Pediatric Hemothorax: A Rare Presentation of Ewing Sarcoma

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A 13-year-old boy was admitted to the hospital with acute-onset dyspnea and chest pain, which had started 4 hours prior to presentation. The pain was described as persistent and pleuritic, radiating up the chest wall, without alleviating factors. A week prior, he had noted intermittent right-sided rib pain of waxing and waning quality, but these episodes were short lived.

History

His family had just returned from a camping trip at a nearby lake where the boy had been swimming, but he denied any overt trauma to the chest from his activities. His family history was notable for small cell carcinoma in his father, diagnosed at age 51 years after a prolonged smoking history.

Physical examination

Upon presentation, the patient had dyspnea and was unable to speak in full sentences. He had an episode of altered mentation and facial pallor, lasting a few seconds, en route to the emergency department.

His vital signs were within normal limits, except his heart rate was elevated at 106 beats/min. He was in moderate respiratory distress with subcostal, intercostal, and supraclavicular retractions. His breath sounds were markedly diminished throughout the right lung fields with dullness to percussion from right subcostal margin to the second rib space.

Diagnostic testing

An initial chest radiography scan was conducted, results of which revealed a large, right loculated pleural effusion. It was further evaluated with a computed tomography (CT) angiography of the chest with contrast (**Figures 1-3**). Results revealed a large, multiloculated, right-sided pleural effusion with compression atelectasis of the right lung without underlying pneumonia.

Upon chest tube placement the following day, frank blood of about 600 mL in volume was observed. With this finding of massive hemothorax, concern was turned to possible etiologies such as arteriovenous malformation or even malignancy. Results of an initial complete blood cell count revealed mild anemia, with a hemoglobin level of 11.5 g/dL (reference range, 10.5-13.5 g/dL) and a hematocrit level of 32 g/dL (reference range, 33-39 g/dL). Results of a comprehensive metabolic panel showed mild hyperglycemia with a blood glucose level of 182 mg/dL (reference range, 74-106 mg/dL). Results of a coagulation panel and peripheral blood smear, as well as the patient's uric acid level, lactate dehydrogenase level, erythrocyte sedimentation rate, reticulocyte count, were all within normal limits. However, his C-reactive protein level was significantly elevated at 14.7 mg/L (reference range, 0-2.99 mg/L).

A magnetic resonance imaging (MRI) scan of the chest showed generalized pleural thickening in the right middle and lower lung, involving both visceral and parietal pleura. After sufficient drainage of the hemothorax by hospital day 6, video-assisted thoracic surgery (VATS) was performed, and a small mass was removed, emerging from the 7th right hemivertebrae. Upon pathological and genetic evaluation of this tissue, an unexpected diagnosis was realized. Microscopic evaluation and immunohistochemical staining of the specimen were consistent with malignant cells. Genetic evaluation of the specimen revealed *EWSR1* gene arrangement, confirming the diagnosis of

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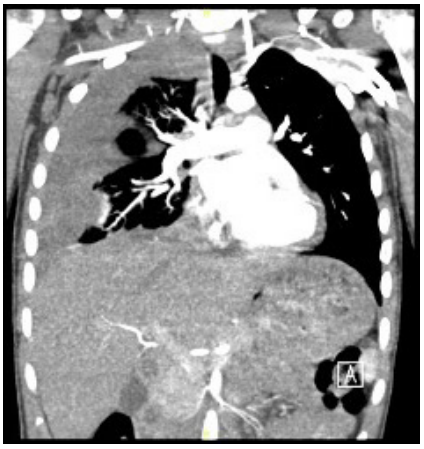

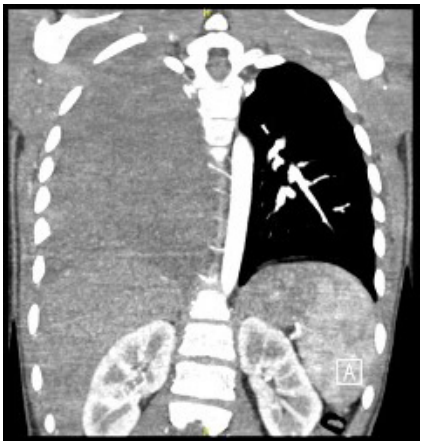

Figure 1. A CT scan of the chest, coronal views, showed a large, multiloculated, right-sided pleural effusion with compression atelectasis of the right lung without underlying pneumonia.

Ewing sarcoma. A whole-body positron emission tomography/CT scan confirmed that it was malignant disease in the right middle and lower chest wall, as well as in the paratracheal region, but without extrathoracic involvement. A bone marrow biopsy was conducted, results of which were normal.

Patient outcome

With this diagnosis, the patient was initiated on chemotherapy according to the Children's Oncology Group protocol starting with vincristine, doxorubicin, cyclophosphamide with mesna. To date, he has completed cycle 6 of chemotherapy with good response. A repeat MRI scan showed significant improvement in pleural thickening that had been previously noted, and the patient has begun radiation therapy.

Discussion

Pediatric hemothorax is a rare presentation. In most settings, a history of chest wall trauma exists.[1] However, nontraumatic etiologies are an important category to consider in the workup. The incidence of hemothorax in pediatric thoracic trauma is estimated to be about 7% to 29%.[2] Other causes of pediatric hemothorax include iatrogenic causes, tuberculosis, hemothorax secondary to pneumothorax, coagulopathy, pulmonary vascular malformation, and malignancies.[3] Herein, we present a patient with pediatric hemothorax secondary to Ewing sarcoma, and a discussion of this particularly rare presentation may assist in the recognition and treatment of future cases.

Pediatric hemothorax, when secondary to thoracic trauma, is associated with a high mortality rate and is, therefore, a medical emergency.[3] The relatively increased compliance of the pediatric thoracic cage can cause a hemothorax, with only minimal external signs of injury. Therefore, it is imperative to ask about recent history of chest trauma in all patients presenting with pleural effusions on chest radiograph or thoracentesis-confirmed hemothorax.[2] Recent hospitalizations are pertinent to the history as well, given that iatrogenic hemothorax is likewise possible because of recent intrathoracic surgery and placement of chest drains or central lines.[3] Furthermore, the differential diagnosis, which all are evaluated based on clues in the history and physical examination, includes:

Tuberculosis, which is rare in the United States but can present with hemothorax

- Pneumothorax
- Coagulopathy
- Pulmonary vascular malformation
- Collagen vascular disorders with spontaneous rupture
- Malignancies

Regardless of the etiology, these pediatric patients must be managed carefully but efficiently because of a myriad of possible complications, including empyema, fibrosis, hemodynamic collapse, and cardiac arrest.[2]

After immediate stabilization of the airway, breathing, and circulation, as well as identification of an effusion on imaging, hemothorax is diagnosed with needle aspiration and an output of frank blood or bloody fluid with a hematocrit value more than 50% of that of the circulating hematocrit.[2] As hemothorax usually cannot be differentiated from other causes of pleural effusion by imaging alone, needle aspiration is performed before laboratory studies, as hemodynamic stabilization is the priority. If trauma is indeed the underlying cause, the pleural effusion will often be accompanied by pneumothorax, pulmonary contusions, rib fractures, spinal fractures, or injury to the mediastinum, and these are important clues for the final diagnosis.[3] In our patient's case, the pleural effusion was not associated with any of these imaging features, which made traumatic hemothorax less likely as well as did an absence of trauma in the

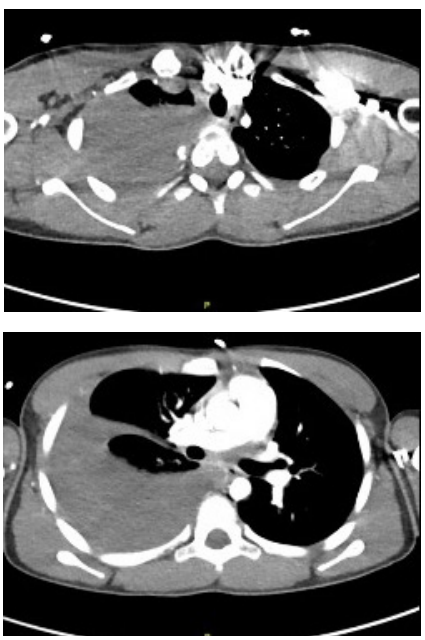

Figure 2. The transverse view of a CT angiography scan with contrast showed a large, multiloculated, right-sided pleural effusion with compression atelectasis of the right lung without underlying pneumonia.

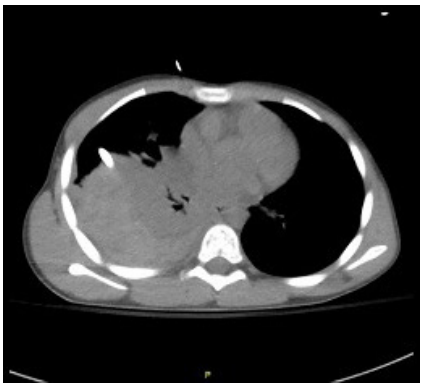

Figure 3. The transverse view of the CT scan with contrast of the thorax confirmed a large, multiloculated, right-sided pleural effusion with compression atelectasis of the right lung without underlying pneumonia

history. However, blunt chest trauma in children can lead to shearing of thoracic vessels without rib damage because of the high compliance of the chest wall.[2] An absence of travel history or exposure to crowded living situations or exposure to incarcerated individuals made tuberculosis infection less likely. Initial laboratory studies should include a complete blood cell count and coagulation profile to determine whether thrombocytopenia or consumptive coagulopathy is the inciting factor. This is also to establish either a baseline hemoglobin/hematocrit level or to determine degree of blood loss.

Malignancies, either intrathoracic or metastatic, are an important category of causes for which health care providers should have a high index of suspicion. Ewing sarcoma is a bone malignancy in the primitive neuroectodermal tumor family of tumors, which most commonly affects the pediatric and young adult populations.[4] The disease most commonly affects the major long bones but is also notorious for affecting the pelvis and ribs. However, the presentation of Ewing sarcoma as a hemothorax is quite rare. One case report described a 12-year-old girl presenting with a 1-month history of dyspnea.[5] She was found to have pleural effusion on imaging and a hemothorax after diagnostic thoracentesis. A thoracic MRI scan demonstrated a solid extraparenchymal mass in the right costodiaphragm, for which CT-guided biopsy was used to diagnose primary extraskeletal Ewing sarcoma of the diaphragm.[5]

A similar case highlighted a 21-year-old man with a history of smoking who developed a hemothorax as the presenting manifestation of a primitive neuroectodermal tumor.[6] It was successfully treated via VATS tumor removal, chemotherapy, open thoracotomy with pleurectomy for complete tumor removal, and limited radiation.[6] It is worth noting that a traumatic hemothorax does not exclude the possibility of malignancy, as in the case of a 15-year-old girl who presented with a left-hemothorax following a snowboarding accident; her hemothorax recurred 1 month after treatment despite pleural drainage and radiographic evidence of resolution.[8] She also reported weight loss and fatigue and was also found to have anemia. Results of a thoracotomy revealed a massive tumor that was found to be Ewing sarcoma.[9]

Conclusions

These cases demonstrate the insidious nature of Ewing sarcoma of the thoracic cavity and its ability to go undetermined yet cause significant morbidity before discovery. Diagnosis of Ewing sarcoma requires immunohistochemical staining or histopathological examination of the lesions of interest.[8] The most effective treatment protocol is to first drain the hemothorax and stabilize the patient, with follow-up wide-margin surgical resection of the sarcoma.[9] Surgical resection is often via VATS6,[9] or thoracotomy[7] and is followed by chemotherapy with high-dose radiation.[5] The standard chemotherapy regimen includes vincristine, actinomycin, cyclophosphamide, and doxorubicin,[10] and early treatment initiation improves prognosis considerably.[5] Although trauma is the most common cause of a hemothorax, pediatric health care providers should keep insidious etiologies such as Ewing sarcoma on the differential diagnosis, as early diagnosis is one of the most important prognostic factors for outcomes with this disease.

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