on the farm. BAL done during an acute febrile attack (December 1990) showed a remarkably high number of neutrophils. An increase in BAL neutrophils has been described after provocation challenges of HP and in acute HP. The neutrophil count decreased at the control BAL obtained during remission (January 1991). His BAL cellular findings were very different from what is seen in ODTS, where a high neutrophil count and no increase in lymphocytes is found.4

Our patient presented with recurrent bouts of acute febrile episodes over at least eight winters without ever developing clinically significant evidence of lung involvement, that is, progression to overt FL. The repeated occurrence of symptoms without obvious massive antigenic exposure (even while wearing a protective mask), the results of BAL, and the absence of symptoms in the other workers on the same farm rule out the diagnosis of ODTS.

The lack of persisting precipitins to common antigens responsible for FL may be significant. However, in our community, 40 percent of farmers who develop FL have such a negative result when tested only by double diffusion. A normal chest roentgenogram in acute FL is not unusual but would be very unlikely after multiple recurrences of the disease.5 The normality of pulmonary functions also goes against a diagnosis of FL. The patient did have some lung involvement: abnormal BAL, cellular infiltrates on histopathologic study, and the presence of inspiratory crackles on one occasion. These lung abnormalities did not cause clinical lung disease and are probably not different from what is seen in some asymptomatic farmers.6

This subject had an acute lung inflammatory reaction similar to what is seen in acute FL or in asymptomatic exposed FL patients. As do subjects with acute FL, he presented systemic symptoms on antigenic exposure. However, contrary to acute FL, his lung inflammation was not associated with dyspnea, inspiratory crackles, interstitial infiltrates, or a decrease in lung function. We therefore believe that this recurrent fever, although related to barn contact, did not meet the diagnostic criteria for FL.

We have recently demonstrated that Micropolyspora faeni, the most frequent causative antigen in FL in our community, can induce the liberation of proinflammatory cytokines by macrophages.8 We believe that it is possible that such mediators were released by lung cells of our patient, producing the systemic febrile reaction, but have no explanation as to why he did not develop respiratory symptoms, abnormal lung function, or lung infiltrates.

Whether this case represents an isolated observation or a more common not reported entity awaits further studies. The relationship between this case and FL or ODTS remains unclear. Perhaps some of the farmers reported as having febrile reactions, assumed to have ODTS, in the epidemiologic survey of Malmberg et al.,8 had an entity similar to that of our subject.

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Pulmonary Hemorrhage in Association with Autoimmune Chronic Active Hepatitis*

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Extrahepatic manifestations have been reported in a majority of patients with ACAH. We describe an 11-year-old girl with CAH who developed life-threatening AH and was successfully managed with mechanical ventilation and high-dose steroids. Although a number of pulmonary manifestations have been described in association with ACAH, this article is the first showing its association with AH.

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AH = alveolar hemorrhage; ACAH = autoimmune chronic active hepatitis; ANA = antinuclear antibody; CAH = chronic active hepatitis; PT = prothrombin time; FTT = partial thromboplastin time; SLE = systemic lupus erythematosus

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Pulmonary Hemorrhage in Chronic Active Hepatitis (Kagalswalla et al)
Autoimmune chronic active hepatitis is a disorder that is not confined to the liver alone. In one large series, 57 percent of the patients had involvement of at least one organ other than the liver. A number of pulmonary abnormalities including pleurisy, pleural effusion, segmental collapse and fibrosing alveolitis have been described. We report a case of life-threatening AH in a child with AIHCA. To our knowledge, this extrahepatic manifestation has not been described previously.

CASE REPORT

An 11-year-old girl was transferred to the ICU of our hospital with shortness of breath and hemoptysis. The Hb value was 50 g/L, which was reduced from 83 g/L two weeks earlier. The accompanying chest x-ray film showed extensive confluent alveolar infiltrates in both lungs.

Four months prior to this admission, she had jaundice which resolved in ten days and recurred six weeks later with hepatosplenomegaly. The Hb value was 94 g/L. Sedimentation rate was 97 mm/h. The total protein level was 93 g/L, and albumin value was 29 g/L. Predominantly direct hyperbilirubinemia was present and values for transaminases were 14 times normal. Serology for hepatitis A, hepatitis B, hepatitis C, cytomegalovirus and Epstein-Barr virus were negative. Four weeks later, the cholestasis and the liver function tests were unchanged. Serum ceruloplasmin and alpha-antitrypsin were normal. The ANA titer was 1:160 and SMA was positive. Anti-liver kidney microsomal, anti DNA and antimitochondrial antibodies were negative.

Two weeks later, she developed a febrile illness with respiratory distress requiring the recent admission to the ICU. Her pulse was 150/min; respirations, 80/min; temperature, 39.8°C; and blood pressure, 120/50 mm Hg. Coarse crepitations were present bilaterally. Liver was 5 cm and spleen 6 cm below the costal margin. The Hb value was 50 g/L, which was reduced from 83 g/L two weeks earlier. The WBC count with a differential cell count was normal. Platelet count was 19.4 x 10^9/L; PT, 17.5 s (normal, 10 to 14 s); PTT, 39.1 s (control, 25 to 35 s). Serum electrolytes, blood urea nitrogen and creatinine were within normal limits. Urinalysis was negative for red blood cells, proteins and casts. Creatinine clearance was normal. Total bilirubin level was 34 µmol/L, with a direct value of 23 µmol; alkaline phosphatase, 365 U/L; SGOT, 322 U/L; SGPT, 253 U/L. Admission chest x-ray film compared with the previous x-ray film done 18 h earlier showed extensive alveolar infiltrates compatible with AH (Fig 1).

Arterial blood gas value analysis revealed hypoxemia with a pH of 7.40; Pco₂, 33 mm Hg; Po₂, 49 mm Hg; oxygen saturation, 79 percent; and HCO₃⁻, 20 mEq/L. During intubation, 12 ml of fresh blood was suctioned from the endotracheal tube. Tracheal aspirate, which was negative for acid-fast bacilli and Pneumocystis carinii, had a few hemosiderin-laden macrophages. Mechanical ventilation with high positive end-expiratory pressure and therapy with corticosteroids were started. Treatment with ampicillin and gentamycin was begun; blood and fresh frozen plasma also were administered. Fresh frozen plasma was continued to correct the coagulopathy. Marked clearing in both lungs was seen within 48 h (Fig 2). She was extubated after 17 days and discharged on a regimen of prednisone after seven weeks of hospitalization.

Three months later, a percutaneous liver biopsy showed piecemeal necrosis, bridging fibrosis and regenerating nodules consistent with a diagnosis of CAH with very early cirrhosis (Fig 3).

FIGURE 1. Admission chest radiograph showing confluent alveolar consolidation in the right lung and the central portion of the left lung compatible with extensive bilateral pulmonary hemorrhage.

FIGURE 2. Chest radiograph 48 h after admission showing marked clearing of the alveolar opacities.

FIGURE 3. Needle biopsy of the liver showing moderately severe CAH with fibrous septae extending from the enlarged portal tract. The interface between parenchyma and connective tissue is irregular (hematoxylin and eosin, original magnification ×200).
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.7 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.8 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.8,9 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.10,11 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.

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Pneumoblastoma in Neurofibromatosis*

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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.

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CASE REPORT

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,14 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,15,16 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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