Ordinary Yet Bizarre: A Rare Pulmonary Lymphangioleiomyomatosis (LAM) Case In Pregnant Lady With Common Presentation

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INTRODUCTIONS

Lymphangioleiomyomatosis (LAM) is a rare and progressive cystic lung disease that occurs usually in women in child-bearing age. It is most commonly diagnosed during pregnancy as it is related with increasing level of estrogen in the body. Spontaneous pneumothorax which can easily be diagnosed with good clinical examinations is one of the most usual preliminary symptoms of LAM. This is an ordinary case of near miss pneumothorax in a pregnant lady who presented with acute dyspnea due to right pneumothorax secondary to a bizarre condition which was LAM yet was diagnosed as pulmonary embolism at first encounter due to incomplete physical examination.

CASE REPORT

31 years old lady with no known medical illness, in her fourth pregnancy at 38 weeks period of gestation presented with short history of shortness of breath and right sided chest pain. She immediately went to the nearest Klinik Kesihatan and initial impression of pulmonary embolism was made. The plan was to immediately send the patient to tertiary center which was 2 hours away. The plan was cancelled as the patient was tachypneic and the team at tertiary center insisted for them to stop at a district hospital for an evaluation. At district hospital via thorough physical examinations revealed that this patient was actually having right sided pneumothorax and chest tube immediately inserted to relieve the symptoms. She was then transferred to tertiary hospital for further treatment and managed to deliver her baby without complications during the stay. The chest tube was removed after few days, however reinserted after few hours due to recurrent pneumothorax. She was put on low-pressure suction and antibiotics to cover for possible lung infection. Despite all treatment, the pneumothorax still persists. High Resolution CT (HRCT) was performed and suggestive for LAM. She was then transferred to other center with cardio-thoracic surgery service availability for thoracospopy.

DISCUSSION & CONCLUSION

LAM is a rare idiopathic and progressive cystic lung disease affecting women in reproductive age group. The prevalence estimated to be around 2.6 patients in 1,000,000 females worldwide. Previously it has mortality up to 100% after 10 years most of it due to respiratory insufficiency, but now overall survival has increase to 71% after 10 years due to early diagnosis and aggressive treatment. There are 2 types of LAM, sporadic and congenital. The congenital is an autosomal dominant genetic disorder as a result of mutation in tuberous sclerosis complex (TSC) gene. LAM characterized by proliferation of unique smooth-muscle like cell in the lung tissue causing air-trapping in the alveoli leading to cyst formation. The gold standard for diagnosing it is via lung biopsy for histopathology and immunohistochemistry. HRCT also can be used to aids in diagnosing LAM where multiple thin-walled round well-defined air-filled cyst can be found. Pneumothorax (50%) is one of the most common presentation after dyspnea (57%), followed by cough (39%), chest pain and pleural effusion. Clinical manifestations of LAM such as pneumothorax usually accelerated during pregnancy as it postulated related to the increase level of estrogen. As for this case, she presented with acute dyspnea and the most common diagnosis to be excluded was pulmonary embolism, we should not underrate the importance of clinical examinations where other diagnosis can be rule-in or rule-out. In-fact after proper examinations, she was having right sided pneumothorax which turn out to be LAM later. As a conclusion, even though LAM is rare and required high end laboratory and radiological investigations for the diagnosis, the early presentation of it such as pneumothorax can be detected clinically after detailed history taking and physical examinations and all medical personal should not underestimate the importance of it

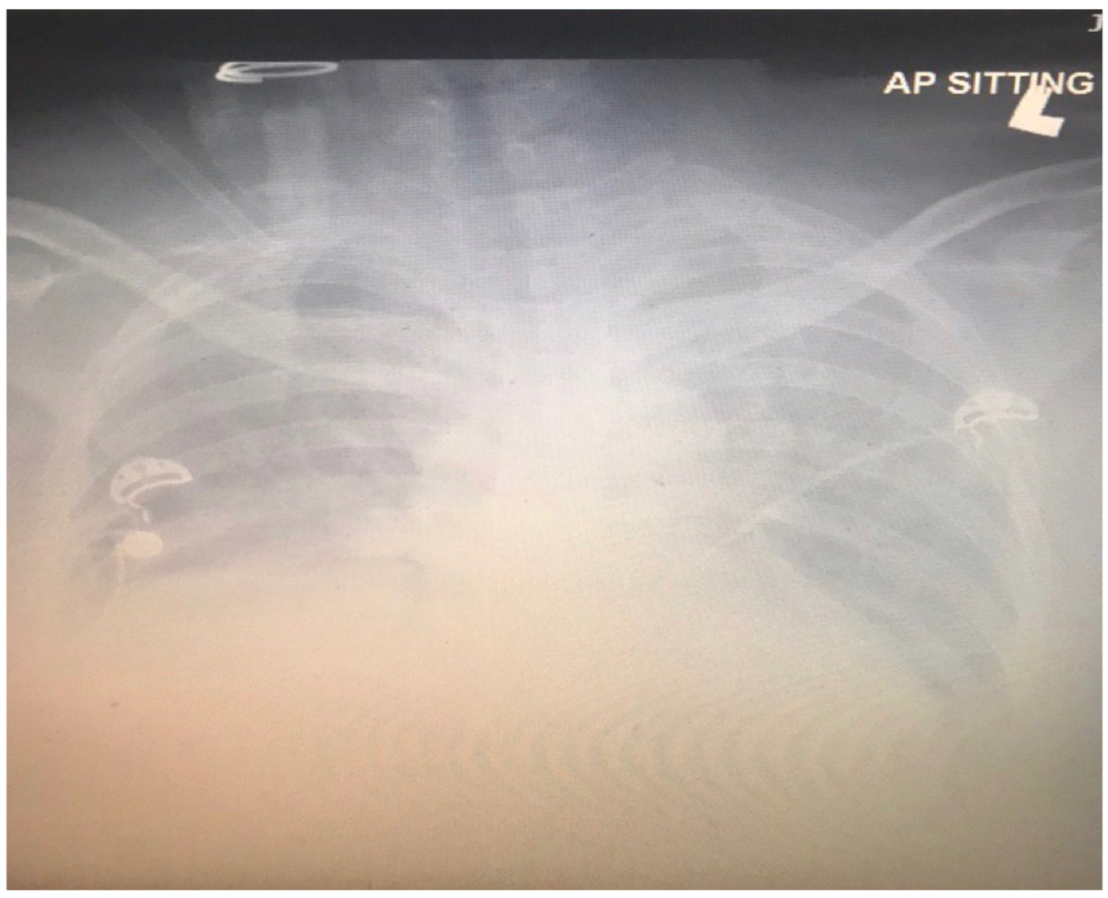

FIGURE 1: First xhest x-ray upon arrival to our emergency department showing right sided pneumothorax with chest tube in-situ

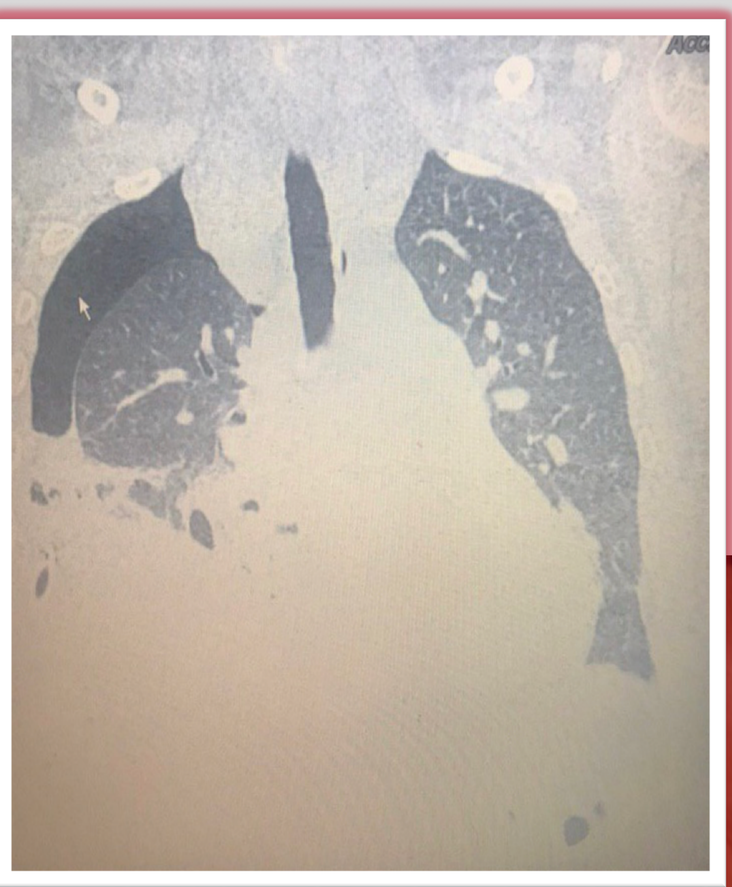

FIGURE 2: HRCT Thorax of the patient after 14 days admission showing persistent pneumothorax

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