Baku
LUNG HEALTH CONFERENCE
13-14 October 2023 Radisson Hotel, Baku/Azerbaijan

PS-02

Due to a Case with Pulmonary Arteriovenous Malformation with Delay in Diagnosis: Osler-Weber Rendu Syndrome

Beran Karakoca1, Kadirhan Gürsoy1, Sümeyye Nur Çalışkan1, Esra Geçgel1, Şebnem Durmaz2, Şermin Börekçi1, Akif Turna3

1-İstanbul University-Cerrahpaşa, Cerrahpaşa Faculty of Medicine, Pulmonology Department, İstanbul
2- İstanbul University-Cerrahpaşa, Cerrahpaşa Faculty of Medicine , Radiology Department, İstanbul
3- İstanbul Üniversitesi-Cerrahpaşa, Cerrahpaşa Faculty of Medicine, Thorasic Surgery Department, İstanbul

INTRODUCTION: Osler-Weber-Rendu(OWR) syndrome (Hereditary Hemorrhagic Telangiectasia) is an autosomal dominant disease characterized by telangiectasias in the skin and mucous membranes and arteriovenous malformations in the internal organs. Pulmonary arteriovenous malformation is common in disease with visceral involvement. Curacao diagnostic criteria are used for diagnosis. These are; epistaxis, telangiectasia, visceral lesions and arteriovenous malformations in other organs, and OWR syndrome in first-degree relatives. We wanted to present our case, which has been diagnosed with a delay in diagnosis despite repeated visits to a doctor for 10 years due to tuberculosis contact, complaints of facial numbness and temporary paralysis, in order to draw attention to OWR Syndrome.

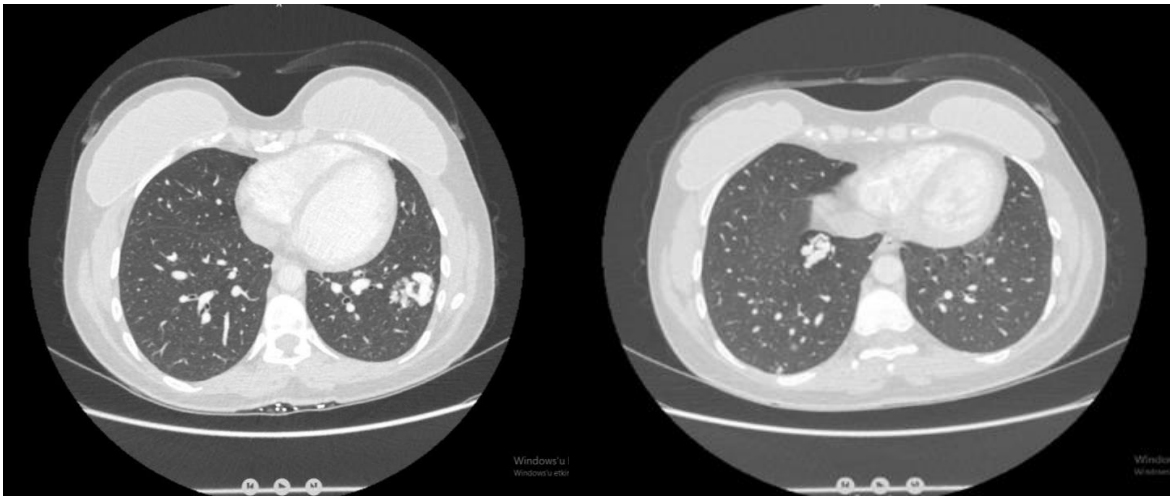
CASE: A 30-year-old female patient with no known history of chronic disease stated that she was screened for exposure to tuberculosis 10 years ago, and at that time a lesion was observed in her lung, but it was said that it might be due to the infection she had. There were repeated doctor applications in the following years, most recently in July 2023, it was evaluated as a transient ischemic attack in an external center due to numbness in the left half of the face, slippage in the mouth, loss of strength in the arms and legs, acetylsalicylic acid was started, and additional hematological evaluation due to polycythemia is normally interpreted. Afterwards, the patient who applied to our outpatient clinic due to exertional dyspnea had a fingertip saturation <90% in room air, and multiple arteriovenous malformations were detected in both lungs on thorax CT angiography (Figure-1 and Figure-2). Areas of ground glass density were observed around the lesion in the superior lower lobe of the left lung (alveolar hemorrhage?). Cranial diffusion MRI revealed a cerebellar infarct area on the left, which may be compatible with embolism, and bilateral embolism sequelae in the posterior system. In the otorhinolaryngological examination of the patient who described intermittent epistaxis, telangiectatic areas were found in the septum and turbinate mucosa in the left nasal cavity, and vascular ectatic areas in the soft palate mucosa. In the abdominal CT, a prominent focus was seen in the arterial phase in the liver. Transthoracic echocardiography was evaluated as natural. Spirometry values were normal and DLCO value was 56%.

Our patient had 4 criteria from Curacao Diagnostic criteria: nosebleed, A-V malformation in the lung, telangiectatic areas in the nasal mucosa and similar findings in first degree relatives, and OWR Syndrome was diagnosed. It was thought that repetitive intracranial embolisms were caused by anastomoses between artery and vein, and the malformation area with frosted glass areas around the left lower lobe was thought to be both hemorrhage area and source of thrombus, and thoracic surgery planned left lower lobectomy for our patient.

CONCLUSION: Although Osler Weber Rendu syndrome is a rare syndrome that appears between 1:5000 and 1:8000, it is a disease that can cause cerebral infarction and stroke with catastrophic consequences if it is not diagnosed and overlooked. For this reason, arteriovenous malformations should be kept in mind in the differential diagnosis of lesions detected in the lung.

Key words: arteriovenous malformation, epistaxis, telangiectasia, iron deficiency anemia, hereditary hemorrhagic telangiectasia, Osler Weber Rendu syndrome

Radiological Images



Picture-1 and Picture-2: Dilated arteriovenous structures associated with each other, compatible with multiple AVMs, are observed in all lobes of both lungs, the largest being in the lower lobe mediobasal on the right (Pic.-1) and the lower lobe anteromediobasal on the left (Pic.-2). In the left lung lower lobe superior, areas of centrilobular ground glass density are observed adjacent to the mentioned structure (alveolar hemorrhage?).

Baku
LUNG HEALTH CONFERENCE
13-14 October 2023 Radisson Hotel, Baku/Azerbaijan

PS-03

Golden Hit in a case of Pulmonary Langerhans Cell Histiocytosis X: Quitting Smoking

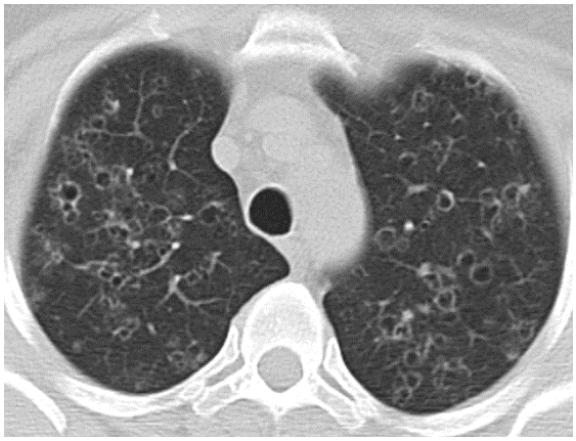
Kadirhan Gürsoy1, Beran Karakoca1, Şebnem Durmaz2, Şermin Börekçi1

1-İstanbul University-Cerrahpaşa, Cerrahpaşa Faculty of Medicine, Pulmonology Department, İstanbul
2- İstanbul University-Cerrahpaşa, Cerrahpaşa Faculty of Medicine, Radiology Department, İstanbul

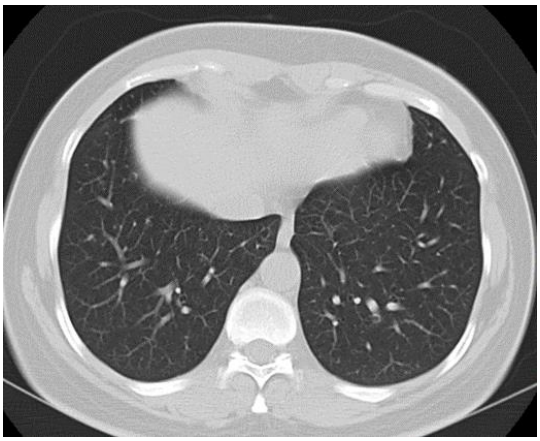
INTRODUCTION: Pulmonary langerhans cell histiocytosis X, a subgroup of langerhans cell histiocytosis, is a rare interstitial lung disease in which langerhans cell infiltration is detected in the lungs, the cause of which has not been fully elucidated. Most of the patients are young, with a dry cough, shortness of breath and a history of spontaneous pneumothorax, more than 90% of them are smokers. Reticulonodular and cystic appearances in the upper and middle zones on high-resolution computer tomography are characteristic. We wanted to present a case of pulmonary langerhans cell histiocytosis, whose cystic lesions regressed significantly after smoking cessation, to emphasize the importance of smoking cessation.

CASE: A 40-year-old female patient presented with back pain and cough in June 2015. In the thorax computed tomography (CT) of the patient, who had a 20 pack-year smoking history and did not use any medication, there were multiple, relatively thick-walled cystic lesions of similar size and appearance, measuring 1 centimeter (cm) in both lungs (Picture-1, Fig. Picture-2). Lower lobe basal segments were seen as preserved. Acute phase values were normal, rheumatological examination was unremarkable, and there was no significant finding except that antinuclear antibody (ANA), one of the collagen tissue markers, was slightly positive as speckled (small grain). The c-ANCA and p-ANCA values of the patient who had no clinical findings in terms of vasculitis were also negative. Abdominal CT was unremarkable, Spirometry and DLCO values were within normal limits. Induced sputum ARB negative, tuberculosis culture did not grow. The patient refused further examination and lung biopsy. Considering the typical thoracic CT findings, age and smoking history, the patient was evaluated as pulmonary langerhans cell histiocytosis. As soon as the diagnosis was made, the patient quit smoking. Due to the absence of clinical signs and normal respiratory functions, no additional treatment was given other than smoking cessation. The patient was followed up with clinical and pulmonary function tests at six-month intervals. In the thorax CT taken 8 years after the first diagnosis of the patient, who continued her daily life without any problems, “In the examination dated June 2015, it was observed that the thick-walled, different sizes and shapes of multiple cysts, more numerous in the upper lobes of both lungs, were totally regressed in the current examination (Picture-3, Picture 4).

CONCLUSION: In patients with pulmonary langerhans cell histiocytosis, smoking cessation is the most important component of the treatment, and in mild cases without complaints, the cysts can completely regress by quitting smoking alone, and the patients can continue their lives without recurrence for many years.

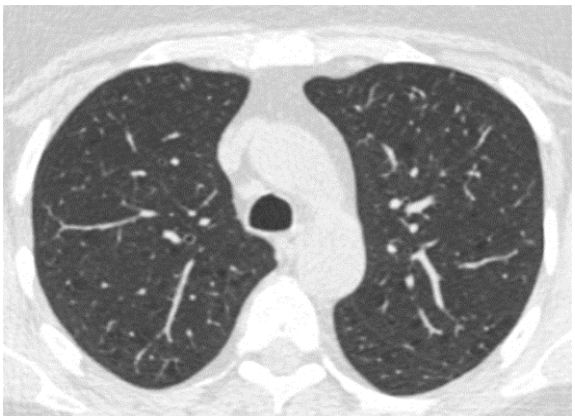


Picture-1

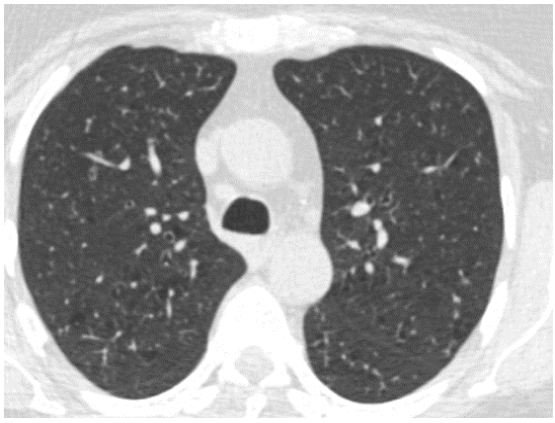


Picture-2

There were multiple, relatively thick-walled cystic lesions up to 1 centimeter (cm) in diameter, similar in size and appearance, in both lungs (Picture-1). Lower lobe basal segments were preserved (Picture-2)



Picture-3



Picture-4

Picture-3, Picture-4: In the examination dated June 2015, it was observed that multiple cysts with thick walls, different sizes and shapes, more numerous in the upper lobes of both lungs, were totally regressed in the current examination.