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Unmasking Ewing's Sarcoma Behind a Hemothorax

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Abstract

Hemothorax is a rare de novo presentation of Ewing's sarcoma, which is a common malignancy of bones in children and young adults with a male predilection. Ewing's sarcoma can rarely affect extra-skeletal sites including the lung and when this sarcoma arises from extraosseous sites, they are found to have neuroectodermal tissue [3]. The incidence of hemothorax presenting as the first manifestation of Ewing sarcoma is unknown, as there are so far, to our knowledge, only a few case reports that have described this phenomenon, of which the majority of cases were reported in children <15 years old [1-5]. Here we present the case of a 20 year old female patient who presented with a massive hemothorax leading to hemorrhagic shock as her initial presentation of Ewing's sarcoma.

Case Presentation

A 20 year old female with no prior medical history who presented with acute on chronic left sided chest pain. She reported a 4 month history of worsening left shoulder pain and pleursiy with no reports of recent trauma or falls. She reported no fevers, chills, hemoptysis or recent sick contacts. She reported no recent travel. Upon arrival to the ED, patient was afebrile, tachypneic to the 30s, tachycardic to the 150s, and hypotensive with MAPS in the 50s. CXR had a possible LUL mass with left sided hemithorax opacification and tracheal and mediastinal deviation to the right, suggestive of a large pleural effusion (Figure 1) She received her 30 mg/kg of LR for sepsis treatment and when her shock persisted, a Levophed infusion was started, at a low dose of 0.03 ng/kg/min. and admitted to the medical icu. Labs revealed a leukocytosis to 17K, a hg of 6.0 (unclear baseline), normal platelets of 255. BMP was unremarkable without evidence of uremia or renal failure. She was immediately started on broad spectrum abx with IV vancomycin, cefepime, flagyl and a unit of packed red blood cells was ordered. Blood cultures and sputum cultures returned negative. Given worsening shock and downtrending hemoglobin levels, there was a concern was for a hemothorax with ongoing bleeding leading to hemorrhagic shock. A bedside 20 French surgical chest tube was placed after bedside ultrasound of the left hemithorax was completed. There appeared to be a large mass in the upper portion of the left chest with basilar free flowing fluid. The chest tube was aimed basilarly and immediately after placement, the chest tube dumped 1.5L of frank blood that clotted in the tubing. The pleural fluid hematocrit was 70%, consistent with a hemothorax.

The cardiothoracic surgery service was consulted. Patient underwent a CTA chest after the hemothorax was partially evacuated with the tube thoracostomy which confirmed a large LUL bilobed pleural based mass measuring 7.6 x 8.7.7cm (image 2) with persistent hemothorax. She unfortunately continued to have high bloody chest tube output with worsening hemodynamic instability, rising pressor requirements, and downtrending hg and was taken to OR for VATs procedure. In the OR she was given 3 units PRBC, 1.8 crystalloid, with 500cc EBL. Intraop note stated there was a large amount of clot and bleeding from mass with multiple hemostatic methods needing to be employed. There was mention in the operative note that the mass appeared to have fractured possibly from pressure. Once the hemothorax was evacuated, multiple biopsies of the mass were taken. Surgical chest tubes were upsized and patient was sent to the surgical ICU to convalesce post-surgery. Biopsies of the pleural based mass revealed small round blue cell tumor with features suggestive of Ewing sarcoma. She recovered well post op and soon after was staged with a PET scan which was concerning for Stage 4 disease due to interval development of left lung nodules, right lung nodules, and right chest wall masses. She was evaluated by cardiothoracic surgery and not deemed to be a surgical candidate due to metastatic disease in the right chest, and started on alternating courses of vincristine, Doxorubicin cyclophasphamide and then ifosfamide and etoposide. She also received radiation to the LUL mass. She had progression of disease s/p 8 cycles VDC/IE and single agent ifosfamide 2 cycles. Pazopanib was discussed as next line therapy, but unfortunately she succumbed to neutropenic septic shock after one of her chemotherapy and passed away.

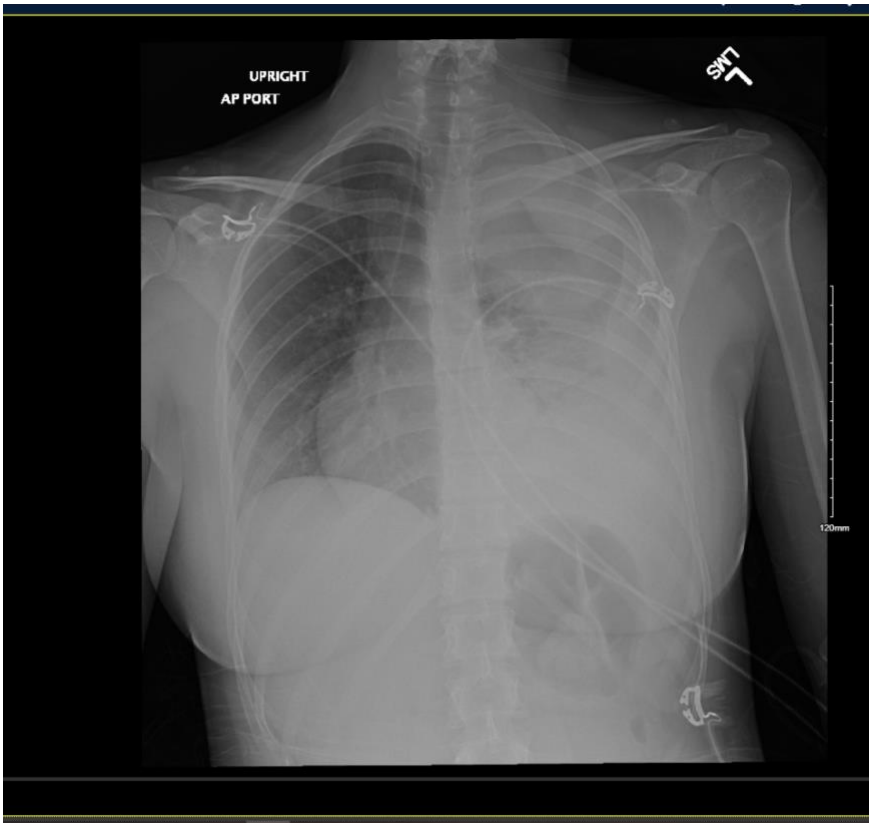

Figure 1: CXR on admission with Left upper lobe mass and left sided hemithorax opacification with mediastinal shift to contralateral side, suggestive of large left sided effusion.

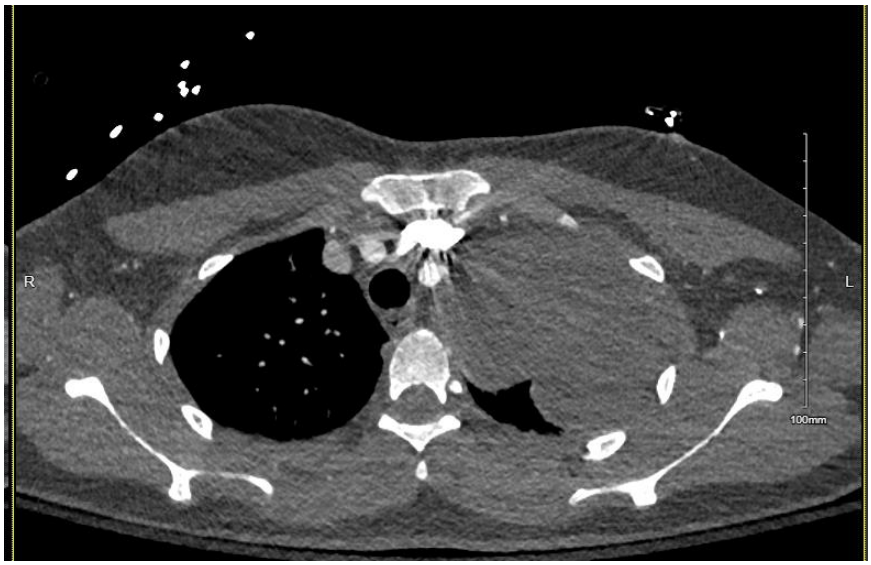

Figure 2: CT chest with large bilobed 7 cm mass with heterogeneous density (blood) in the mass. This was later biopsied and found to be consistent with Ewing's sarcoma.

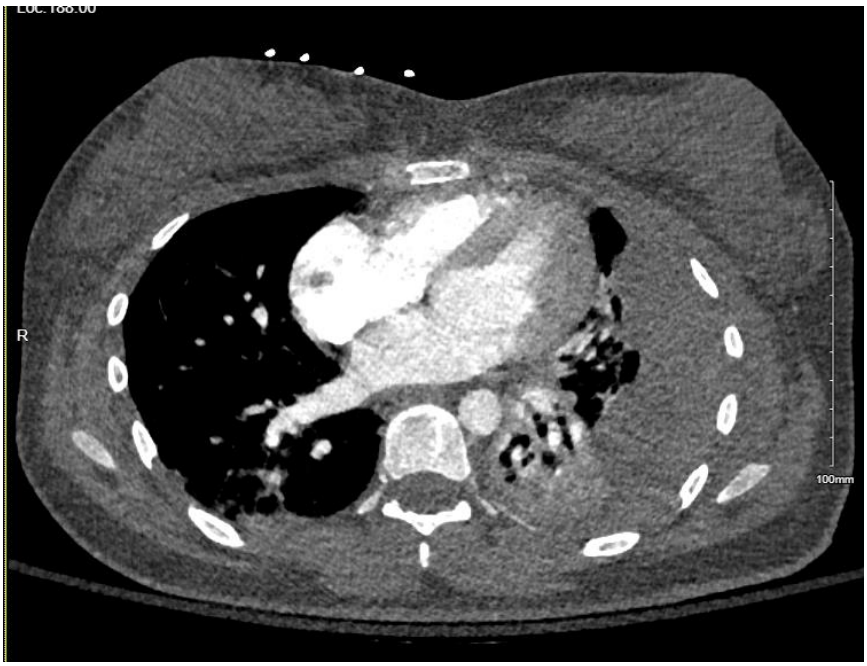

Figure 3: CT chest image of left sided pleural effusion with high density material, consistent with hemothorax.

Discussion and Conclusion

A hemothorax is the accumulation of blood in the pleural cavity, diagnosed by a pleural hematocrit >50% of the serum hematocrit. The etiologies for hemothorax are wide and include: trauma; blunt or penetrating (most common etiology), bleeding tumors, hematologic disorders, procedural complications, vascular abnormalities, infectious disease, endometriosis or idiopathic. Treatment includes drainage of the blood from the pleural space with tube thoracostomy and if bleeding continues a more invasive procedure for hemostatic measures are required, like surgical intervention with a VATS. It is postulated that Ewing sarcoma can be associated with development of a hemothorax through several mechanisms [3]: tumor invasion into blood vessels causing bleeding into the pleural space, tumor rupture and bleeding into the pleural space, and trauma associated fractures due to weakened bone structure. Finally, certain oncologic treatments (particularly VEGF inhibitors) have been associated with increased risk of bleeding and hemothoraces. Based on the Operating Room note during evacuation of her hemothorax, it was suggested that the etiology of our patient's spontaneous nontraumatic hemothorax was due to a ruptured and fractured mass.

Therapy for localized disease is surgical excision. Standard first line therapy for metastatic Ewing Sarcoma is alternating courses of 5 drugs: vincristine, doxorubicin, cyclophosphamide, ifosfamide, and etoposide; the regiment our patient received. Pazopanib efficacy in pulmonary Ewing's sarcoma has been reported in case studies but is not standard of care and more clinical trials need to be done [4]. It is an oral tyrosine kinase inhibitor with activity against vascular endothelial growth factor receptor. Perhaps in the future it will be added to the treatment regimen earlier in the detection of Ewing's sarcoma.

Timely identification and diagnosis of patients is crucial, as the 5-year overall survival rate for localized Ewing sarcoma is 70%, compared to less than 30% for metastatic disease. Therefore, early diagnosis is essential. As Ewing's sarcoma is a malignancy typically found in children, adult physicians must remain vigilant to keep this esoteric adult malignancy in the differential of a nontraumatic hemothorax, given early detection is key as the prognosis varies based on stage at diagnosis [2].

REFERENCES

1. Wolf G, Aigner RM, Schwarz T. Massive pleural effusion with dyspnea in a 17-year-old boy as the first sign of ewing sarcoma. J Pediatr Hematol Oncol. 2002; 24: 420.
2. Al-Sukhni L, Maniam G, Dean R, et al. Pediatric hemothorax: a rare presentation of Ewing sarcoma. Consultant. 2023; 63: 12-14.
3. Ling X, Tong J, Wang L, et al. Primary pulmonary Ewing's sarcoma: rare cause of massive hemothorax in a young girl-case report. BMC Pediatr. 2021; 21: 194.
4. Sanna S, Bertolaccini L, Brandolini J, et al. Uniportal video-assisted thoracoscopic surgery in hemothorax. J Vis Surg. 2017; 3: 126.
5. Eroğlu A, Kürkçüoğlu İC, Karaoğlanoğlu N, et al. Extraskeletal ewing sarcoma of the diaphragm presenting with hemothorax. The Annals of Thoracic Surgery. 2004; 78: 715-717.