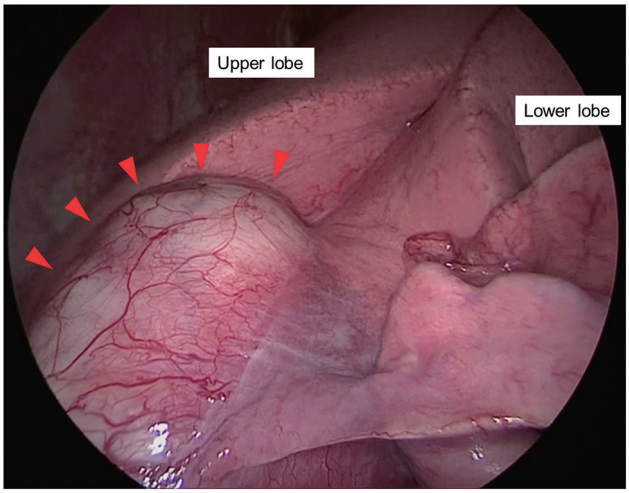

Fig. 2 Intraoperative findings. Thoracoscopy shows the tumor arising from the left lower lobe without adhesion to the other organs (red arrowhead).

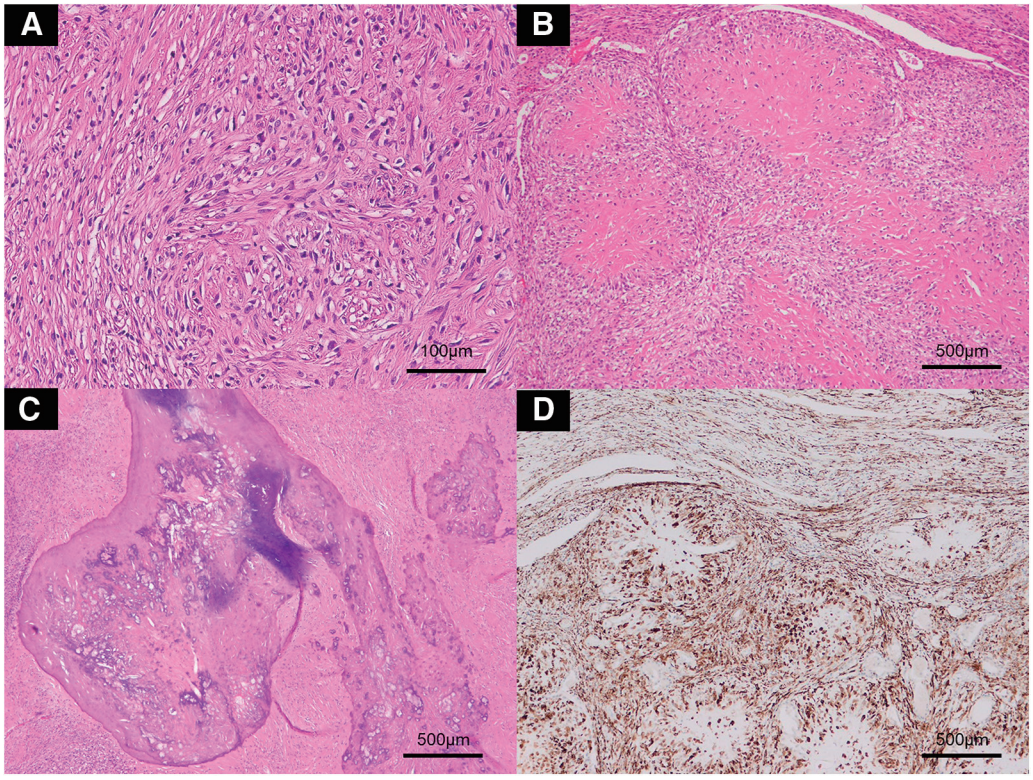

Fig. 3 Microscopic findings of the surgically resected tumor. The tumor comprises (**A**) uniform and bland-appearing spindle-shaped cells with a fibrous area and (**B**) hyalinized rosettes (i.e., giant rosettes). (**C**) There are coarse calcification and vitreous components in the other areas. (**D**) On immunohistochemistry, the tumor cells are diffusely positive for MUC4.

disease. Immunohistochemical analysis revealed that the tumor cells were diffusely positive for MUC4 and negative for STAT6, CD34, SMA, desmin, S-100, HMB-45, β-catenin, CD31, calretinin, ALK, CKAE1/AE3, and TTF-1. These microscopic findings and diffuse positivity for MUC4 were compatible with LGFMS.

Discussion

LGFMS, which is generally known as Evans tumor, arises mainly on the proximal extremities and trunk and rarely originates from the lung. In fact, only five cases of LGFMS originating from the lung have been previously

reported.[4–8] As a result, the clinical characteristics of this tumor are not well known. On CT scan, LGFMS was previously described to have low attenuation components that were hypodense relative to the skeletal muscle; these findings were likely secondary to the myxoid component.[10–12] Although PET has demonstrated ^{18}F-FDG uptake in both primary and metastatic LGFMS lesions,[13] some primary tumors had no ^{18}F-FDG uptake.[3] Therefore, preoperative PET imaging may not be useful for differential diagnosis. Although calcification has been reported in LGFMS,[14] obvious and massive calcification on chest X-ray, which was seen in our case, is atypical. Heterotopic calcification is generally considered nonspecific, metaplastic, or reactive; according to a previous report, it might be the consequence of a long-standing tumor.[15] These cases with heterotopic calcification may represent a diagnostic challenge because the differential diagnosis would include other tumors that have more frequent bone formation (e.g., extraskeletal osteosarcoma, synovial sarcoma, sclerosing epithelioid fibrosarcoma, dedifferentiated liposarcoma, and epithelioid sarcoma). Microscopically, these portions were reported to contain small osteocytes and osteoblasts without atypia.[16,17] Therefore, preoperative clinical diagnosis of LGFMS is challenging. In our case, SFT, which also rarely presents with calcification,[18] was suspected preoperatively. SFT must be differentiated from LGFMS originating from the lung because one of the five reported latter cases also presented with calcification. However, compared with the previously reported case, our case showed massive calcification, which may contribute to diagnosis by clinical imaging in the future.

The treatment for all the previously reported cases of LGFMS originating from the lung was anatomical resection, including lobectomy and pneumonectomy.[4–8] In our case, although the tumor had a diameter of 8.0 cm, it was pedunculated and had no evidence of invasion into the surrounding organs. Therefore, we were able to remove the tumor with enough margins by wedge resection. In addition, the preservation of the patient's lung function, because he was young and had Duchenne muscular dystrophy, was considered to be directly related to the prolonged prognosis. Furthermore, a more extensive surgical procedure, such as lobectomy, might have aggravated the postoperative aspiration pneumonia and led to a more severe clinical course after surgery. Therefore, we chose wedge resection and could safely remove the tumor completely. To the best of our knowledge, this was the first report on wedge resection under thoracoscopic surgery for LGFMS originating from the lung. Insufficient margins warrant consideration of anatomic resection. As noted in previous studies,[4–6] the choice of surgical method may influence preoperative diagnosis, typically achieved through CT-guided needle biopsy or thoracoscopic biopsy. Our case presented challenges for CT-guided needle biopsy due to the tumor's location, making thoracoscopic biopsy followed by planned surgery a potential alternative. Notably, we did not perform mediastinal lymph node dissection. Primary lung sarcomas infrequently metastasize to lymph nodes; however, Régnard et al. found N2 involvement in two out of 20 patients who underwent resection and N1 involvement in three.[19] Preoperative diagnosis could necessitate considering mediastinal lymph node resection.

LGFMS has the potential to metastasize to atypical sites and to relapse long after the primary treatment.[2] This affects the choice of the best imaging method as well as the optimal duration of follow-up. The best recommendation for diagnosing LGFMS and detecting any disseminated lesions is full-body magnetic resonance imaging (MRI), which was shown to have higher sensitivity compared with PET; in fact, the latter may not detect lesions and delay proper management.[3] Locally recurrent LGFMS was reported to appear as a cluster of masses on CT and MRI.[10] The multiplicity of masses seen with the local recurrence of LGFMS could be secondary to multiple residual tumor foci after an incomplete surgical resection, based on a previous histologic demonstration of variable degrees of infiltration into the adjacent soft tissues.[2] The previous case reports did not observe a recurrence of LGFMS originating from the lung, although the follow-up periods were unknown or short.[4–8] In our case, there were no signs of recurrence on CT during the six-month follow-up. Notably, we need to pay particular attention to the lung dissection line because we performed a wedge resection. Further long-term follow-up is required for early detection of late recurrence.

Conclusions

In summary, we presented a rare case of surgically resected LGFMS with massive calcification originating from the lung. Physicians should consider this entity as a differential diagnosis for a solitary lung mass with massive calcification in young adults.

Declarations

Ethics approval and consent to participate

Informed consent was not obtained for this Case Report as it is not mandated in such cases. However, all procedures were conducted in accordance with the ethical standards of the 1964 Declaration of Helsinki and its later amendments.

Funding

None.

Disclosure statement

None to declare.

Data availability statement

Not applicable.

Authors' contributions

HW conceived and designed the study, acquired and analyzed data, and drafted the manuscript and figures.

KN, HU, and TK contributed to the study conception and design, analyzed data, and reviewed and edited the manuscript.

YS and TFC-Y participated in the study conception and design and reviewed and edited the manuscript.

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