Primary Pulmonary Myxoid Malignant Fibrous Histiocytoma

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Primary pulmonary malignant fibrous histiocytoma (MFH) is a rare mesenchymal neoplasm of the lung. Myxoid variant is an uncommon subtype of primary pulmonary MFH. In this report we present a case of a young adult who had chest discomfort and was found to have a huge mass in the left lung. Left pneumonectomy was performed and a myxoid variant of primary pulmonary MFH was proven by immunohistochemistry. Intercostal color-doppler ultrasound showed a heterogeneous cystic mass with internal echogenicity of the cystic content and thick wall but without obvious blood flow. The chest CT showed a huge cystic mass in the lung parenchyma with mural nodules. MRI demonstrated more findings than CT, including chest wall invasion and characteristic signal intensity of the cystic content compatible with mucin. The imaging findings of this myxoid variant of primary pulmonary sarcoma with cystic change were unique and correlated with pathologic diagnosis.

Key words: Lung neoplasm, malignant fibrous histiocytoma

Primary pulmonary sarcomas are infrequently encountered [1]. Sarcomas in the lung are usually metastatic. Primary sarcomas of the lung are extremely rare and are categorized into three groups by the site of origin: parenchymal, large-vessel, and small-vessel sarcomas. Parenchymal sarcomas consist of malignant fibrous histiocytomas, fibrosarcomas, leiomyosarcomas, and osteosarcomas from different origins [2]. Malignant fibrous histiocytoma (MFH) is the most common malignant soft tissue tumor of older adults, most frequently involving the extremities, retroperitoneum and trunk, but it is rare as a primary tumor of lung [3-5].

Most of the pulmonary sarcomas appear to be a single solid nodule or mass without calcification. They are rarely infiltrative in lung parenchyma or cavitated [2-5]. Myxoid change of the pulmonary sarcomas is very rare [5-7]. We reported a case of primary pulmonary myxoid MFH with cystic change in which findings of ultrasound, computed tomography (CT) and magnetic resonance (MR) imaging have not been reported to our knowledge.

CASE REPORT

A 24-year-old male was rather well previously until one month before this admission. He began to experience fever, chest discomfort, easily fatigue and progressive shortness of breath. Physical examination revealed pale sclera and lip, increased dullness on percussion and decrease of breathing sound over his left hemithorax. A chest radiograph was obtained which showed a huge well-defined opacity about 16 cm in diameter in his left lung. During the admission, the laboratory data showed severe anemia (hemoglobin 8.7 g/dl) and hypoalbuminemia (2.5g/dl). The tumor
markers including alpha-fetoprotein, CEA, beta-HCG and CA 19-9 were normal. Serial imaging studies including abdominal sonography, barium study of upper gastrointestinal tract, chest CT, MR study, and isotope bone scan failed to detect any tumor other than the left pulmonary mass.

Color Doppler ultrasound study through left intercostal space showed a cystic mass with heterogeneous content and irregular mural nodules. There was no detectable signal of blood flow by ultrasound study (Figure 1). There was some echogenic cloud-like component of the cystic tumor. The contrast-enhanced chest CT showed a huge cystic pulmonary mass with irregular thick wall and heterogeneous-density content within the cyst (Figure 2) The area of faintly high density of the cystic content was demonstrated, suggestive of intracystic hemorrhage. No separable fat plane between this mass and chest wall was demonstrated. More characteristic findings of content of the cystic lung mass by the chest MR were demonstrated. The T1-weighted MR image (TR 750/TE 33) showed high signal in the fluid content of this

Figure 1. Color Doppler ultrasound through left intercostal space showed a cystic mass with heterogeneous internal echogenic materials and irregular wall (arrow). The internal echogenic component was “cloudy” like appearance. There was no detectable blood flow noted within this tumor.

Figure 2. Axial enhanced CT scan at the level of inferior pulmonary vein. There was a huge cystic mass with irregular and nodular wall (arrow) in the left lung. There was slightly swelling of the left chest wall.

Figure 3. a. Axial spin-echo T1-weighted image showed inhomogeneous high signal intensity in the central part of the cystic mass. b. Coronal Gadolinium-DTPA enhanced spin-echo T1-weighted image showed intense enhancement of the wall of the cystic mass and mural nodules (arrow). There was tumor invasion of the chest wall, notably in the intercostal space (arrowhead). In the right side chest wall, there was normal intermediate signal intensity of the muscles between adjacent ribs.
cystic mass, consistent with proteinaceous and/or hemorrhagic fluid. Gadolinium enhanced T1-weighted images revealed well enhancing mural nodules of the cystic wall (Figure 3). Mediastinal lymph nodes were also noted. The mass was inseparable from the chest wall. Because of the respiratory symptoms related to lung compression, surgery was performed under the impression of a cystic pulmonary neoplasm through median thoracotomy. A huge intrapulmonary mass (15×12×12 cm) in the left thoracic cavity involving most of the upper lobe and the upper half of the lower lobe was found. There was darkish red necrotic tissue and bloody mucoid content in this cystic mass. During surgery, it was found that this tumor invaded left chest wall and left major fissure and was associated with abnormal enlarged mediastinal lymph nodes. Left pneumonectomy with lymph node dissection was then performed and the patient recovered from the surgery in stable condition.

Grossly, the tumor was predominantly cystic with mucoid and gelatinous content associated with some dark brown hemorrhagic fluid. Microscopically, a cellular area was only noted along the wall of the cystic mass, where predominant spindle fibroblasts admixed with some histiocytes arranged in a storiform pattern were found (Figure 4). The immunohistochemical study results were positive for vimentin and antichymotrypsin, and negative for S-100 protein, desmin, actin, myosin and myoglobin. A primary pulmonary myxoid MFH with mediastinal and hilar lymph node metastasis was diagnosed [5].

Adjuvant radiation therapy of the chest was performed after surgery. He also received chemotherapy. The patient experienced no discomfort until he had an episode of back pain five months after tumor resection. Ultrasound study showed rapid abdominal distention due to tumor metastasis of the liver and invasion to inferior vena cava one month later. The follow-up chest and abdominal CT revealed local tumor recurrence, liver metastasis and an adrenal mass. Rapid disease progression, including hemoptysis, respiratory failure, septic shock, occurred in spite of intensive medical care. Unfortunately, he died of multiple organ failure nine months after diagnosis.

**DISCUSSION**

Patients with parenchymal sarcomas of the lung are usually older in age. Primary pulmonary sarcomas in young adults are usually with a grave prognosis [1-8]. MFH is the most common malignant soft tissue tumor of older adults, most frequently involving the extremities, retroperitoneum and trunk. It rarely presents as a primary tumor of lung [5]. The primary pulmonary MFH consists of 0.04% of primary lung cancer [2]. There was slight male predominance with the range of age from 41 to 75 of years (average age of 59 years) according to a review of 16 cases of primary pulmonary MFH [4]. Only one of twenty-two cases (4.5%) of primary pulmonary MFH had been reported as a myxoid variant [7] while the majority was storiform-pleomorphic subtype. In this report, we described the imaging findings of a rare primary pulmonary myxoid MFH in a young adult who passed away nine months after diagnosis. The imaging findings of a primary pulmonary myxoid MFH have not been reported to our knowledge. In this case, the initial finding of chest radiograph was a huge nonspecific left pulmonary mass. The ultrasound findings of myxoid MFH showed a cystic mass devoid of blood flow and associated with heterogeneous echogenic content and nodular cystic wall (Figure 1). The “cloud”-like echogenicity of the internal content of the cystic lesion might be indicative of mucoid component. On CT images, this myxoid MFH was a large cystic mass with heterogeneous fluid density and enhancing wall (Figure 2). The high attenuation

![Figure 4.](image-url)
content in the cyst correlated well with the hemorrhagic fluid. The MR study of this case showed the location of the pulmonary mass and the morphology of the mural nodules. Furthermore, the fluid content consistent with hemorrhagic mucoid fluid by the MR signal and chest wall invasion of the tumor were characteristically depicted by MR (Figure 3A and B). From the imaging findings, it was suggestive of a cystic mucoid pulmonary tumor. Myxoid sarcomas include MFH, leiomyosarcoma, liposarcoma, chondroma, chondrosarcoma, ... etc. [9, 10]. The most common imaging finding of pulmonary sarcoma was mass-like lesion [1,2, 4, 5, 7]. However, large-vessel sarcoma, which are primarily intravascular tumors, arises from within the pulmonary artery. The definite diagnosis should rely on the study of immunohistochemistry [5,10].

Sonography of the chest can differentiate solid from cystic lung lesion if there is good contact between lung lesion and pleura [11]. The depth resolution and limited field of view of the intercostal ultrasound render complete evaluation of lung lesion impossible. CT can be very helpful in the evaluation of tumor location, nodal status and lung parenchymal change, but less sensitive in tissue characterization and detection of chest wall invasion. MRI showed more definite evidence of tumor invasion of chest wall (arrow in Figure 3B). Both CT and MRI can predict resectable tumors involving chest wall, but cannot distinguish inflammatory chest wall adherence from early invasion [12,13]. In this case, tumor extended into the intercostal space on the coronal MR image which was in favor of tumor invasion of the chest wall. Some of the primary pulmonary MFH as other pulmonary sarcomas could have initial chest wall invasion or metastasis to lymph nodes and solid organs [2, 7].

The differential diagnosis of an intrapulmonary fluid-filled cystic lung lesion was few. Bronchogenic cyst and pulmonary sequestration are possible cystic lesions of the lung parenchyma. The majority of bronchogenic cysts occur at the middle mediastinum close to the junction of tracheobronchial tree and carina. In some rare cases, intrapulmonary bronchogenic cyst may appear as a cystic pulmonary lesion usually associated with smooth and thin wall [14, 15]. The CT attenuation of the content of bronchogenic cyst may range from water to soft-tissue density, which depends on the component of the fluid. The characteristic T2 hyperintensity of the content of bronchogenic cyst has been reported associated with smooth or slightly lobulated wall. Pulmonary sequestration is usually found at basal lung with the appearance of consolidation, mass or associated with cystic change [16]. Recurrent pulmonary infection is usually found. Communication with bronchial tree results in the appearance of air cysts or air-fluid layering in a cystic lung lesion. By using spin-echo MRI or MR angiography, an anomalous feeding artery supplying the sequestrated lung is usually visible. Neither bronchogenic cyst nor pulmonary sequestration has the same radiologic picture as this pulmonary sarcoma.

Though surgical resection of primary pulmonary sarcoma is the treatment of choice, this patient died due to multiple metastases and organ failure after tumor resection. The prognostic factors were significantly related to the presence of metastasis, local recurrence and significant tumor necrosis [8].

In conclusion, the imaging findings of this primary pulmonary myxoid MFH were very unique and not been reported in the literature to our knowledge. The definite diagnosis relies on study of immunohistochemistry. A pulmonary parenchymal lesion with mucoid component and irregular and nodular wall at ultrasound, CT or MRI was suggestive of a myxoid pulmonary sarcoma such as myxoid MFH.

REFERENCES

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primary pulmonary myxoid malignant fibrous histiocytoma

原發性肺部黏液性惡性纖維組織瘤

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肺部原發惡性纖維組織瘤（malignant fibrous histiocytoma）為罕見之肺部間質腫瘤，而黏
液性（myxoid）變異形態更罕見。本案例報告一位年輕男性在胸部不適後即出現一在胸部
X光為非特異性巨大左肺腫塊，經左全肺切除後，病理免疫組織學檢查證實為罕見之肺臟原發
惡性纖維組織瘤之黏液變異。術前肋間彩色多普勒超音波為一異質性無血流之肺內囊性腫塊，
電腦斷層則發現一具壁上節結之囊性肺腫瘤，磁振造影發現更具組織特性的黏液狀農號變化於
囊內含物與腫瘤之胸壁侵犯。比較病理結果顯示，此種具異性之原發性肺臟黏液惡性纖維
組織瘤有特殊影像發現並具診斷價值。

關鍵詞：肺腫瘤、惡性纖維組織瘤