Giant Primary Mesenchymal Chondrosarcoma of the Lung: Case Report and Review of Literature

Bo Mei, MD, Ying-Long Lai, MD, Guang-Jie He, MD, Yun-Nan Shou, MD, and Jun Liu, MD

Mesenchymal chondrosarcoma, a rare malignant tumor, was predominantly occurring in the bone and may involve somatic soft tissue but it is extremely rare in the lung. We report the case of a 20-year-old female who presented with a 2-month history of irritant nonproductive cough and chest pain. The histopathologic examination revealed the tumor composed of atypical undifferentiated small cells and islands of matured chondroid matrix typically presented as bimorphic appearances. Immunohistochemical examination revealed that the tumor cells were positive for vimentin and CD99 for all components, and to S-100 limited to the areas of cartilage. In addition, previously reported cases of primary lung mesenchymal chondrosarcoma were reviewed, and the relevant clinical knowledge regarding its clinical manifestations, diagnosis, and treatment were discussed.

Keywords: lung neoplasms, mesenchymal chondrosarcoma

Introduction

Mesenchymal chondrosarcoma is a rare malignant tumor composed of atypical undifferentiated small cells and islands of matured chondroid matrix typically presented as bimorphic appearances. Mesenchymal chondrosarcoma, first described in 1959 by Lichtenstein and Bernstein, was common in young adults and teenagers (no significant sex predilection), and was predominantly occurring in the bone and may involve somatic soft tissue but it is extremely rare in the lung. To date, only 3 cases have been published (online PubMed search) as primary mesenchymal chondrosarcoma of the lung.1-4

We report a case whose tumor was larger one than ever before reports.

Case Report

A 20-year-old female patient presented with a 2-month history of irritant nonproductive cough and chest pain. Physical examination revealed decreased breath sounds over the left basal lung field. Chest contrast enhanced computed tomography (CT) demonstrated an ill defined margin large mass measuring approximately 11.6 cm × 9.2 cm × 6.0 cm in the left lower lung and occluding the lumen of the left lower lobe bronchus. The tumor was nonhomogeneous in density and intensified unevenly in the arterial phase and associated with almost entire consolidation of the left lower lobe. Most of the mass was closely attached to the parietal pleura, whereas there was no ribs destruction (Fig. 1). Bronchoscopy revealed a whitish tumor obstructing the left lower bronchus. Skeletal system without abnormalities was diagnosed by technetium-99m methylene diphosphonate (Tc-MDP) bone scan. Ultrasound-guided needling biopsy revealed small round cells and short shuttle-like cells with a high suspicion of malignancy.
Contrast-enhanced computed tomography revealed a large inhomogeneous mass (11.6 cm × 9.2 cm) that was associated with almost entire consolidation of the left lower lobe.

A left pneumonectomy and mediastinal lymph node dissection was performed. Intraoperative findings included gigantic tumor located at the left lower lobe of the lung, which involved the upper left lobe of the lung, and several mediastinal lymph nodes enlarged. The resected mass measured 13.0 cm × 8.0 cm × 6.0 cm in size (Fig. 2), the cut surface of which was gray-white to dark red, texture of which is not uniform that some areas were very firm and other soft, some areas of the tumor possessed clear boundaries but some poorly circumscribed. The cutting margin was checked by examination of frozen sections to ensure that the resection was clear.

Microscopically, the majority of tumor cells were small round cells and short shuttle-like cells, both of its size and morphology were disparity, which shows vacuolar, scant cytoplasm and nuclear mitoses. A few immature cartilage cells could be found in immature cartilage matrix in some field of view (Fig. 3A). The immunohistochemical stains shows that the cells presented diffuse positive reactions to vimentin (Fig. 3B) and CD99 for all components, and to S-100 limited to the areas of cartilage. In contrast, the tumor cells were not reactive to synaptophysin, epithelial membranous antigen, chromogranin, neurospecific enolase, cytokeratins, or glial fibrillary acidic protein.

Surgically resected margins was tumor free, unfortunately, one of subcarinal lymph nodes was metastasised. Mesenchymal chondrosarcoma was diagnosed. Postoperative course was satisfactory, and the patient was discharged after operative day 12. The patient was lost to follow-up (1 month after operation).

Discussion

Mesenchymal chondrosarcoma, a subtype of chondrosarcoma, derived from primitive mesenchymal tissue with potential chondroblast. Mesenchymal chondrosarcoma most commonly presents as a primary neoplasm of the bone, and may occur in other sites, such as the kidney, thyroid, prostate, mediastinum, and meninges. As a primary tumor, mesenchymal chondrosarcoma of the lung is an extremely rare tumor with only about 3 cases been reported in the literature, so metastasis from the more common primary sources must be convincingly ruled out before considering this diagnosis. In our case, there is no evidence of another primary site as determined by physical and image detection.

The clinical manifestations of pulmonary mesenchymal chondrosarcoma lacked specificity. The majority of
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Fig. 3 (A) The majority of tumor cells showed vacuolar, scant cytoplasm and nuclear mitoses. Besides, a few immature cartilage cells with mild atypia could be found in immature cartilage matrix in some field of view (hematoxylin-Eosin ×200). (B) Tumor cells positive to vimentin (×200).

patients presented with cough, chest pain, dyspnea, and hemoptysis. In addition, patients may be asymptomatic during their presentation, and the findings were incidentally detected simply by a chest roentgenogram.2

Although the radiologic manifestations have no differential points from other lung tumors, they can define of the size and location of the tumor and the extent of the adjacent tissues involved. The confirmation is done by microscopic and immunohistochemical studies. Mesenchymal chondrosarcoma is composed of atypical undifferentiated mesenchymal cells and islands of matured chondroid matrix typically presented as bimorphic appearances. The mesenchymal cells with less nuclear atypia and mitotic activity, round or shuttle-like, are uniform with hyperchromatic nuclei, inconspicuous nucleoli, and scant cytoplasm. There are more meshes of the reticular fibers between mesenchymal cells, it is like Ewing’s sarcoma. In addition, the chondrocyte with lacking of atypia may be calcification or ossification in the focus of cartilage, and the size and/or number of the islands of matured chondroid matrix is different in different area in one case or other cases. Immunohistochemically, the lesion is positive for vimentin, CD99 and S-100 in the areas of cartilage, may be positive for neurospecific enolase or leu-7, but negative for desmin, action, cytokeratins, or other epithelial markers. Although it has typical character feature in pathology, clinically, the histological sample collected by fine needle aspiration is usually small, pathologic images may only contain one material or lack cartilage, so the disease is easily to be misdiagnosed as hemangiopericytoma, Ewing’s sarcoma, poorly differentiated synoviosarcoma or other small round cells tumor. In our cases, ultrasound-guided needling biopsy was undiagnosed before the operation.

The optimal therapy for mesenchymal chondrosarcoma of the lung is not established in the current study. Resection, radiation, chemotherapy, or combination of them were used in treating the mesenchymal chondrosarcoma of usual sites. The surgical resection may be the mainstay. The complete surgical resection followed by radiation and/or chemotherapy therapy results better in long-term control of disease than surgery alone.6,7 Tuncer et al.7 reported a case of 5-month-old girl with mesenchymal chondrosarcoma of the orbit who received one course of vincristine, actinomycin-D, and cyclophosphamide and two courses of ifosfamide, epirubicin and cisplatin (IEC), and then radiotherapy, exenteration was carried out. Postoperatively, three courses of IEC chemotherapy were administered. She was alive with no evidence of disease after 4 years of follow-up. The prognosis for patients with mesenchymal chondrosarcoma is generally poor, because it is an aggressive tumor with high potential of local relapse and distant metastases.

Conclusion

Primary mesenchymal chondrosarcoma of the lung is a extremely rare malignant tumor. Patients with primary mesenchymal chondrosarcoma of the lung must be carefully evaluated to rule out a metastatic origin. This disease is easily to be misdiagnosed as hemangiopericytoma, Ewing’s sarcoma, or other small round cell tumors by needling biopsy. The final diagnosis of mesenchymal chondrosarcoma depends on pathology and immunohistochemistry. Multimodality treatment
(surgery, chemotherapy, and radiotherapy) may lead to long-term survival.

Disclosure Statement

This case report do not exist potential conflict of interest.

References