**DISCUSSION**

Autoimmune chronic active hepatitis was suspected on the basis of prolonged jaundice, hepatosplenomegaly and exclusion of known viral and metabolic causes of chronic hepatitis. The presence of ANA, SMA and the hepatic histology confirmed this diagnosis. The acute drop in the Hb level from 83 to 50 g/L, hemoptysis, respiratory distress and radiologic findings were consistent with AH.

Life-threatening AH is rare in children. Local causes of hemorrhage such as tuberculosis were excluded by the subsequent clinical course. Systemic lupus erythematosus, which may be associated with AH, was considered. However, she did not fulfill the 1982 revised clinical and immunologic criteria for the diagnosis of SLE. Goodpasture syndrome was excluded since there was no renal involvement. Systemic vasculitides account for 40 to 60 percent of adult cases of immune pulmonary hemorrhage. In systemic vasculitides, glomerulonephritis almost always accompanies the AH, and they are thus excluded in our patient. The AICAH rules out idiopathic pulmonary hemosiderosis which is a diagnosis of exclusion. In this case, AH is thus secondary to AICAH.

The pathogenesis of AH in our patient is speculative, since she was too ill to have a lung biopsy. The coagulopathy contributed to the bleeding but the lack of overt bleeding from other orifices and puncture sites makes it unlikely that this was the sole reason for the AH. Factors such as immune complex vasculitis and infection have been incriminated in the pathogenesis of AH. Since the alveolar infiltrates cleared within two days, it is unlikely that an infection triggered the AH. The cultures were negative and the fever could be a manifestation of the underlying immune process. We conclude that a combination of coagulopathy and vasculitis contributed to the AH in this patient with AICAH.

**REFERENCES**


**Pneumoblastoma in Neurofibromatosis**

Luc P. Bron,† Nigel R. Howarth, M.D.; and Alex F. Muller, M.D.

Pneumoblastoma is a rare tumor composed of two histologic cell types, arising from epithelium and stroma. Patients with von Recklinghausen's disease are known to develop certain types of tumors. A rare, and possibly first case of pneumoblastoma arising in a patient with neurofibromatosis is described.

**CtE SYSTEM**

A 34-year-old man was admitted for investigation of a large left hypochondriacal mass, having complained of a dull and constant abdominal pain for one month. He had lost 4 kg in weight over the same period. A diagnosis of neurofibromatosis had been made three years ago on the basis of multiple "café au lait" type skin lesions, and he had undergone left pneumectomy for excision of pneumoblastoma two years ago. The margins of the specimen obtained at surgery along with sampled mediastinal nodes were clear of tumor, and neither radiotherapy nor chemotherapy had been proposed.

During the last two years, he had led a normal life, returning to full-time employment as a post office worker. He had been followed every three months by his physician.

On admission, he appeared thin with at least ten "café au lait" patches over his upper trunk with no other skin lesions. A tender, smooth, fixed, 13 cm mass could be palpated in the left hypochondrium. Laboratory studies disclosed normal values for the following: full blood cell count; erythrocyte sedimentation rate in the first hour; urea and electrolytes; hepatic function, LDH; and CEA. The C-reactive protein value was increased at 1.13 mg/dl. Abdominal ultrasonography revealed an extensive mass filling both the left hemithorax and progressing through the diaphragm into the abdominal cavity. A percutaneous biopsy specimen revealed recurrent pneumoblastoma.

Arteriography (Fig 1) showed that the mass was supplied by intercostal, inferior left diaphragmatic and branches from the splenic artery. Magnetic resonance imaging (Fig 2) demonstrated that the tumor was invading all of the left hemithorax, the left thoracic wall, and progressing through the diaphragm into the abdomen, displacing the spleen and the kidney medially. The pericardium appeared not to be involved. Surgical resection was deemed impossible, and the patient underwent a course of palliative radiotherapy.

**DISCUSSION**

Pneumoblastoma is a rare tumor. Only 130 cases had been documented in the literature by 1990, three having been operated on in the past 11 years at the Geneva University Hospital.

This tumor can occur from childhood to old age, with an average age at diagnosis of 40 years. There is a male to female predominance of 3 to 1.

Pathologically, pneumoblastoma typically is a large, well-defined mass located in the periphery of the lung. Extension to and growth within adjacent bronchial lumina is unusual. Hemorrhage and necrosis are frequent. Microscopically, the tumor is an admixture of primitive epithelium and stroma that superficially resembles the pseudoglandular period of lung development. The epithelial cells

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are arranged in small slit-like spaces or branching tubules and are surrounded by polygonal or spindle-shaped stromal cells with hyperchromatic nuclei. Metastases may show predominance of either epithelial or stromal components and may sometimes show both.

Surgery is the only relatively satisfactory treatment. Neither chemotherapy, which has been tried empirically with multiple drug schedules including vincristine, cyclophosphamide, adriamycin, CCNU, and VP16, nor radiotherapy, administered at different doses, appears to influence the lifespan of treated patients.

The prognosis of pneumoblastoma is difficult to determine for any individual. Of 39 patients reviewed by Fung and colleagues,17 developed metastases, and only two survived more than two years. Despite this, occasional patients survive longer, even in the presence of metastatic disease.5,7,8

Compared with the three cases seen previously at the Geneva University Hospital, for whom survival was from three to eight months, our patient, operated on in March 1989, had a survival of two years before tumor recurrence was detected. To our knowledge, a case of a pneumoblastoma arising in the context of neurofibromatosis has not been described previously in the literature.

von Recklinghausen’s disease is a common autosomal disorder linked to chromosome 17. The condition presents in its classic form with a triad consisting of multiple nerve tumors, pigmented skin nodules called “café au lait patches,” and pigmented nodules of the iris.9

Abnormalities of other organ systems are common. Skeletal lesions such as scoliosis of the spine and multiple intraosseous and subperiosteal cysts have been described. Affected individuals are at high risk of developing various tumors. In particular, there is a common association between von Recklinghausen’s disease and tumors of the peripheral and central nervous system, irrespective of histologic condition. Many other tumors have, however, been reported in neurofibromatosis.10 They are grouped as follow: (a) those of presumed neural crest origin such as neuroblastoma, phaeochromocytoma, medullary thyroid carcinoma, and melanoma; and (b) those traditionally thought not to be derived from the neural crest such as Wilm’s tumor, rhabdomyosarcoma, and leukemia.

Common adult cancers, including breast, colon, and lung, seem to be underreported in association with neurofibromatosis. The first case of colon cancer associated with von Recklinghausen’s disease was reported in 1972.11 Yet, the presence of common malignancies, including colon cancer in Danish and American studies,12 seems to refute any notion that the neurofibromatosis gene may protect against certain malignancies.

In this context, our case appears to be the first pneumoblastoma described in the literature in association with von Recklinghausen’s disease. A better understanding of the pathogenesis and etiology of both pneumoblastoma and neurofibromatosis may help elucidate the role of the neurofibromatosis gene in the development of unusual tumors in this common disorder.

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REFERENCES
Idiopathic Hemorhorax*

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Development of spontaneous hemothorax without predisposing conditions is extremely rare. We report a young man with a history of a seizure disorder who presented to the emergency department with spontaneous hemothorax. Exploratory thoracotomy evacuated 2,000 ml of old blood. No source of hemorrhage was identified. To our knowledge, this is the first report of spontaneous hemothorax proved by thoracotomy. (Chest 1993; 103:638-39)

DIC = disseminated intravascular coagulation

Although hemothorax is a relatively common entity, it is usually secondary to associated factors such as trauma, coagulopathy, pleural or other anatomic abnormalities, pulmonary infarction, necrotizing infection, aortic dissection, or malignant neoplasm. In managing cases of acute hemothorax, a search to identify the source of hemorrhage is critical. Invasive procedures such as chest tube placement or exploratory thoracotomy may be inevitable.

Case Report

A 35-year-old man presented to the emergency department with a four-week history of poor oral intake, generalized weakness, dyspnea on exertion, and lower extremity purpuric skin lesions. No chest pain, hemoptysis, hematemesis, or melena were reported. There was no history of recent traumatic injury or seizure.

Posttraumatic seizures had been diagnosed one year prior to hospital admission, for which he took phenytoin sodium. He drank 12 beers per day, but had had none for the ten days prior to hospital admission. He had smoked one pack of cigarettes per day for five years, but denied intravenous drug abuse, homosexual contact, or transfusions.

Physical examination revealed a cachectic young man with blood pressure of 106/68 mm Hg, pulse of 90/min, respiratory rate of 18/min, and temperature of 36.6°C. Multiple nontender purpuric lesions were present diffusely on both legs. Decreased breath sounds and dullness to percussion were appreciated over the right lung. Results of his examination were otherwise completely normal.

His hematocrit was 24.7 percent, decreased from 42.7 percent two weeks prior to hospital admission. Other laboratory analyses, including complete blood cell count with differential and platelets, electrolytes, liver injury panel, bleeding time, and coagulation profile were normal. His serum albumin, however, was decreased (2.5 g/dl).

His chest roentgenogram (Fig 1) showed multiple pleural-based masses. These were shown to be fluid collections by ultrasonography. Eight hours after hospital admission, his hematocrit had fallen to 14 percent, his pulse had risen to 104/min, and his blood pressure had fallen to 86/60 mm Hg. A chest roentgenogram (Fig 2) showed a significant increase in the sizes of the fluid collections. A chest computed tomographic (CT) examination confirmed multiple loculated pleural fluid collections. A thoracentesis revealed frank blood.

The patient was transfused with two units of packed red blood cells, and thoracotomy was performed the following morning. Old blood, 2,000 ml, was evacuated but no source of hemorrhage was identified. The pleura appeared completely normal and no biopsies were performed.

The patient's hospital course was complicated by postoperative acute adult respiratory distress syndrome (ARDS) and disseminated intravascular coagulopathy (DIC). Additional studies included normal complement levels (C3, C4, and CH50), negative human immunodeficiency virus (HIV), ANA, RF, Raji cell, and antinuclear cytoplasmic antibody. Biopsy specimen of his lower extremity lesions showed only nonspecific dermatitis.

The patient eventually made a full recovery and is currently functioning well in his daily activities.

Discussion

To our knowledge, only two cases of idiopathic spontaneous hemothorax have been reported in the English literature. Promisloff and Friehling reported hemothorax in a healthy young man following heavy lifting; however, no surgical exploration was performed. Dimitri described a woman with left hemothorax, in whom thoracoscopy and multiple pleural biopsy specimens revealed nonspecific inflammatory changes.

Our patient presented initially with a hemothorax and rebled during the first 24 h of hospitalization. Thoracotomy with careful surgical exploration failed to demonstrate any

FIGURE 1. Posteroanterior chest roentgenogram taken at the time of hospital admission showing multiple pleural fluid collections.