Case report - Pulmonary
Low-grade pulmonary myxoid liposarcoma

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Abstract

Primary pulmonary liposarcoma is extremely rare. There are only 12 cases reported in the literature. We presented a case of low-grade primary pulmonary myxoid liposarcoma, which was diagnosed and underwent surgery in our clinic. The diagnosis was established after imaging investigations (X-ray and CT), histologic and immunohistochemical examinations. The patient was followed up one year after the operation.

Keywords: Pulmonary liposarcoma; Primary; Myxoid

1. Introduction

Primary pulmonary liposarcomas are mesenchymal tumors accounting for 3% of all lung neoplasms [1,2]. The histologic types described include fibrosarcoma, leiomyosarcoma, rhabdomyosarcoma, hemangiopericytoma, malignant schwanoma and others. Liposarcoma is one of the rarest types of primary pulmonary sarcomas. There are only 12 cases reported in the literature [3–5].

2. Case report

We presented a case of a 49-year-old man who had complained of dry cough, right-sided chest pain and a subfebrile temperature. The laboratory findings of blood and urine tests were within the normal ranges. The X-ray examination of the lungs revealed a strong shadow firmly delineated, 20 mm in size, located in the 4th right intercostal space. The CT scan investigation of the lungs showed a low-density lesion 25HE, situated in the right lung dorsally, which measured 16 mm. The mediastinal lymph nodes, the abdominal cavity and retroperitoneal space were not involved (Fig. 1). The probable conclusion of the CT scan examination was pulmonary echinococcosis. The physical examination of the upper and lower extremities showed no abnormal changes.

The patient underwent surgery. Right-sided thoracotomy was performed. The operative finding was: well-outlined solid tumor formation 15/10/10 mm in size, located in the 2nd segment of the right lung. Segmentectomy of the involved segment was performed. The urgent examination of the species revealed inflammatory pseudotumor of the lung.

3. Pathological finding

At gross examination the tumor was a well-circumscribed, solid nodule 15/10/10 mm in size, with yellowish outer surface, situated subpleurally in the periphery of the pulmonary parenchyma. For the histologic examination, we used not only the traditional method with hematoxylin and eosin-stained sections, but also histochemical and immunohistochemical methods (Fig. 2).
The histological diagnosis was low-grade pulmonary myxoid liposarcoma. The lesion consisted of pleomorphic (round or spindle-shaped) cells amongst myxoid stroma, plexiform capillary net, scattered multivascular lipoblast. There were microcysts filled with a mucoid substance in the tumor parenchyma. There were a lot of lymphocytes, which formed lymphoid follicles in the tumor stroma. No necrosis was seen in the samples. Mitoses were 3–5/10 HPF (High Power Field). The proliferative activity of the tumor cells, examined by Ki-67 was less than 5% in 10 HPF. Histochemical staining for lipids and mucopolysaccharides (Sudan III and PAS reaction) were positive.

The postoperative period was uneventful. The patient was followed up one year after the operation; he had no subjective complaints; CT scan of the lungs showed no pathologic changes.

4. Discussion

Liposarcoma is the commonest soft tissue tumor in adults. It is seen more frequently in the lower extremities or in the retroperitoneal space. The primary thoracic liposarcoma is rare. It affects more frequently the mediastinum, but sporadic cases involving the lung parenchyma, pleura and chest wall have also been reported. Possible pathogenetic factors for the development of primary pulmonary liposarcoma are malignant degeneration of pulmonary lipoma or pleuropulmonary atelectasis [4,6]; we did not find data for that in our case. The literature review shows that the primary pulmonary liposarcoma has no sex predilection with a reported patient age range of 9–59 years [6].

Liposarcoma is subclassified histologically into well-differentiated, myxoid, round-cell, pleomorphic, dedifferentiated and mixed type. In accordance with the TNM classification of the lung tumors from 1997, the tumor in our case was stage T1N0M0. When using the FNCLCC grading system, and the following criteria – tumor differentiation, mitotic count and tumor necrosis, the lesion in our case was Grade 1 (total score 3). After detailed examination of the described liposarcoma, we considered that the cell differentiation, the mitotic index, the absence of necrosis and the proliferative activity of the tumor cells, defined the lesion as low grade myxoid liposarcoma. The abundant lymphocytes infiltration in the stroma of the tumor with the formation of lymphoid follicles was an interesting finding, which was not previously reported; that is why the result of the urgent pathological examination was inflammatory pseudotumor.

The primary pulmonary liposarcoma often causes diagnostic difficulties. It demonstrates nonspecific findings at conventional radiography. CT appearance differs depending on the amount of fat tissue within the formation [6]; the probable diagnosis in our case was hydatid cyst.

The most important differential diagnosis of primary pulmonary sarcoma is metastatic spread from an extrapulmonary sarcoma [7] as well as all tumors and tumor-like processes, which appear as well-demarcated solid nodules in the lungs. The correct diagnosis is pathomorphologic; the most important examination is the immunohistochemistry and the examination of the proliferative activity of the tumor cells.

Myxoid liposarcoma presents typically as a slow growing mass; there is a risk of recurrence, and metastases are rare. Based on the reports in the literature, the tumor responds poorly to both chemotherapy and radiotherapy. The treatment is operative-wide complete excision [2,5,8].

In conclusion, the primary pulmonary liposarcoma is a rare tumor with difficult preliminary diagnosis. The prognosis depends on the precise pathologic diagnosis, the stage of making the diagnosis and the completion of the surgical intervention.

References