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A RARE CASE OF CYSTIC LUNG DISEASE

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ABSTRACT
Lymphangioleiomyomatosis (LAM) is a multisystem disorder, predominantlyaffecting post and premenopausal women, which is characterized by cysticlung lesions, abdominal angiomyolipomas (AML) and lymphaticabnormalities, for example, lymphatic tumors, chylous effusions.[1–5] These pathologic features are caused by the proliferation of a neoplasticsmooth muscle–like LAM cell that also has characteristicsof melanocytes.[6] Inherited and sporadic forms of LAM have beendescribed. Sporadic LAM is caused by somatic mutations in anunknown susceptible cell of the tuberous sclerosis complex 2(*TSC2*) gene.[7,8] Sporadic LAM is an uncommon disease occurringin approximately 4.9/1,000,000 women.[9] LAM also occurs in TSC, an autosomal dominantdisorder resulting from germline mutations in the *TSC1* or *TSC2*genes that is characterized by widespread hamartomas in severalorgans including the brain, heart, skin, kidney, eyes, lung, and liver,and occurs in 1 of 6000 live births.[10]

KEYWORDS: Lymphatic tumors, chylous effusions.

CASE REPORT

A 44 years old female, married, and housewifepresented to us with chief complaints of shortness of breath, right sided chest pain, cough and fever for 5 days. Shortness of breath was acute in onset, mMRC grade 4, aggravated on exertion and was relieved partially on lying down inrightlateral decubitus position. Patient had chest pain for 5 days, which was acute in onset and dull aching in nature. It was localized to righthemi-thorax and was non-radiating and non-migratory. It aggravated on exertion and was relieved partially with rest. She complained of cough for last 5 days. It was acute in onset, dry in nature and was not associated with diurnal or postural variation. She also had fever for 5 days. It was present throughout the day and was not recorded. It was not associated with chills and rigor or rash and was relieved on taking medications. No complain of haemoptysis, loss of weight, loss of appetite, orthopnea, abdominal pain, nausea, vomiting, trauma, altered sensorium or loss of consciousness.

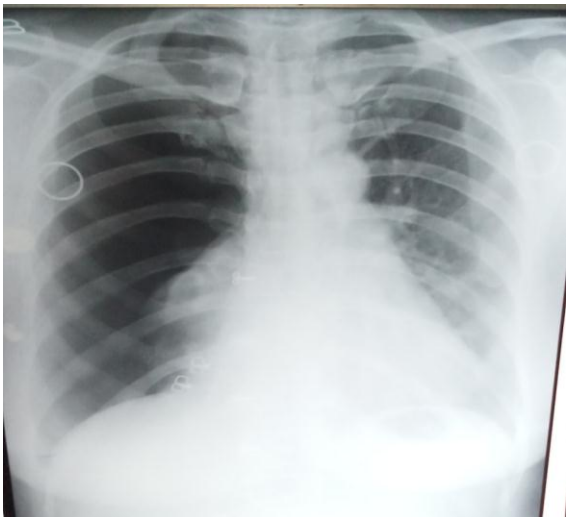
She had history of pulmonary tuberculosis 10 years back. Took ATT for the same for 9 months from a private practioner. She was having uterine mass for 3 months and was on progesteron preparations for the same from private practioner. No history of DM / HTN / CAD / thyroid disorders / asthma or inhaler use in the past.

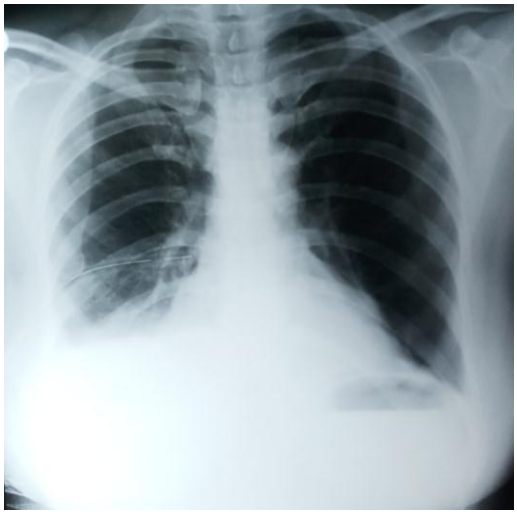
Menstrual history - polymennorhic for last 5 months with 15 days cycle with normal bleeding period and amount.

On general examinationthere was no pallor, icterus, clubbing, cyanosis, pedal edema or lymphadenopathy. JVP was not raised. Then respiratory system was examined. On inspection, therewas assymetrical chest movement with decreased movements onrightside. On palpation, chest movement decreased onrightside and trachea central in position, vocal fremitus decreased onrightside in all regions. On percussion, hyper-resonant notes were present in all regions of righthemithorax and upon auscultation air entry was absent on rightside.

INVESTIGATIONS

X-ray chest- rightpneumothorax





ABG- Type 1 respiratory failure

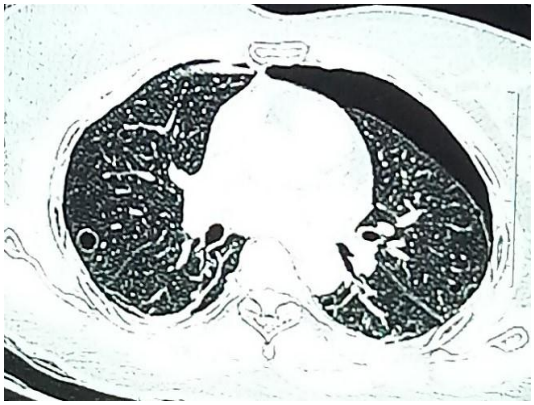
- CBC, LFT, RFT, RBS – WNL
- ECG- WNL
- Cardiac profile- WNL
- D-dimer - WNL

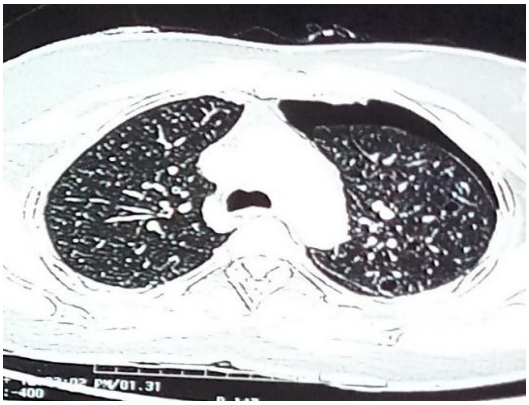
Right pneumothorax resolved with ICTD, but on day-4 patient developed left side pneumothorax

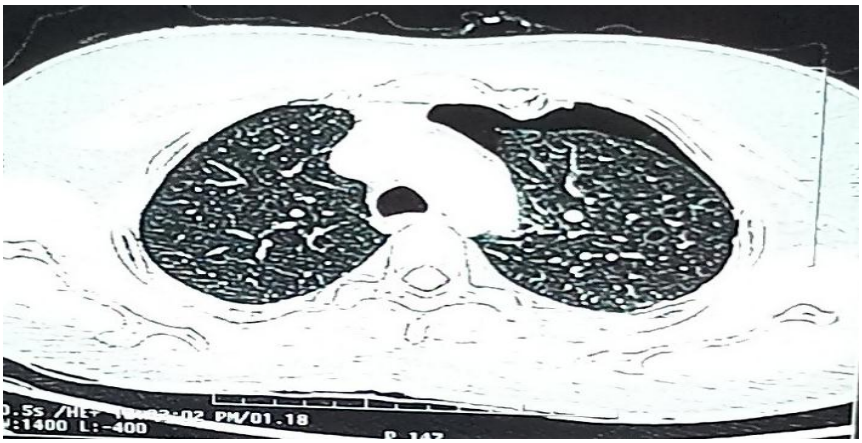
X-ray chest- left pneumothorax with ICTD insitu right side.

Pleural fluid analysis- Straw coloured , TLC-1019, DLC-$N_{78}L_{18}E_4$, total protein-1.67gADA- 22, AFB – not seen, G/S and C/S – sterile.

CECT thorax- evidence of multiple thin walled air filled cystic lesions diffusely distributed in bilateral lung parenchyma. Left hydro-pneumothorax with intercostal drain in-situ.

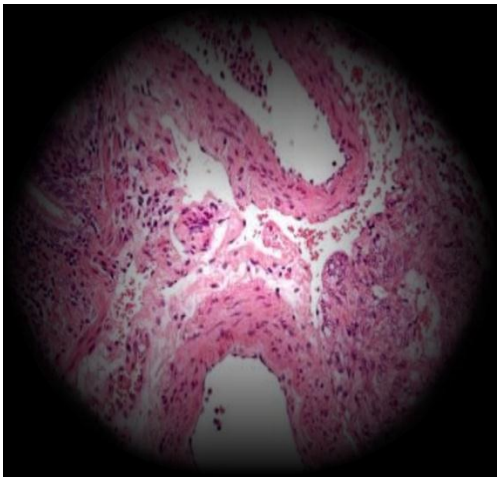






CECT abdomen- bulky uterus measuring 10.5 * 5.9 cm with ill-defined iso-attenuating mass lesion suggestive of intra-mural fibroid in posterior myometrium (approximate size 2.5 * 2.9 cm).

- Trans-thoracic lung biopsy - Few alveoli were seen. Smooth muscle cells surrounding the wall of cystic structures were seen and they are infiltrating into pulmonary parenchyma, airways and blood vessels, suggestive of lymphangioleiomyomatosis.



DIAGNOSIS

This is a definite case of LAM. Definite LAM may be diagnosedin the presence of a characteristic HRCT and a lung biopsyshowing the pathologic features of LAM.

TREATMENT AND FOLLOW-UP

- ICTD was done to correct pneumo-thorax and B/L pleurodesis was done to prevent recurrent pneumo-thorax
- Regular exercise was encouraged. Avoid sports with physical contact like marshall arts and karate
- Gynaecology consultation was done
- Patient is doing well & is on our regular follow up.

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