Primary myelolipoma of the lung: a case report and review of literature

Wenbin Huang¹, Youcai Zhao¹, Xindao Yin², Qiong Qi¹

¹Department of Pathology, Nanjing Medical University Affiliated Nanjing Hospital (Nanjing First Hospital), China
²Department of Medical Image, Nanjing Medical University Affiliated Nanjing Hospital (Nanjing First Hospital), China

Myelolipoma of the lung is a rare benign tumor composed of mature adipose tissue and normal hematopoietic cells. Only 8 cases of primary myelolipoma of the lung have been reported up to date. We report here the ninth case which was misdiagnosed as a malignant tumor and relevant literature is reviewed.

Key words: lung, myelolipoma.

Introduction

Myelolipoma is a rare benign tumor of mesenchymal origin which consists of mature adipose tissue and normal hematopoietic cells. It occurs most often in adrenal gland [1]. Extraadrenal myelolipoma is very rare. They have been reported in the presacral region [2], mediastinum [3], retroperitoneum [4], liver [5], spleen [6], stomach [7], leptomeninges, and thoracic cavity including lung. To the best of our knowledge, only 8 cases of primary myelolipoma of the lung have been reported [8-13]. In the present paper, we report a case of pulmonary myelolipoma misdiagnosed as a malignant tumor before surgery.

Case report

A 57-year-old woman was admitted to our hospital with fever for thirteen days. The patient presented with intermittent fever up to 38.6°C. She felt chills and had no cough or sputum. CT scanning at a local hospital revealed bronchiectasis with infection in the lower lobe of the right lung and a small nodule of the right lung (Fig. 1). After she was treated with some drugs such as patulin and antibiotics, she had a CT scan performed again. The second CT scan showed that there was a massive shadow in the right lower lobe. She was clinically diagnosed with lung cancer and the right lower lobectomy with the assisted thoracoscope was performed. The frozen section during an intraoperative consultation revealed benign mature adipose tissue and hematopoietic cells characteristic of myelolipoma.

Pathological findings

Grossly, the excised lower right lobe (segmentectomy) measured 13 cm × 10 cm × 2.5 cm and a circumscribed, grey-red nodule measuring 1.6 cm in the greatest diameter was observed on the cut surface.

Microscopically, the lesion was located in the vicinity of bronchiole and was well-demarcated on low power (Fig. 2). The tumor was composed of mature adipose tissue and hematopoietic elements including myeloid cells, erythroid cells, and megakaryocytes (Fig. 3). The proportion of adipose and hematopoietic tissue was approximately equal. The hematopoietic tissue displayed normal maturation. No trabecular bone, and calcification or ossification was seen in the tumor.

Discussion

Myelolipomas were first described at the beginning of the 20th century. Although myelolipomas are predominantly associated with adrenal glands, extraadrenal myelolipomas are also reported. Extraadrenal myelolipoma is very rare. Since 2006, only 37 cases have been reported in the English-language literature. The sites of extraadrenal myelolipoma have been described in the presacral soft tissue, retroperitoneum, pelvis,
mesentery, spleen, liver, stomach, mediastinum, and leptomeninges [2-7]. Only eight cases of myelolipoma arising from the lung have been reported [8-13]. Of the previously reported cases, five of them were solitary, and two were multifocal. The mean age at presentation is 60 years (ranging from 45 to 81). The sex distribution is predominantly male. The patients usually present with pneumonia, with one case presenting with bronchial carcinoid [10]. CT scanning showed a completely or partially encapsulated mass which density depends on the relative proportion of fat (low density) versus hematopoietic tissue (high density).

Pulmonary myelolipomas including the present case are mostly small nodules of less than 2.5 cm in diameter except for a case of myelolipoma measuring 7 cm in the greatest diameter [9]. Histologically, the tumors resemble myelolipomas in the adrenal gland, which consist of different ratios of mature adipose tissue and hematopoietic cells including myeloid, erythroid, and megakaryocytic elements, and occasionally lymphocytes. Immunohistochemical and special staining may demonstrate various hematopoietic line cells. Areas of internal hemorrhage, calcification, small spicules, and metaplastic cartilages are found in the tumor.

Pulmonary myelolipoma should be distinguished from extramedullary hematopoiesis and hamartoma. Extramedullary hematopoiesis usually occurs as a manifestation of myeloproliferative diseases or is a compensatory phenomenon in various chronic anemias. In contrast to myelolipoma, extramedullary hematopoiesis of the lung frequently presents with multiple occurrences and ill-defined lesions. Microscopically, extramedullary hematopoiesis is composed predominantly of hematopoietic cells and erythroid hyperplasia. The lesion typically lacks lymphoid cell aggregates and there is no or only little adipose tissue. Lung hamartomas generally consist of cartilage, fat, and connective tissue [14]. Some cases may contain smooth muscle and bone. No hematopoietic cells can be seen within the hamartoma. When myelolipoma is composed predominantly of fat tissue, it is difficult to distinguish it from lipoma. Because myelolipoma always has hematopoietic cells, it is necessary to obtain more samples to avoid misdiagnosis as a lipoma.

Although several hypotheses have been proposed as to the cause of myelolipoma, at present the exact pathogenesis of myelolipoma still remains unclear. Historically, there are 3 theories of the pathogenesis of myelolipomas. The first is that myelolipomas are derived from bone marrow emboli that lodge in the adrenal gland or other sites. The second theory suggests that myelolipomas are derived from embryonic primitive mesenchymal cells. The third theory suggests that myelolipomas arise from metaplastic transformation of adrenal (or other sites) stromal cells in response to stimuli. However, Bioshop et al. found that both the hematopoietic elements and the fat have the same
pattern of X-chromosome inactivation in the majority of cases, which suggests that the majority of these entities are clonal proliferations and are derived from a common, pluripotential stem cell [15].

Given the likelihood of subsequent hemorrhage or compressing of adjacent bronchus by myelolipomas, surgical removal is the best choice. No recurrence or malignant transformation has been reported, so conservative resection is recommended.

References


Address for correspondence

Wenbin Huang
No. 68, Changle Road, Nanjing, China 210006
tel. 86-25-52271192
fax 86-25-52271192
e-mail: wbhuang348912@126.com