Massive Hemothorax Due to Intrathoracic Extramedullary Hematopoiesis in a Patient With Thalassemia Intermedia*

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We report the case of a 49-year-old woman with thalassemia intermedia who developed a massive hemothorax due to hemorrhage from a large intrathoracic, paraspinal hematopoietic mass. Thoracotomy was required for initial control of bleeding. Postoperatively she received a total of 1,500 rads to the mass and has not had recurrence of the hemothorax. This complication of extramedullary hematopoiesis has not been previously reported, to our knowledge. (Chest 1988; 94:659-60)

Massive extramedullary erythropoiesis is an unusual phenomenon, most frequently observed as a response to the intense intramedullary hemolysis associated with the homozygous β-thalassemias and related conditions, such as hemoglobin E-thalassemia. The subset of such patients who do not require transfusion (thalassemia intermedia, or mild thalassemia) is most likely to display this phenomenon, since their erythropoiesis is not suppressed. The erythropoietic masses are usually asymptomatic, although spinal cord compression has been reported. We describe a patient with thalassemia intermedia in whom hemorrhage from a large erythroid mass resulted in massive hemothorax, to our knowledge a heretofore unreported occurrence.

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had cul-de-sac is in for many years. retrosternally and to the left side (open arrow). A second chest tube was placed but did not result in significant improvement. A left thoracotomy was performed. The chest was filled with approximately 3 L of clotted blood. There was an 8 × 6-cm mass in the left paravertebral area just above the diaphragm, and a 3 × 2-cm mass just lateral to it which had not been appreciated on CAT scan. The capsule of the larger mass had several oozing stellate lacerations and a larger, rapidly bleeding longitudinal tear. The smaller lacerations were readily closed with sutures. The larger laceration required packing with Gel-foam soaked in thrombin, and apposition of the edges with pledgetted mattress sutures to control the bleeding. The intraoperative blood loss was estimated to be 8 L. Biopsy of the mass revealed extramedullary hematopoiesis. Postoperatively, the left and later the right mediastinal masses were irradiated with 1,500 rads in divided doses. There has been no recurrence of bleeding.

**CASE REPORT**

The patient is a 46-year-old black woman with thalassemia intermedia. Hematologic and globin synthetic data concerning the patient and her family have been reported. Aside from severe anemia associated with her only pregnancy at age 21, and again with secondary hypersplenism at age 35 (successfully alleviated by splenectomy), her hemoglobin has been stable at about 8.0 g/dL. She has had long-standing hepatomegaly with mild abnormalities of liver chemistry studies. A liver biopsy specimen at age 40 showed hemochromatosis with slight fibrosis. At age 44, iron unloading with continuous, subcutaneous desferrioxamine infusion was initiated and has continued to date. She remained actively employed and clinically well.

She was long known to have a left posterior mediastinal mass on chest roentgenograms, which has gradually increased in size over the past 11 years (Fig 1) but produced no symptoms. A mass in the cul-de-sac had also been noted, and both were assumed to be extramedullary erythropoietic tissue. Extensive osteopenia, consistent with expansion of the hematopoietic marrow, had also been present for many years.

On the morning of admission she awoke with the sudden onset of left pleuritic chest pain radiating to her left shoulder, weakness, and rapidly increasing dyspnea. She later recalled that her husband's arm had struck her chest when he turned over in his sleep the night before. On admission the blood pressure was 90/60 mm Hg, pulse rate, 120 and regular; respirations, 28/min; and temperature 38.7°C rectally. Physical examination results showed signs of a massive left pleural effusion, which a chest roentgenogram showed to extend almost to the lung apex. The WBC count was 8,400/mm, and platelets were 412,000/μm. The bleeding time, prothrombin time, and partial thromboplastin time were normal. Thoracocentesis yielded gross blood with a hematocrit reading of 20 percent which was equal to the venous hematocrit. Pleural fluid analysis was negative for malignant cells and for microorganisms on smears and cultures.

A chest tube was placed and drained 3,400 ml of bloody fluid over the next 48 hours. The patient's condition improved with transfusions of packed RBCs. The chest roentgenogram showed a reduction in the hematopericardium. A CAT scan showed the large left posterior mediastinal mass, surrounded by blood and clot in the left pleural cavity. A smaller mass was seen in the right paravertebral area, with a small right pleural effusion. Visualization of the masses was markedly enhanced following administration of IV contrast medium (Fig 2).

On the fourth hospital day, her condition deteriorated; her left effusion was found to have increased again, almost filling the left postcardiac area. A new anterior mass was noted, and the left lung was flattened. Left thoracotomy was performed. A 5-cm, partially cystic mass was found in the left pleural cavity. The surgical specimen contained 3 L of clotted blood, and a 3 × 5-cm mass is in the left retrosternum. The mass was excised, and a left thoracotomy was performed. A left chest tube was placed but did not result in significant improvement. A left thoracotomy was performed. The chest was filled with approximately 3 L of clotted blood. There was an 8 × 6-cm mass in the left paravertebral area just above the diaphragm, and a 3 × 2-cm mass just lateral to it which had not been appreciated on CAT scan. The capsule of the larger mass had several oozing stellate lacerations and a larger, rapidly bleeding longitudinal tear. The smaller lacerations were readily closed with sutures. The larger laceration required packing with Gel-foam soaked in thrombin, and apposition of the edges with pledgetted mattress sutures to control the bleeding. The intraoperative blood loss was estimated to be 8 L. Biopsy of the mass revealed extramedullary hematopoiesis. Postoperatively, the left and later the right mediastinal masses were irradiated with 1,500 rads in divided doses. There has been no recurrence of bleeding.

**DISCUSSION**

Intrathoracic extramedullary hematopoiesis is a rare condition and is usually asymptomatic. Verani et al reviewed the literature in 1983 and collected 55 cases. With the exception of one patient who had dyspnea relieved with resection of an intrathoracic mass of erythropoietic tissue, the only previously reported complications have been due to spinal cord compression. Luyendijk et al in 1975 collected five cases of spinal cord compression from the literature and added one of their own. To our knowledge, our patient is the first reported case of hemothorax in this condition.

Extramedullary hematopoiesis is seen in a variety of hematologic disorders. These include severe hemolytic anemias such as thalassemia and conditions such as myelofibrosis and myelophthitic anemia, where there is extensive replacement of normal marrow.

The most common sites of extramedullary hematopoiesis are the liver, spleen, and lymph nodes. Other sites are involved less frequently. The foci of heterotopic marrow are usually microscopic but they may be macroscopic and grow to a large size, as in this case. In the chest, tumor-like masses are formed which have a characteristic appearance. They are usually located in the lower paravertebral areas and are usually multiple and bilateral. They may have a lobulated appearance. Destruction of adjacent ribs and vertebrae is not seen. Pathologically, the masses are soft, deep red, and resemble splenic tissue on the cut surface. Histologically, they are formed of hematopoietic elements mixed with adipose tissue.

The origin of the erythropoietic tissue in intrathoracic extramedullary hematopoiesis is unclear. It may arise by extrusion of hyperplastic marrow through the thinned cortex of ribs or vertebrae, with the periosteum forming the capsule of the mass. Alternatively, embryonic rests may transform into hematopoietic tissue under circumstances of bone marrow stress.

The diagnosis of intrathoracic extramedullary hematopoiesis can be established with reasonable certainty on the basis of the characteristic radiologic findings in a patient with a predisposing hematologic condition. Neurogenic tumors in the posterior mediastinum are the main considerations in differential diagnosis. Features that help to distinguish the two conditions are the frequent finding of lobulation and the absence of bone destruction in intrathoracic extramedullary hematopoiesis. Bone erosion and sclerosis occur in about 50 percent of patients with neurogenic
tumors and lobulation is not seen. In addition, enhanced visualization of the masses on postcontrast computerized tomography supports the diagnosis of intrathoracic extramedullary hematopoiesis as demonstrated in our case.

Although invasive diagnostic procedures, including thoracotomy and aspiration needle biopsy, have been used, they are usually not necessary and are potentially hazardous because of the highly vascular nature of the thoracic masses. At thoracotomy Andras et al found the blood supply to derive from intercostal arteries and the esophageal plexus. Venous drainage was into the azygous system. In several reports surgical resection has been complicated by extensive hemorrhage requiring multiple transfusions. Our case provides a dramatic example of the hemorrhagic potential of this condition.

Treatment is required only in the presence of complications. Treatment may in fact be deleterious due to removal of a compensatory source of erythropoiesis and exacerbation of the underlying anemia. Extramedullary hematopoietic tissue is highly radiosensitive, and relatively small doses of radiation have been effective after surgical decompression in cases of spinal cord compression. In our patient radiotherapy was administered to prevent recurrent hemorrhage after surgical control of the acute bleeding.

It is unclear whether there was some factor in our patient which provoked the hemorrhage resulting in massive hemothorax. The lacerations in the capsule of the larger mass seen at thoracotomy may have resulted from spontaneous rupture. The role of the apparently superficial trauma that she sustained prior to admission remains speculative.

Awareness of this potential complication of intrathoracic extramedullary hematopoiesis may make it possible to treat future cases with radiation and chest tube drainage without resorting to thoracotomy.

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Pathogenesis of Cerebral Air Embolism during Neodymium-YAG Laser Photoablation*

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We describe a case of air embolism complicating neodymium-YAG laser photoablation of an endobronchial carcinoid tumor. A 27-year-old man experienced an acute neurologic syndrome during laser photoablation which responded to acute hyperbaric therapy. (Chest 1989; 94:660-62)

The phenomenon of arterial air embolism has been described as a sequela to major thoracic trauma, diving accidents, selective arterial angiography, and central venous catheter malfunctions. Recently, arterial air embolism has been reported as a consequence of transbrachial needle lung aspiration. In addition, the occurrence of cerebral air embolism has been reported with transbrachial lung biopsy in a patient with miliary tuberculosis and, recently, in a patient with pulmonary amyloidosis.

In this report we present a case of acute air embolism complicating YAG laser photoablation of an endobronchial carcinoid.

CASE REPORT

The patient is a 27-year-old, heterosexual, nonsmoking, Caucasian man who was admitted with persistent left lower lobe (LLL) pneumonia. Fiberoptic bronchoscopy was performed and demonstrated a large, exophytic, and hypervascular lesion totally occluding the LLL bronchus. The patient reported a long history of flushing associated with childhood "hyperactivity" and "panic attacks" and had been given various triyclic medications. He described a previous episode of "tachycardia" while taking these medications, which required brief treatment with propranolol.

On physical examination, his systemic blood pressure was 160/70 mm Hg, pulse rate 98/min, respirations 20/min, and rectal temperature was 38.3°C. Examination of his head and neck was unremarkable. Cardiac examination demonstrated a regular rhythm, normal S1 and S2, without murmurs. There was evidence of decreased breath sounds in the area of the LLL. The results of remainder of the examination were normal. The patient's data base included a WBC count of 23,900/cu mm, with a left shift. The electrolytes and hemoglobin values were normal.

Course

Approximately six weeks prior to his transfer to our institution, the patient had developed fever (40.0°C), pleuritic chest pain, and a productive cough of green sputum which failed to resolve on treatment with oral erythromycin on an outpatient basis. At another institution, the patient was noted to have a LLL pneumonia and pleural effusion, while the sputum demonstrated "mixed flora." He was treated with IV ampicillin and metronidazole. His course was

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