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RESEARCH ARTICLE OPEN ACCESS

Lymphangioleiomyomatosis

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Abstract:

Lymphangioleiomyomatosis (LAM) is a rare, systemic disease of unknown etiology, which is devastating to women of reproductive age, with a variable course of non-specific symptoms common to many respiratory diseases, and with a poor prognosis. Because it is a rare disease, little is known about its pathogenesis and treatment. It is important to record confirmed cases to have more information on its clinical course. (1)

CASE REPORT:

IDENTIFICATION: The patient SEGL, a 39-year-old healthy female, living in a common-law marriage, evangelical religion, born in Mombaça, living in Fortaleza for 26 years, with high school educational level, working as an operator for 12 years.

MAIN COMPLAINT: “Shortness of breath, pain when breathing, and fever”.

About a year ago, the patient noticed that she was having shortness of breath on great exertion, as after being transferred to another job, she started to walkfour blocks instead of one. She thought this was due to the fact that she was obese. She reports that while accompanying her mother to the hospital, she started to experience several flu-like episodes, with nasal obstruction, coryza, waking at night with shortness of breath, dyspnea at rest,fever, and dry cough, around May or April. These flu-like episodes lasted an average of 10 days, intermixed by 2 to 3 days of improvement. She has had 3 episodes since the beginning. During this period she noticed an increase in

fatigue, while taking care of her mother. The dry cough and chest pain worsened at night, leading to hemoptysis episodes.At the moment she has dyspnea even at rest, but currently has no cough, fever, and accepts her diet well. She has normal sleep-wake cycles. She continues to feel a stabbing pain when she takes deep breaths at the base of the LHT. Herurine is reddish in color and she has not evacuated for 8 days.

PHYSICAL EXAMINATION: The patient is in good overall status, has normal skin color, slightly dyspneic, oriented and cooperative.

PHYSIOLOGICAL ASPECTS:The patient is G1P1A0 and has a history of eclampsia during pregnancy. Before the pregnancy, she had an irregular menstrual cycle and after that,it normalized. She used oral contraceptives (Diane-35) for two years before her pregnancy. Her menarche occurred at 12 years of age, with complete pubarche and telarche. She had her second menstrual cycle at 14 years, and thereafter,she had a cycle of 8 months to 1 year. Psychomotor development:she had a preterm birth with a gestationalage of 29 weeks. Her child is currently 6 years of age and has epilepsy.

FAMILY HISTORY: Her mother had eclampsia while pregnant with her. Both parents smokehand-rolled cigarettes. Her mother has heart disease; the father has cough and chest pain. One aunt died of lung cancer and another of TB. One sister, who is asmoker, had TB and seizure episodes.

SOCIAL HISTORY: The patient was a cigarette smoker from age 25 to 36 years and used to smoke 6 cigarettes a day. She drinks alcohol socially and denies using illicit drugs.

RECENT HISTORY:She denied recent trips, bathing in ponds, rivers and lakes. Her vaccination history is unknown.

PATHOLOGICAL HISTORY:She denied having SAH, DM, TB, asthma, allergies. She reported having had measles, mumps and chicken pox as a child. She received anlodipine (1 tablet/day) during her pregnancy.

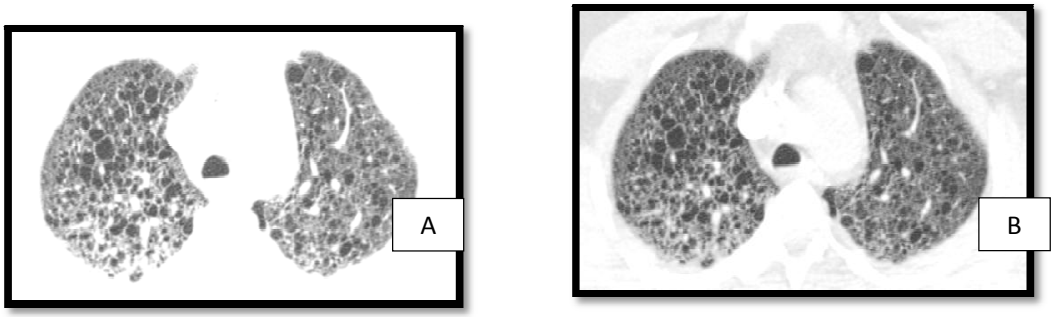

Figure 1: (A) (B) Multiple cystic lesions with thin walls, diffusely distributed in the parenchyma

PHYSICAL EXAMINATION: The patient's overall status was impaired by dyspnea, with abroken speech pattern, using home oxygen therapy, afebrile at the time of the examination, oriented and cooperative. At the moment of the examination, her oxygen saturation was 91%, despite nasal oxygen (2L / min).

Cardiac Auscultation– Regular heart rate 2T, normal heart sounds; Heart rate: 110 bpm.

Pulmonary Auscultation – VM present universally. Expiratory wheezing distributed in both lungs. Absence of crackles.

Abdomen –Round, without palpable enlargements, hydro-aerial sounds present.

Extremities –Slight edema, palpable peripheral pulses.

Keywords: Lymphangioleiomyomatosis; angiomyolipoma; interstitial lung disease; tuberous sclerosis complex.

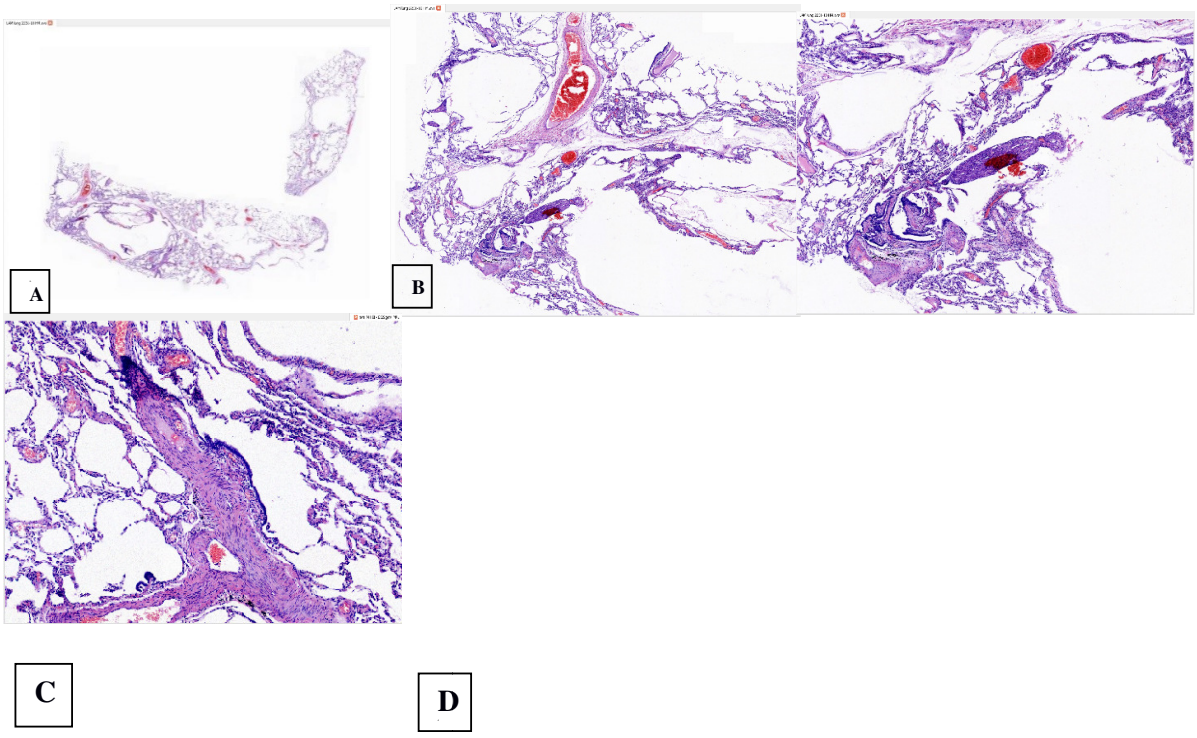

Figure2:

A. Panoramic picture showing pulmonary interstitium with parenchymal cystic dilatations, close to the axial axis;

B. Larger magnification shows thin-walled cysts alongside normal wall structures

C. On one side of the cystic wall, one observes spindle-cell proliferation of young, smooth muscle cells on the internal surface,with an elongated shape.

D. Details of smooth muscle cells and their associationwith the cyst, which arises in part from the obstruction of the bronchioles and the destruction of the alveolar walls.

Discussion:

The diagnosis of lymphangioleiomyomatosis is established by HRCT, where diffusely distributed pulmonary cysts can be observed, and it is almost always abnormal at the time of diagnosis, whereasthe chest x-ray is normal or only shows pleural fluid or pneumothorax(9). The cysts are defined as rounded areas, circumscribed by epithelial or fibrous tissue that form a thin, regular, low-attenuation wall, varying in diameter inside the pulmonary parenchyma, ranging from 2 to 60mm and are usually filled with air. However, they may occasionally be filled by fluid or solid material (6)(10).

A definitive diagnosis of LAM requires a tissue biopsy and an expert pathologist's report. The HRCT may establishthe diagnosis if it shows characteristic findings associated with a renal angiolipoma; in such cases,a biopsy cannot be performed.

LAM is probably an underreported disease that develops only during adulthood, showingprogressiveparenchyma destruction, and often culminating in therapeutic failure and need for lung transplantation. LAM may occur sporadically, but screening studies indicate a frequency of up to 30% of association with Tuberous Sclerosis Complex (TSC).The disease etiology and treatment effectiveness are debatable; however, epilepsy was the most frequent cause of hospital admission in TSC and pneumothorax was the most common cause in patients with LAM. (5)

Sirolimus therapyhas shown a decrease in circulating lymphangioleiomyomatosis cells and a decrease in patients with chylous ascites (6)(7). In some cases, pleurodesis is indicated in recurrent pneumothorax, since it is a chronic pulmonary disease, for which lung transplant is indicated as early as possible.

Conclusion:

Lymphangioleiomyomatosis (LAM) is a rare disease that shares symptoms with many respiratory diseases, in many cases being mistaken with asthma, bronchiolitis etc., of unknown etiology, which makes it necessary for these cases to be notified and published in their most diverse clinical presentation types, so that more knowledge can be acquired about the disease.

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