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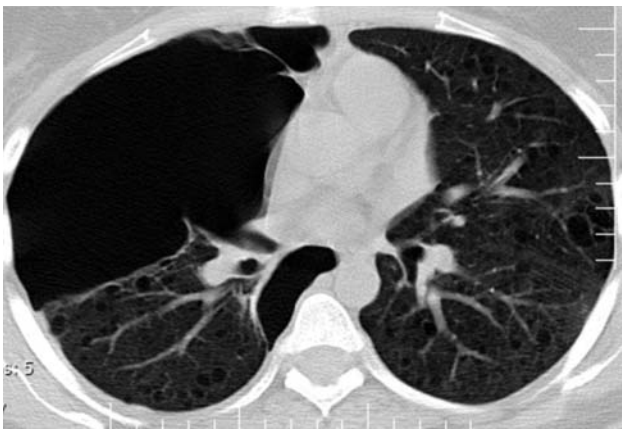


Figure 1. Computed tomography of pneumothorax in lymphangioleiomyomatosis

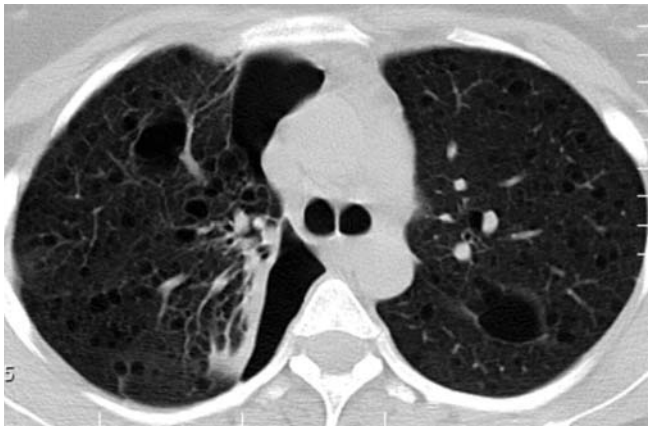
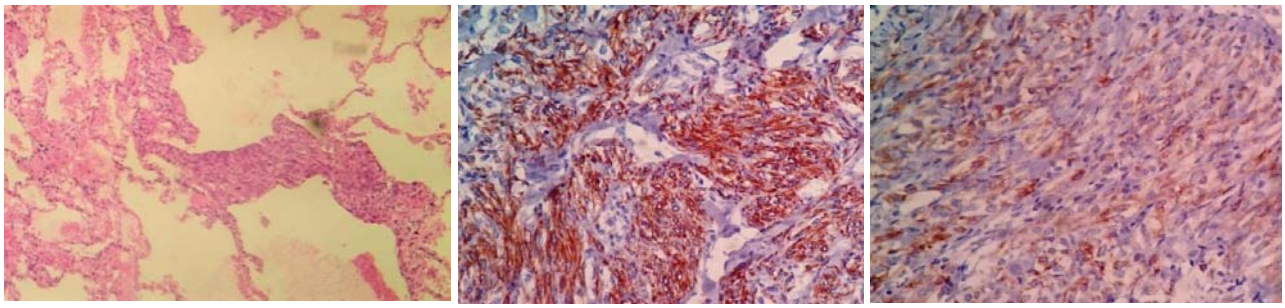


Figure 2. Multiple small cysts penetrating the entire parenchyma of the lung



(A) **(B)** **(C)**

Figure 3. H&E and Immunohistochemical studies (H&E, ×100); **A)** Shows portion of lung tissue alveolar walls disruption with thin walled cystic air spaces and patchy, disordered, nodular proliferation of bland spindled and cuboidal epithelial cell around airways. In IHC proliferating cells were positive for **B)** Desmin; **C)** HMB-45.

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A 29-year-old non-smoker woman presented to emergency department of Shariati Hospital with shortness of breath and right-sided chest pain with dry cough since 4 days ago which was acute in onset and progressive in nature. There was no history of fever, paroxysmal nocturnal dyspnea, palpitation or hemoptysis. She had no drug history. On admission to the hospital, the vital signs were stable with decreased breath sounds on the right side of the chest. Cardiovascular and central nervous systems were unremarkable. An electrocardiogram was normal. Chest X-ray showed right-sided pneumothorax. Subsequent Computed Tomography (CT) scan showed diffuse bilateral cystic lesion in both lungs with severe right pneumothorax (Figures 1 and 2). A tube thoracostomy with a 28F tube was placed. Biopsy from apical segment of the upper lobe of the right lung was taken, which showed a portion of lung tissue with marked hemorrhage in alveolar spaces and alveolar walls disruption with thin walled cystic air spaces and patchy, disordered, clustered to nodular proliferation of bland spindled and cuboidal epithelial cell around airways, lymphatic and blood vessels. On Immunohistochemistry (IHC), the proliferating cells were positive for HMB-45/Desmin (Figure 3).

What is your diagnosis?
See the next page for diagnosis

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Photoclinic Diagnosis: Sudden Onset of Dyspnea in Healthy Woman: Lymphangiomyomatosis

Lymphangiomyomatosis (LAM) is a rare, progressive cystic destruction of the lung seen in women of the reproductive age.[1] The prevalence of this disease is estimated to be about 2:1,000,000.[2] Lung involvement in LAM is due to proliferation of smooth muscle cells that invade all lung structures including the lymphatic, air way, blood vessels and interstitial spaces which lead to progressive dyspnea, recurrent pneumothorax and hemoptysis.[3]

This disease can be sporadic (S-LAM) or associated with tuberous sclerosis (TSC-LAM). LAM occurs in about 30% of females with tuberous sclerosis complex but TSC-LAM constitutes only 15% of LAM patients.[4]

The clinical symptoms of LAM include fatigue, cough, chest pain, hemoptysis, chylothorax and chylous ascites. The most common symptom is shortness of breath. Recurrent pneumothorax occurs in 60% – 70% of LAM-patients.[5] LAM is sometimes diagnosed by chest CT scans in patients with spontaneous pneumothorax.

The typical CT scan shows diffuse round, bilateral, thin walled cysts of varying size ranging from 2 to 20 mm in diameter. The single most important diagnostic test is a High Resolution Chest CT scan view and confirmed by biopsy. LAM has two types of cells, myofibroblast-like shaped cells and epithelioid-like cells.[6] Immunohistochemistry for HMB-45 and smooth muscle actin and desmin is an important tool in the diagnosis and increases the specificity and sensitivity.[5]

Sirolimus is used for patients with abnormal pulmonary function test. This medication stabilizes lung function, reduces symptoms and improves the quality of life.[7] After the first pneumothorax, pleurodesis is recommended, not waiting for a second event. Lung transplant is one of the last options for patients with advanced disease.

References

1. Meenakshi CJ. Lymphangiomyomatosis presenting as recurrent pneumothorax in a female child: A case report. *Journal of Evolution of Medical and Dental Sciences.* 2015; 4(51): 8930 – 8933.
2. johnson SR. The ERS guidelines for LAM: Trying a rational approach to a rare disease. *Respir Med.* 2010; 104(Suppl 1): S33 – S41.
3. Prieto VC, Zambrano CD, Del CPE, Torrado C, Macías JP. Linfangioleiomiomatosispulmonar: presentación de un caso de evolución fulminante y revisión de la literatura. *Rev Esp Anestesiol Reanim*. 2004; 51: 595 – 599.
4. Harari S, Torre O, Moss J. Lymphangioleiomyomatosis: What do we know and what are we looking for? *European Respiratory Review*. 2011; 20(119): 34 – 44.
5. Spiliopoulos K, Tsantsaridou A, Papamichali R, Kimpouri K, Salemis NS, Koukoulis GK, Tsilimingas NB. Recurrent spontaneous pneumothorax in a 42 years old woman with pulmonary lymphangioleiomyomatosis: Insights and pitfalls of the surgical treatment. *J Clin Med Res*. 2013; 5(1): 70 – 74.
6. Mahishale V, Patil B, Ramannavar A, Suresh G, Rathi A, Sindhuri A. Lymphangioleiomyomatosis: A rare case report. *J NTR Univ Health Sci*. 2014; 3: 189 – 191.
7. Freitas CSG, Baldi BG, Araújo MS, Heiden GI, Kairalla RA, Carvalho CRR. Use of sirolimus in the treatment of lymphangioleiomyomatosis: favorable responses in patients with different extrapulmonary manifestations. *J Bras Pneumol*. 2015; 41(3): 275 – 280.