Pediatric Idiopathic Pulmonary Hemosiderosis Diagnosed by Sputum Analysis: Plain Radiography and Computed Tomography Findings

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Key Words

Idiopathic pulmonary hemosiderosis · Computed tomography · Chest radiography · Hemoptysis

Abstract

Objective: Idiopathic pulmonary hemosiderosis (IPH) is an uncommon disorder, which is characterized by recurrent hemoptysis, iron deficiency anemia and diffuse parenchymal infiltration on chest radiographs in pediatric patients. We wish to present clinical and radiological (plain radiography and CT) findings of this rare pathology. **Clinical Presentation and Intervention:** A 14-year-old girl was admitted to the pediatric emergency department with complaints of cough, dyspnea, fatigue and bloody sputum for 6 months. She had been hospitalized 3 times during this period and received antibiotics and blood transfusion. Chest X-rays revealed prominent perihilar and bibasilar consolidation. CT showed a ground glass pattern and consolidated areas with increased density. Sputum analysis yielded hemosiderin-laden macrophages. With presumptive diagnosis of IPH, prednisolone was administered. Her symptoms improved on the 5th day of treatment and 1 month later, plain chest radiography demonstrated marked improvement. **Conclu-**

sion: Although IPH is a rare condition, the diagnosis of IPH should be considered, among others, in a patient with hemoptysis and bilateral infiltration in the chest X-ray. This may prevent antibiotic misuse and risk of death due to severe hemorrhage.

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Introduction

Idiopathic pulmonary hemosiderosis (IPH) is an uncommon disorder, which is characterized by recurrent hemoptysis, iron deficiency anemia and diffuse parenchymal infiltration on chest radiographs [1–3]. This disease is usually encountered under age 10, however it may arise in the second decade and is diagnosed by open lung biopsy [2]. If the biopsy cannot be performed, the clinical presentation of the patient, in the absence of other causes of pulmonary hemorrhage, may establish the diagnosis [3]. We present a case of IPH with typical clinical and radiological findings and hemosiderin-laden macrophages in the sputum.

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Fig. 1. Posteroanterior chest radiograph showing bilateral air space consolidation areas.

Case Report

A 14-year-old girl was admitted to the pediatric emergency department with complaints of cough, dyspnea, fatigue and bloody sputum for 6 months. She was hospitalized 3 times during this period and received antibiotics and blood transfusion. She had no family history of lung disease. On physical examination her blood pressure was found to be 110/65 mm Hg. The patient was pale and had tachypnea, tachycardia, crepitating rales and wheezing. Laboratory data included hemoglobin of 9.7 g/dl, hematocrit of 28%, and MCV of 61.2 fl. The platelet count was 380,000/mm³ with iron deficiency anemia. Acute phase reactants and WBC were normal. Sputum culture, PPD test and acid-resistant bacteria tests were negative. Coagulation and blood-urine biochemical tests were within normal limits. The stools were positive for occult blood and sputum analysis yielded hemosiderin-laden macrophages elevated 3-fold. Chest X-ray revealed prominent perihilar and bibasilar consolidation (fig. 1). CT (10 mm thickness, 120 kVp, 200 mAs, Hitachi 1000, Tokyo, Japan) showed a ground glass pattern and consolidated areas with increased density as seen in patients with alveolar hemorrhage (fig. 2). The patient refused to undergo an open lung biopsy. Milk radioallergosorbent test, antineutrophilic cytoplasmic antibody test, rheumatoid factor, anticardiolipin antibody and anti-DNA tests were all negative. Immunoglobulins (IgG, M, A) and complement (C3, C4, CH5O) were normal. Electrocardiography and echocardiography revealed findings of slight right ventricular overload. With presumptive diagnosis of IPH, prednisolone (2 mg/kg/day) was administered. The patient's symptoms improved on the 5th day of treatment and she was discharged from the hospital on the 14th day. One month later, plain chest radiography demonstrated marked improvement (fig. 3).

Fig. 2. Contrast-enhanced CT (120 kVp, 200 mAs, thickness: 10 mm, normal reconstruction algorithm) of the chest demonstrating bilateral marked air space consolidation and increased density areas supporting alveolar hemorrhage. a Mediastinal window, b Lung window.

Discussion

Virchow first described the pathologic findings of IPH in 1864 and clinical and radiological diagnosis was described by Waldenstrom in 1944 [cited after ref. 4]. Estimated incidence of this disorder is 0.24–1.23 per 1,000,000 population [3]. There is no gender (female:male ratio of 1:1), racial or geographic propensity [5]. Familial tendency has been described [6]. Recurrent multifocal alveolar capillary hemorrhage is responsible for the clinical

Fig. 3. Follow-up chest radiograph showing marked improvement compared to the previous findings.

cal and laboratory findings [7]. Etiology is obscure but the dramatic response to steroid treatment suggests an immunologic defect in the basal membrane of the pulmonary capillaries [7]. The clinical course may vary but most patients die in the early phase of the disease because of acute pulmonary hemorrhage. However, complete remission can be achieved with early diagnosis and treatment. Some patients may progress to chronic pulmonary fibrosis, chronic respiratory insufficiency, and ultimately pulmonary hypertension [3]. The differential diagnosis includes Goodpasture's syndrome with or without renal involvement, hemosiderosis secondary to vasculitis, pulmonary embolus, pulmonary venous hypertension such as long-standing mitral valvular stenosis, pulmonary vein stenosis and veno-occlusive disease, and bleeding diatheses due to cancer or infections and lymphangioleiomyomatosis [8]. Our case could not be linked to any of these diseases. Therefore, in the absence of a gold standard test, IPH was the diagnosis of choice by exclusion, given the histologic evidence.

Histologically, the unifying features of diffuse pulmonary hemorrhage are the presence of recent hemorrhage in alveolar spaces and hemosiderin-laden macrophages in alveolar spaces and interstitium [2]. The presence of the

hemosiderin-laden macrophages in the sputum reflects the presence of pulmonary alveolar hemorrhage, but does not reflect any specific cause.

Typical radiographic findings are air space consolidation and ground glass appearance [2]. Perihilar areas and both lung bases are mostly affected [9]. The apexes and costophrenic angles are typically unaffected. However if the parenchymal involvement is extensive, as in our case, costophrenic angles and apexes may also be affected [9]. Consolidation areas may turn to a reticular pattern within a few days that may then be resorbed completely, as in our case, or may advance to fibrosis with recurrent episodes of pulmonary hemorrhage. Lymphadenopathy and pleural effusion are not common [9]. While the sensitivity of CT is superior to that of the plain film in displaying the characteristic acinar pattern [1], the CT findings are nonspecific because pulmonary edema and alveolar hemorrhage or other diffuse air space disorders, all display similar CT changes. Nonetheless, a diffuse dense acinar pattern supports the alveolar nature of the pulmonary pathology [1]. The reason of the increased density in the CT is intra-alveolar hemorrhage and/or hemosiderin-laden macrophages. The pattern of acute alveolar hemorrhage may be more easily distinguished by CT [1] than by plain X-ray films, which may provide images that mimic pneumonia and thus result in diagnostic delay and unnecessary antibiotic therapy [10]. Sometimes the source of hemorrhage is extrapulmonary and the blood gets aspirated into the alveoli, as in endobronchial tuberculosis, or the blood may spread from one segment of the lung to another or from one to the contralateral side [11, 12].

Corticosteroids are the initial treatment of choice. If the steroids are ineffective, cytotoxic drugs may be used. Long-term prognosis of IPH is poor and most patients develop pulmonary fibrosis 5 years after the initial diagnosis [7].

Conclusion

Although IPH is a rare condition, in a patient with hemoptysis and bilateral scattered air space disease in the chest X-ray, the diagnosis of IPH should be considered, among others. This may prevent antibiotic misuse and risk of death due to severe hemorrhage.

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