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(Citation)
Molecular and Clinical Oncology,12(3):244-246

(Issue Date)
2020-03

(Resource Type)
journal article

(Version)
Version of Record

(URL)
https://hdl.handle.net/20.500.14094/90007316

A case of leiomyosarcoma originating from a bronchogenic cyst: A case report

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Received July 1, 2019; Accepted January 2, 2020

DOI: 10.3892/mco.2020.1977

Abstract. Mediastinal leiomyosarcoma is an aggressive tumor that primarily occurs in the posterior mediastinum. A bronchogenic cyst is a benign lesion that often develops in the mediastinum close to the thoracic trachea and has been reported to be the origin of certain malignancies. The present study reports an extremely rare case of an anterior mediastinal leiomyosarcoma that was considered to have originated from a bronchogenic cyst. An 82-year-old woman complained of chest pain, presenting an abnormal 70 mm mass shadow in the anterior mediastinum of a chest CT scan. Mediastinal tumor resection was performed and the tumor was diagnosed as a leiomyosarcoma. As the tumor was located adjacent to a bronchogenic cyst, it was considered to have originated from the remnant tissue of the cyst. Recurrent lesions were noted in the right lower lobe on CT scan 24 months after surgery.

Introduction

Primary sarcomas of the mediastinum have been reported to be rare, and leiomyosarcomas account for approximately 9% of the mediastinal sarcomas (1). A leiomyosarcoma is an aggressive malignant tumor that originates in the mesenchymal tissue (2). Mediastinal leiomyosarcomas mostly occur in mediastinal organs, such as the heart, great vessels, and esophagus, usually manifesting as posterior mediastinal tumors, and rarely as anterior mediastinal tumors (3). Only few cases of anterior mediastinal leiomyosarcomas have been reported to date (3). Here we report an extremely rare case of an anterior mediastinal leiomyosarcoma that was considered to have originated from a bronchogenic cyst.

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Key words: leiomyosarcoma, bronchogenic cyst, mediastinum, sarcomas, mesenchymal tissue

Case report

An 82-year-old woman with a medical history of hypertension presented to us with chest pain. Her chest radiograph revealed a giant mass in the left lung field, and chest CT scan revealed a 70-mm mass in the anterior mediastinum (Fig. 1A). Positron emission tomography-CT scan revealed strong fluorodeoxyglucose accumulation in the tumor (maximum standardized uptake value, 10.9), and no other lesion was detected (Fig. 1B). A needle biopsy of the tumor was not performed owing to the possibility of tumor implantation. Mediastinal tumor resection was performed via the median sternotomy approach. The tumor adhered to the aorta, pericardium, and left upper lung. As tumor infiltration of the left upper lung was suspected, a part of the left upper lobe and the thymus was resected along with the tumor. The operation time was 233 min, and bleeding was 325 ml. The postoperative course was uneventful, and the patient was discharged 15 days postoperatively. Pathological examination showed that the tumor was firm, white-to-yellowish in color, and 65 mm in size; furthermore, it, had a well-demarcated and fibrous capsule (Fig. 2A-C). A cystic cleft was observed in the tumor periphery. Histologically, the tumor comprised eosinophilic spindle cell bundles with focal necrosis and exhibited increased mitotic activity (three/high-power field) (Fig. 2D). On immunohistochemical staining, the tumor was positive for alpha smooth muscle actin and desmin, and negative for cytokeratin, S-100 protein, AE1/3, Cam5.2, and CD34 (Fig. 3). Based on these findings, the tumor was finally diagnosed as a leiomyosarcoma. Leiomyosarcomas, which are present as anterior mediastinal tumors, are extremely rare. The tumor was located adjacent to the cyst wall, which was covered with ciliated columnar epithelium (Fig. 2A-C); therefore, the tumor was suspected to have originated from the remnant tissue of the bronchogenic cyst. The patient's chest pain disappeared after the surgery. Recurrent lesions were noted in the right lower lobe on a CT scan performed 24 months after the surgery. Because the effectiveness of radiation and chemotherapy for leiomyosarcoma remained unclear, aggressive treatments were thought to be difficult considering her age; she now receives best supportive care.

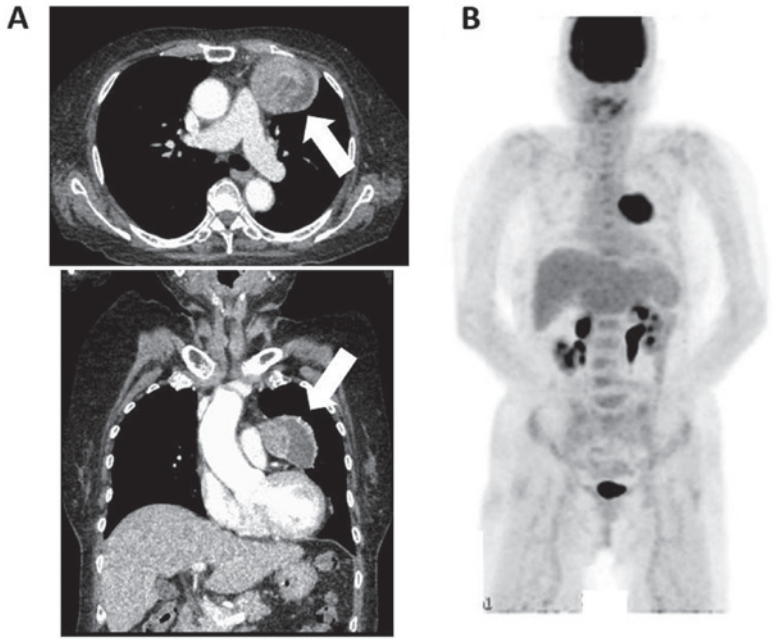

Figure 1. Clinical presentation of the tumor. (A) CT scan presenting a 70 mm mass in the left anterior mediastinum. (B) Positron emission tomography-CT scan revealing strong fluorodeoxyglucose accumulation in the tumor (maximum standardized uptake value, 10.9). White arrows indicate the tumor location.

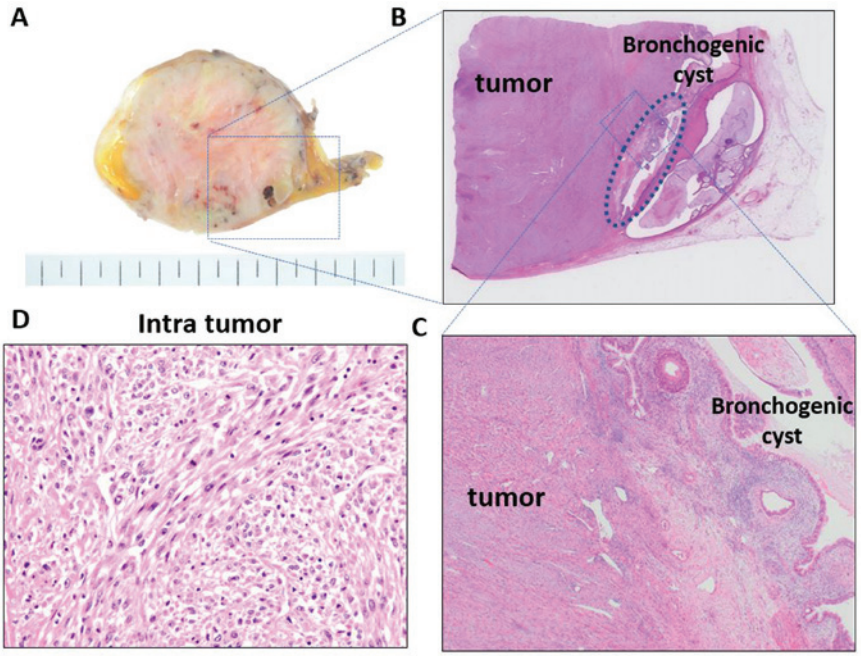

Figure 2. Pathological examination of the tumor. (A) Macroscopic image of the resected tumor. The tumor was located adjacent to a bronchogenic cyst and considered to have originated from the cyst wall. Hematoxylin and eosin staining of the tumor at (B) x10, (C) x40 and (D) x400 magnifications are presented.

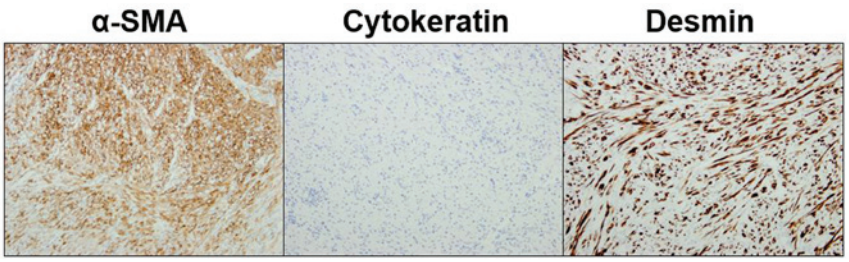

Figure 3. Following immunohistochemical staining (magnification, x200), the tumor stained positively for alpha smooth muscle actin and desmin, and negatively for cytokeratin.

Discussion

We experienced a rare case of a leiomyosarcoma that originated from the wall of a bronchogenic cyst. So far, less than 40 cases of mediastinal leiomyosarcomas have been reported, with most of them occurred in the posterior mediastinum (2). Leiomyosarcomas presenting as anterior mediastinal tumors are extremely rare. Mediastinal leiomyosarcomas seem to originate from major vessels in some cases; however, the actual site of origin is often unclear (2). In this case, there was no tissue connection between the tumor and the wall of major vessels. Bernheim *et al* (4) have reported a case of an anterior mediastinal leiomyosarcoma that was suspected to have originated from the wall of a bronchogenic cyst, similar to our case. A bronchogenic cyst is a benign lesion that often occurs in the mediastinum close to the thoracic trachea, and it has been reported to be the origin of certain malignancies (5). To our knowledge, this is the second case report to show the simultaneous development of a bronchogenic cyst and an anterior mediastinal leiomyosarcoma.

Mediastinal leiomyosarcomas are aggressive, and local recurrence and distant metastasis have been reported to occur in a large number of patients (2). Patients with mediastinal leiomyosarcomas present with symptoms (such as cough, dyspnea, chest pain, fever) because of the local mass effect (6). The principal treatment for a leiomyosarcoma is complete surgical resection; the effectiveness of chemotherapy and radiotherapy remains unclear (1). The most common site of metastasis has been reported to be the lung, followed by the liver and skin (7). Recurrence is more frequent within 2-3 years of diagnosis, and the 5-year survival rate after complete resection has been reported to be 15-20% (8). The risk factors for poor prognosis of leiomyosarcoma are old age and large tumor size (7). The present patient had both these risk factors; hence, she was considered a high-risk patient with regard to survival.

We described the case of an elderly woman with an anterior mediastinal leiomyosarcoma. The tumor was suspected to originate from the remnant tissue of a bronchogenic cyst. The findings of the present case might help in the assessment and treatment of patients with a leiomyosarcoma in the anterior mediastinum.

Acknowledgements

Not applicable.

Funding

No funding was received.

Availability of data and materials

All data generated or analyzed during the present study are included in this published article.

Authors' contributions

KK and HO collaborated in the conception of the present study. NJ prepared the pictures presented in the figures. YM, DH, NJ and YT critically revised the manuscript and were involved in data interpretation. All authors contributed to writing the manuscript and approved the final version.

Ethics approval and consent to participate

Written informed consent was obtained from the patient prior to enrollment.

Patient consent for publication

Written informed consent was obtained from the patient for the publication of all accompanying images.

Competing interests

The authors declare that they have no competing interests.

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