**Case Report**

**Lymphangioleiomyomatosis-associated spontaneous tension hemopneumothorax in multiple organ involvement tuberculosis**

*Multi organ tutulumlu tüberosklerozda lenfanjiyoleiyomiymatomatözise bağlı spontan tansiyon hemopnömotoraks olgusu*

Bayram METIN*, Mustafa Fatih ERKOC, Sevinç SAHİN, Yavuz Selim INTEPE

1Bozok University Faculty of Medicine, Department of Thorasic Surgery, Yozgat/TURKEY
2Bozok University Faculty of Medicine, Department of Radiology, Yozgat/TURKEY
3Bozok University Faculty of Medicine, Department of Pathology Yozgat/ TURKEY
4Bozok University Faculty of Medicine, Department of Chest Diseases, Yozgat/ TURKEY

**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**


**Anahtar kelimeler:** tuberosklerozis; lenfanjiyoleiyomiymatomatözis; hemopnömotoraks
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. 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.

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, TSC2) mutations. This is a rare condition, and most cases are sporadic. The occurrence of LAM in a TBS patient is even rarer. The case presented here highlights the importance of considering rare conditions in the differential diagnosis of pleural effusions.
chromosome 9q32-34 and TSC2 (tuberin, 16p13.3) pathoge-
nic 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 gi-
ant cell astrocytomas, cardiac rhabdomyoma, LAM, angiom-
yolipomas (AML). The minor criteria include "confetti" skin lesi-
on, enamel holes, intraoral fibromas, retinal acromic patches,
multiple renal cysts and non-renal hamartomas. The definiti-
ve 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 les-
ions with lymphangioleiomyomatosis (TSC-LAM), however,
there are reported cases with multifocal micronodular pne-
umocyte 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 sponta-
neous pneumothorax and chylos pleural effusion due to
lymph node involvement (1,4,6). The spontaneous hemopneu-
emothorax 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 im-
age), the hemangioma-like vascular structures and bullae.

The thorax CT of LAM patients can show paraseptal emphy-
sema areas, air cysts, ground-glass densities and mosaic patterns
(4,5). The pathological examination can be done using bronc-
hoscopical biopsy, transthoracic needle biopsy or open lung
biopsies (1). Several changes can be observed in the histopath-
ologic 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 struc-
tures, fusiform cells that proliferate around these cystic forma-
tions and the dilated bronchioles, and micronodular pneu-
moocyte 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 comp-
lication 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 em-
boly 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, oste-
oclastic changes, periosteal new bone formations, cystic chan-
ges in phalanges and scoliosis. If there are other organ invol-
vements, 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 si-
rolimus treatment, mTOR (the mammal target of rapamycin),
can provide regression in AML, LAM and brain astrocytomas.
The corticosteroid and cytotoxic agent treatments (adminis-
tered 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 recur-
rence(1).

Conclusion

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

Declaration of conflict of interest

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