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Sclerosing hemangioma of the lung with mediastinal lymph node metastasis mimicking lung cancer: a case report

Mediastinal lenf nodu metastazı olan akciğer kanserini taklit eden akciğerin sklerozan hemanjiyomu: Olgu sunumu

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Sclerosing hemangiomas of the lung are very rare tumors and they are often considered benign. In this article, we report a case of sclerosing hemangiomas of the lung in a 32-year-old female case. She was admitted with complaints of progressive cough and thoracic pain for the past three months. Thoracic computed tomography showed a large tumor, approximately 9x10x10 cm in diameter involving the upper and lower lobes in the left sided parahilar region of the left lung. Left thoracotomy was performed and the tumor was seemed as lung cancer in surgical exploration. The mass was removed by left pneumonectomy, followed by systematic mediastinal lymph node dissection. The histopathological examination was reported as a sclerosing hemangioma of the lung. There was metastatic involvement in one station (aortic window, no: 5) of the lymph node. Anatomic resection with systematic mediastinal lymphadenectomy should be considered in such cases to avoid possible recurrences or metastasis.

Keywords: Metastasis; sclerosing hemangioma; surgery.

Akciğerin sklerozan hemanjiyomları çok nadir görülen tümörler olup, genellikle iyi huylu olarak kabul edilir. Bu yazıda akciğerinde sklerozan hemanjiyomu olan 32 yaşında bir kadın olgu sunuldu. Hasta son üç aydır artan öksürük ve göğüs ağrısı yakınmaları ile başvurdu. Toraks bilgisayarlı tomografisinde solda, parahiler bölgede alt ve üst lobu etkileyen yaklaşık 9x10x10 cm boyutlarında büyük bir tümör görüldü. Sol torakotomi yapıldı ve cerrahi eksplorasyonda tümör akciğer kanseri görünümü veriyordu. Sistematik mediastinal lenf nodu diseksiyonunu takiben kitle sol pnömonektomi ile çıkarıldı. Histopatolojik inceleme akciğer sklerozan hemanjiyomu olarak bildirildi. Lenf nodu incelemelerinde bir istasyonda (aortikopulmoner pencere, 5 nolu istasyon) metastatik tutulum var idi. Muhtemel nüksleri veya metastazı önlemek için sistematik mediastinal lenf nodu diseksiyonu ile birlikte anatomik rezeksiyon akılda tutulmalıdır.

Anahtar sözcükler: Metastaz; sklerozan hemanjiyom; cerrahi.

Sclerosing hemangioma (SH) of the lung is a rare, benign tumor that was first described in 1956 by Liebow and Hubbell.[1] On a histological examination, SHs contain epithelioid (solid), papillary, sclerotic, hemorrhagic patterns. Although the morphology of pulmonary SH has been thoroughly described, the clinicopathological features and treatment outcomes of these tumors have not yet been fully elucidated.[2] Hence, the optimal therapeutic approach for these lesions remains unclear.

CASE REPORT

Herein, we report the case of a 32-year-old, non-smoking female with a three-month history of left-sided thoracic pain and a cough without sputum who was otherwise asymptomatic.

A thoracic roentgenogram and chest computed tomography (CT) revealed an irregularly circumscribed, lobulated tumor measuring approximately 9x10x10 cm in diameter that was

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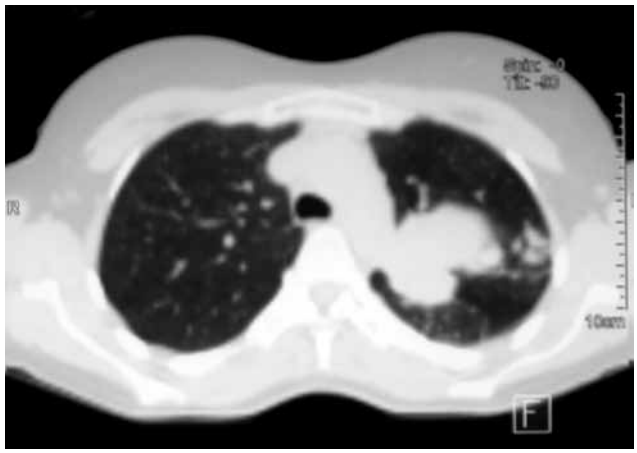

Figure 1. Chest computed tomography shows an irregularly circumscribed, lobulated large tumor in the left side of the parahilar region.

affecting the upper and lower lung lobes in the left side of the parahilar region (Figure 1). To evaluate this lesion, a whole-body 18-Fluoro-deoxyglucose positron emission tomography (FDG-PET) scan was performed which identified a hypermetabolic lesion standard uptake volume (SUV) of 3.8 at the left pulmonary hilum. However, no other abnormalities were found. A bronchoscopy revealed minimal bronchial narrowing in the left upper and lower lobes of the bronchus that this situation coused by external bronchial pressure. A biopsy and bronchial lavage were also conducted, but these revealed no definitive histopathological diagnoses.

Since we could not rule out the possibility of a malignant tumor, we then performed an exploratory thoracotomy that revealed a large, lobulated, solid mass in the hilar and parahilar regions that was affecting both the upper and lower lobes. The histology of the tumor could not be determined clearly by frozen section analysis, but a macroscopic examination showed that it was white and sclerotic, suggesting a malignant lung tumor. Because we could not determine the tumor size and settlement area via a sleeve lobectomy, a pneumonectomy was performed on the left side followed by a systematic mediastinal lymph node dissection.

A histopathological study diagnosed the tumor as pulmonary SH with multiple lung involvement and mediastinal lymph node metastasis. The tumor was composed of round stromal tumor cells that formed solid sheets, and hematoxylin-eosin (H-E) staining (x100) showed that the stalk of the papillary projections was lined with cuboidal surface cells (Figure 2). In addition, immunohistochemical staining (x10) was positive for pancytokeratin in the surface cells of the SH, and thyroid transcription factor-1 (TTF-1) in the surface and round cells of the tumor.

Hematoxylin and eosin staining was also used to determine that there was a metastatic focus in the aorticopulmonary window lymph node, but there were no metastatic invasions in the other lymph nodes in the mediastinum. The metastatic cells of the SH also tested positive for the epithelial membrane antigen (EMA) in the lymph node on an immunohistochemical investigation (x5) (Figure 3).

We consulted the case with the medical and radiation oncology departments about whether the patient should receive adjuvant chemotherapy and/

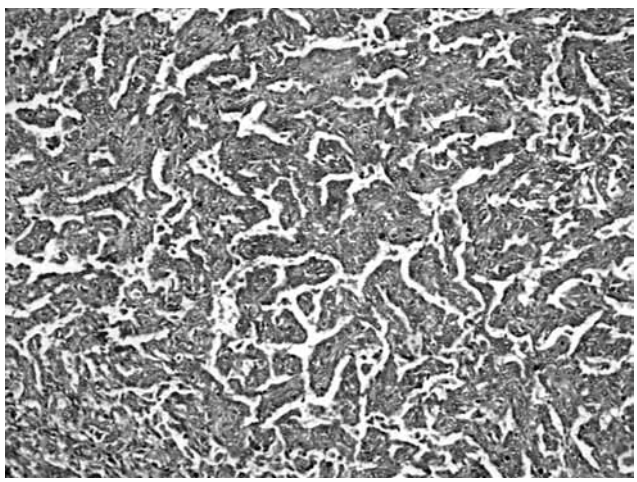

Figure 2. The tumor was composed of round stromal cells that formed solid sheets, and the stalk of the papillary projections was lined with cuboidal surface cells (H-E x 100).

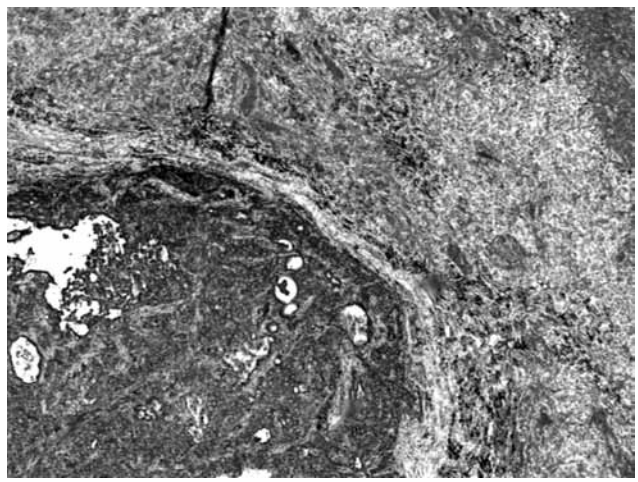

Figure 3. The metastatic cells of the sclerosing hemangioma tested positive for epithelial membrane antigen in the mediastinal lymph node (x5).

or radiotherapy because so little evidence is available regarding SH in the medical literature, not only in the pulmonary organs, but in others as well. In addition, our investigation found no studies on adjuvant therapy and SH. Therefore, we decided that the patient should be followed up and that no adjuvant therapy of any kind would be given after the operation. Three years have now passed, and we have detected no recurrence or metastasis.

DISCUSSION

Sclerosing hemangioma of the lung is a rare tumor that most frequently occurs in middle-aged women, peaking in the fifth decade of life. Furthermore, this tumor is most frequently discovered in Asian individuals. It is more commonly found on the right side, but cases with multiple lesions in both lungs have also been described. However, in our patient, the tumor was located in the central and parahilar regions in the left lung.

Most SH lung tumors arise in the peripheral parenchyma, especially in the subpleura, and are asymptomatic. They primarily are found incidentally (80%) until the time of diagnosis.[2] Moreover, chest X-rays have determined that SHs are sharply-marked and well-defined. In addition to chest CT, a FDG-PET scan can also be used to differentiate between benign and low-grade malignant lesions, which can aid in the planning of the operation.[3]

Sclerosing hemangioma of the lung is usually benign, with only approximately 2-4% of the cases having nodal metastases. However, malignancy does not appear to affect the prognosis. In a study by Miyagawa-Hayashino et al.,[4] four patients with regional (but not mediastinal) lymph node metastases (interlobar, peribronchial, or hilar) who underwent lobectomies and regional lymph node dissection (hilar and peribronchial) were alive at a mean follow-up of 4.7 years and showed no evidence of residual or recurrent disease. However, as of yet, no studies exist that have focused on systemic metastases or mortality for SH of the lung.

In contrast, there are many reports in the literature of pulmonary SH in patients with mediastinal lymph node metastases. However, Katakura et al.,[5] recently reviewed the literature and found only two case reports, including theirs, of mediastinal lymph node metastasis in pulmonary SH, and like their case, ours had mediastinal lymph node metastases without regional or hilar lymph node metastases.

The majority of pulmonary SHs present as solitary pulmonary nodules, with multiple lesions are very rare. Because of this rarity, the natural course of pulmonary SH that feature multiple tumors is not well understood. It is possible that the biological behavior might differ between solitary and multiple cases, whether there is lymph node metastasis or not, but further analysis involving a larger group of patients is required to verify this hypothesis.

In conclusion, although SHs are benign, the possibility of nodal metastases should be kept in mind, particularly during the work-up of enlarged regional lymph nodes in pulmonary SH. Clinicians and pathologists should remain aware that although SH of the lung with lymph node metastasis and multiple lung involvement is a rare occurrence, it is still possible. Therefore, both anatomic resection and mediastinal systematic lymph node dissection are necessary during surgery so that a detailed histopathological analysis can be undertaken to establish the patient's prognosis. In addition, more patient reports related to the diagnostic procedures are needed regarding this condition since currently there is not any evidence about the behavior of metastatic and multiple SH of the lung.

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