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Case Report

Lymphangioleiomyomatosis-associated spontaneous tension hemopneumothorax in multiple organ involvement tuberculosis

Multi organ tutulumlu tüberosklerozda lenfanjiyoleiyomiyomatozise bağlı spontan tansiyon hemopnömotoraks olgusu

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Abstract

Tuberosclerosis complex is a genetic disorder that can affect several organs (such as the brain, lungs kidneys, eyes, the heart, bones and the skin). Pulmonary lymphangioleiomyomatosis is a disease that is mostly seen among young adult women. It is characterized primarily by the non-neoplastic atypical proliferation of smooth muscle cells in the lung parenchyma. Here, we present a case of lymphangioleiomyomatosis-associated spontaneous hemopneumothorax in a tuberosclerosis patient, which we did not encounter in the literature.

Keywords: tuberoussclerosis, lymphangioleiomyomatosis; hemopneumothorax

Öz

Tuberosklerozis kompleksi, beyin, akciğer, böbrek, göz, kalp, kemik ve deri gibi organları etkileyebilen genetik bir hastalıktır. Pulmoner lenfanjiyoleiyomiyomatozis daha çok genç erişkin bayanlarda görülen bir hastalıktır. Akciğer parankimindeki düz kas hücrelerinin neoplastik olmayan atipik proliferasyonu ile karakterizedir. Biz burada literatürde karşılaşmadığımız tuberosklerozis hastasında lenfanjiyoleiyomiyomatozise bağlı spontan hemopnömotoraks olgusunu sunuyoruz.

Anahtar kelimeler: tuberosklerozis; lenfanjiyoleiyomiyomatozis; hemopnömotoraks

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Introduction

Pulmonary lymphangioleiomyomatosis (LAM) is a disease that is mostly seen among young adult women. It is characterized primarily by the non-neoplastic atypical proliferation of smooth muscle cells in the lung parenchyma. Tuberosclerosis complex (TBS) can be observed in 1-2.3% of the cases. The most common pleural complications in these cases are recurrent pneumothorax and chylothorax (1). Here, we present a case of LAM-associated spontaneous hemopneumothorax in a TBS patient, which we did not encounter in the literature.

Case

The 20-year-old female patient admitted to the emergency room with the complaints of chest pain and shortness of breath. The oxygen saturation of the patient was 70%; the pulse, 115/min; the blood pressure, 80/40 mm/Hg. The PA lung image revealed total pneumothorax and pleural effusion in the right, and that the mediastinum had shifted towards the left hemothorax. The patients underwent emergency tube thoracostomy. Approximately 1000 cc hemorrhagic fluid was drained together with some air. The hemoglobin value in the pleural fluid was 6.5, and the relative hemoglobin (blood) value was measured as 13.1. There were no abnormal findings in the other routine blood tests. The patient had been monitored for 10 years for Tuberosclerosis. The physical examination revealed lesions compatible with sebaceous adenoma in the face, lesions compatible with Shagreen patch in the back and periungual fibroma in the side of the right hand's third finger. The tomographic imaging revealed paraseptal emphysema regions in both lungs, increased ground-glass densities and mosaic pattern appearances (Figure 1). After 5 days, the patient was discharged with recovery.

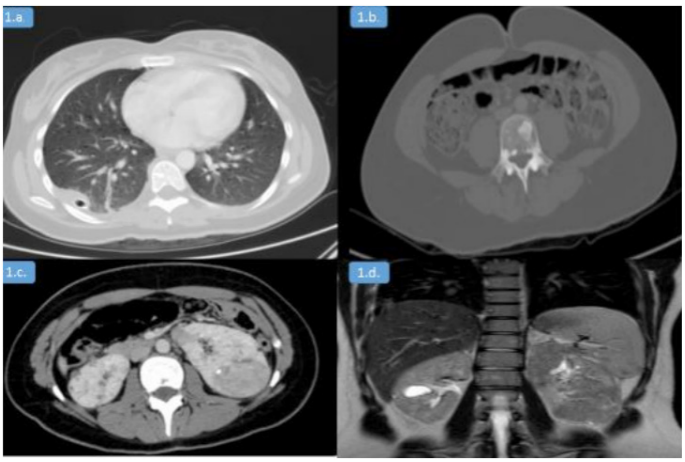

Figure 1a: Thorax CT, paraseptal emphysema regions in both lungs, ground-glass densities and mosaic pattern. **1b:** Abdominal CT sections, sclerotic tubular formations in the body of lumbar vertebra. **1 c. d:** Dynamic MRI examination, angiomyolipomas like solid masses (isointense in T2) in both kidneys that have peripheral contrast retention in the CT with contrast, para-medullary cortical cysts.

The follow-up chest radiography, taken 10 days after discharge, revealed recurrent pneumothorax in the right lung. The patient underwent single-port video-assisted thoracoscopy from the previous drain suture. The thoracoscopy revealed extensive bullous formations and color changes in the parenchyma (subpleural hemangioma type).

The patient underwent apical wedge resection and parietal pleurectomy for pleurodesis. The histopathologic examination of the operation material revealed extensive cystic structures, fusiform cells that proliferated around these cystic formations and the dilated bronchioles, and micronodular pneumocyte hyperplasia foci in bundles. The aforementioned areas were histochemically positive for SMA, HMB45, progesterone receptors, estrogen receptors, CK7 and TTF-1 (Figure 2). There were no postoperative complications and the patient was discharged.

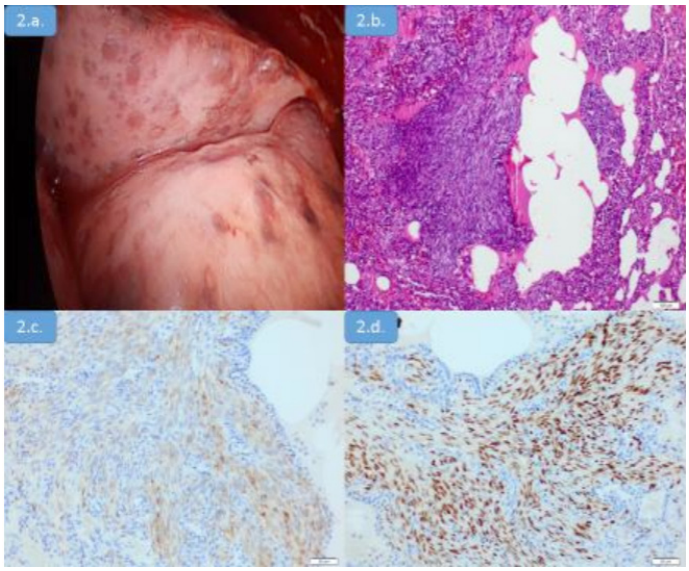

Figure 2a: The thoracoscopic images revealed extensive bullous formations in the lung and color changes compatible with subpleural hemangioma in the parenchyma. **2b:** The light microscopic examination (Hematoxylin-eosin stain, x100), extensive cystic structures, fusiform cells that proliferated around these cystic formations and the dilated bronchioles, and micronodular pneumocyte hyperplasia foci in bundles. **2c:** HMB45-positive lung lesion (avidin biotin peroxidase assay, x 200) . **2d:** Progesterone receptor-positive lung lesion (avidin biotin peroxidase assay, x 200)

Discussion

Tuberosclerosis complex [also known as tuberous sclerosis (TS) and Bourneville disease] is a genetic (autosomal dominant) disorder that can affect several organs (such as the brain, lungs kidneys, eyes, the heart, bones and the skin). The diagnosis can be made according to the criteria stated by the International Association of Tuberculosis (2.3). In this case, the genetic examinations have shown that the DNA had TSC1 (Hamartin,

chromosome 9q32-34) and TSC2 (tuberin, 16p13.3) pathogenic mutations and the diagnosis was confirmed.

The clinical criteria are divided into two groups: major and minor criteria (1). The major criteria include hypomelanotic macules, angiofibromas or fibrous cephalic plaques, fibromas in the nails, Shagreen patches, multiple retinal hamartomas, cortical dysplasia, subependymal nodules, subependymal giant cell astrocytomas, cardiac rhabdomyoma, LAM, angiomyolipomas (AML). The minor criteria include "confetti" skin lesions, enamel holes, intraoral fibromas, retinal acromic patches, multiple renal cysts and non-renal hamartomas. The definitive diagnosis of TBS requires either 2 major criteria or 1 major criterion with 2 minor criteria (1,2).

The TBS patients with lung involvement commonly have lesions with lymphangioleiomyomatosis (TSC-LAM), however, there are reported cases with multifocal micronodular pneumocyte hyperplasia (4). LAM is typically observed among young adult women, which may be associated with estrogen secretion (5). There are sporadic forms of LAM (S-LAM), which are rare and not associated with TSC mutation. These LAM forms can affect the lungs, the lymphatics, the retroperitoneal region and the kidneys, but do not affect the skin, eyes or the central nervous system (5). Clinically, LAM patients are mostly asymptomatic, but the most common symptoms are a chronic cough, hemoptysis, wheezing, chest pain, and effort dyspnea (1,4). The most frequent complaints at the admission are spontaneous pneumothorax and chylous pleural effusion due to lymph node involvement (1,4,6). The spontaneous hemopneumothorax clinic seen in our case has not been found in the literature. Spontaneous hemopneumothorax can develop due to the tear of the adhesion between the parietal and visceral pleura, the rupture of the vascular bullae that lie underneath the lung parenchyma, or the tear of the congenital aberrant veins that are localized in and/or around the bullae in the lung apex (7). In our case, the hemothorax is thought to be caused by the pleural adhesions (observed in the thoracoscopic image), the hemangioma-like vascular structures and bullae.

The thorax CT of LAM patients can show paraseptal emphysema areas, air cysts, ground-glass densities and mosaic patterns (4,5). The pathological examination can be done using bronchoscopic biopsy, transthoracic needle biopsy or open lung biopsies (1). Several changes can be observed in the histopathologic examination; such as the diffuse proliferation of the smooth muscle cells (around the alveolar wall, the bronchi, the lymph ducts and blood vessels) and cystic changes in the pulmonary parenchyma. The immunohistochemical LAM cells are HMB45- and progesterone receptor-positive. It is typical for LAM to visually observe (intraoperative) the multiple and diffuse blebs on the lung surface (1,4,5). The histopathologic examination of our patient has revealed extensive cystic structures, fusiform cells that proliferate around these cystic formations and the dilated bronchioles, and micronodular pneumocyte hyperplasia foci in bundles. The aforementioned areas were histochemically positive for SMA, HMB45, progesterone receptors, estrogen receptors, CK7 and TTF-1.

The kidneys of the TBS can develop AML, fat-density cysts, cortical cysts, and rarely, cancer (8,9). The most dangerous complication of renal AML is bleeding due to vascular rupture (8,9). In our case, the para-medullary cortical cysts and AMLS are shown radiologically shown. There was also a history of emboly in the left kidney due to massive hematuria.

The bone involvement of TBS can include cyst-like lesions, hyperostosis in the internal parts of the calvarial bones, osteoclastic changes, periosteal new bone formations, cystic changes in phalanges and scoliosis. If there are other organ involvements, they are also associated to TBS (8). Our patient had sclerotic tubular formations in the vertebral body (CT image).

The clinical trials among the TBS patients indicate that the sirolimus treatment, mTOR (the mammal target of rapamycin), can provide regression in AML, LAM and brain astrocytomas. The corticosteroid and cytotoxic agent treatments (administered for the improvement of lung functions) were found to have limited benefits (5,10). The literature reports applications of videothoracoscopic wedge resection, limited pleurectomy, and pleurodesis. It is indicated that lung transplant can be effective for the terminal patients, however, it is shown that the LAM cells can migrate to the transplanted lung to cause recurrence(1).

Conclusion

In our case, the 1-year follow-up and the apical wedge resection have indicated that the treatment was sufficient without complications.

Declaration of conflict of interest

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