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CASE REPORT

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A male patient 19 years old, student, with no special habits of medical importance was referred to Ain Shams University Specialized Hospital complaining of rapidly progressive exertional dyspnea started one week before admission, without orthopnea or paroxysmal nocturnal dyspnea, together with left mammary stabbing pain not radiated or referred. No other associated symptoms & the patient denied any history of trauma.

Past History: Apart from history of congenital Pectus excavatum since birth, there was nothing of relevance.

On examination:

- There was a palpable left supraclavicular lymph node, about 1cm in diameter, firm in consistency, not tender with normal overlying skin.
- ***Locally:*** There was Pectus excavatum deformity with bulging over left hemithorax anterior & posterior (the patient reported that this bulge since birth). There was stony dullness over left inframammary, lower axillary and infrascapular areas, with diminished intensity of breath sounds over same areas & shift of trachea to the other side.

CXR & CT chest were done revealing left sided moderate effusion (Fig. 1). The patient started to develop subcutaneous swelling over left mammary area with crepitus sensation.

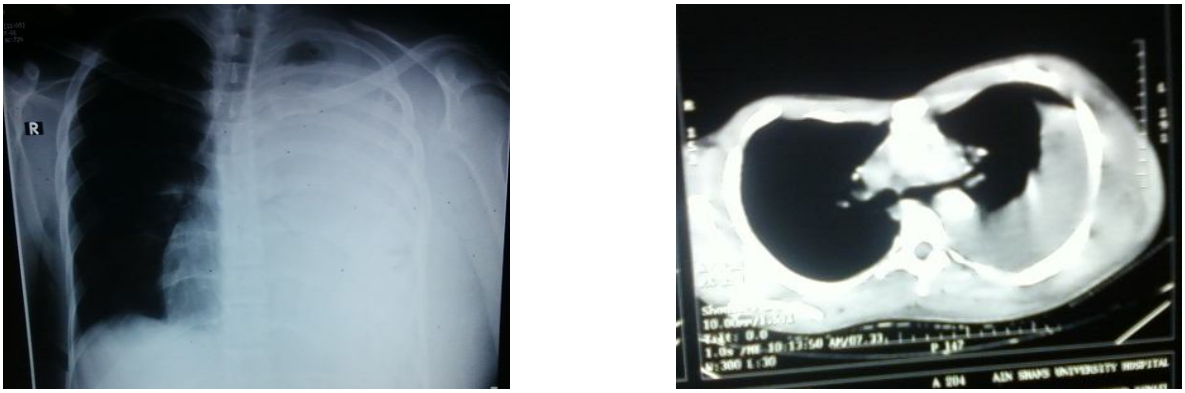

Fig 1. Chest x- ray and CT showed left side pleural effusion with subcutaneous fluid collection.

Hospital course: Tapping of 500 cc of deeply hemorrhagic pleural effusion (not clotted) was done & examination of the fluid revealed exudative fluid (protein: 4.9 gm / dl) rich in lymphocytes & red blood cells with many mesothelial cells.

Chest x-ray hard film for bone examination was done revealing fracture of sixth rib. So, for the possibility of traumatic haemothorax occurrence intercostal tube was inserted. An average of 4–5 liters of deep hemorrhagic effusion was drained daily for two weeks without change in hemoglobin level over this period. All laboratory investigations were within normal values including ESR 4 mm / hr. Repeated cytological examination of the pleural fluid was not conclusive. After a multidisciplinary meeting between the chest medicine, thoracic surgery and thoracic oncology, a decision for thoracoscopic exploration was discarded due to expected adhesions.

However over the third week, hemoglobin level started to decrease down to 10 gm / dl with increase in the size of subcutaneous swelling.

Hence the decision for thoracotomy was taken.

Surgical course:

Exploratory small posterolateral thoracotomy was done. The subcutaneous tissues oozed about 4 liters of serosanguineous fluid. The pleural cavity contained dense adhesions and an amount of 2 liters was drained after the break down of the loculi. The lateral part of the parietal pleura was thickened whereas the mediastinal pleura had some plaques. By gross examination, the lung was completely normal. The lateral pleura was decorticated, hemostasis achieved, 2 drains were left and the patient was transferred to the ICU in a stable condition. Few hours later, the chest tubes surprisingly kept draining blood, hematocrit value was continuously declining, the bleeding profile was increasingly high and thus the patient was transferred to the theater for exploration. During exploration the patient had cardiac arrest and was successfully resuscitated. Massive blood clots were evacuated, but there was no bleeding point. After thoracotomy the bleeding profile was investigated in details and the patient at that time suffered from disseminated intravascular coagulopathy (DIC). In spite of replacing different clotting factors, the same scenario happened few hours later and the patient was re-explored for the second time but the thoracic surgeon packed the patient, closed the skin. The patient after 24 hours was stable and his laboratory profile was satisfactory.

Histopathology: Biopsies from visceral, mediastinal & parietal pleura together with biopsies from lung, pericardium & rib 4 were taken & examined pathologically and by immune phenotyping (Fig. 2). They revealed markedly vascular tumor tissue formed of intercommunicating vascular channels with hyperchromatic lining showing mild pleomorphism.

Tumor tissue markedly infiltrating pleura and infiltrating bony tissue received. There was excess fibrin with extensive hemorrhage.

Diagnosis: Low grade Angiosarcoma, infiltrating pleura and rib bone with congested lung tissue.

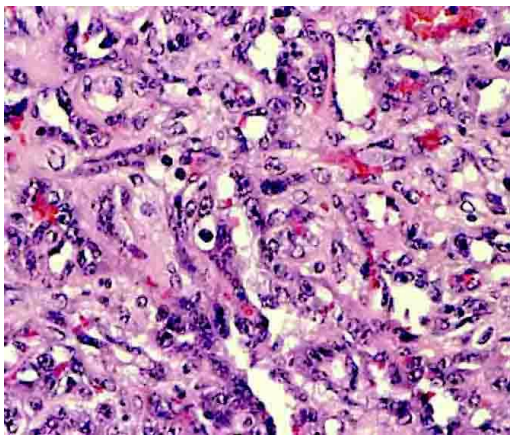

Fig 2. Pleural angiosarcoma.

In the surgical ward, the postoperative course was uneventful. The drains kept weeping and after 21 days the drains were removed consecutively. The patient was referred to the oncology department and he has been receiving chemotherapy in the form of Holoxan / Adriamycin (3 cycles) & Taxol weekly for (3 cycles) with fair general condition.

Comment:

Pleural angiosarcomas are extremely rare account for less than 1% of all soft tissue sarcomas. They arise from endothelial cells and usually originate from small blood vessels. They are extremely rare in the pleura and other serous membranes like the pericardium and peritoneum. Its biological behavior depends on the histological grade, site of origin and multifocality, but generally has a rapidly progressive course.