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Case Report

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Idiopathic Pulmonary Hemosiderosis in Children: A Case Report

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1. Introduction

Idiopathic pulmonary hemosiderosis (IPH) is a rare respiratory disease of unknown etiology characterized by repeated episodes of a diffuse alveolar hemorrhage resulting in the accumulation of hemosiderin inside pulmonary macrophages. We present a case of children with pulmonary hemosiderosis, collected at our department of infectious and respiratory diseases.

2. Case Report

It was a 21-month-old infant female, with antecedents of prematurity 35 weeks of menorrhoea, birth weight 1700g with several hospitalizations for severe anemia. History of the disease goes back to the age of 5 months by dry cough and hemoptysis. Clinical examination finds a conscious infant tonic reactive apyretic, with generalized cutaneous-mucosal heat, discolored conjunctiva and staturopunderal delay. Thus, diagnosis evoked was a pulmonary hemosiderosis. Blood tests had objectified a low hemoglobin level at 3.5, VGM at 63 and CCMH at 28, white blood cells level at 7690, neutrophils at 4710, lymphocytes 2at 450 and platelets at level of 154000. Besides, CRP and VS were normal, TP 100%, TCA 1 and fibrinogen level at 2.2.

X-ray of the lung showed discrete reticulomicronodular opacities at the lung bases (Figure1) and thoracic CT-scan demonstrated ground-glass attenuation in a similar distribution, as seen in a standard chest x-ray associated to homogeneous mediastinal adenopathy (Figure 2), gastric tubing with search for Koch's bacilli was negative, also anti-gliadin and anti-transglutaminase antibodies were negative. However, sputum testing found siderophages with GOLDE index over 200. Treatment consisted of a transfusion of red blood cells and oral steroid therapy, evolution was favorable marked by a regression of the cough and a decrease in the frequency of hemoptysis.

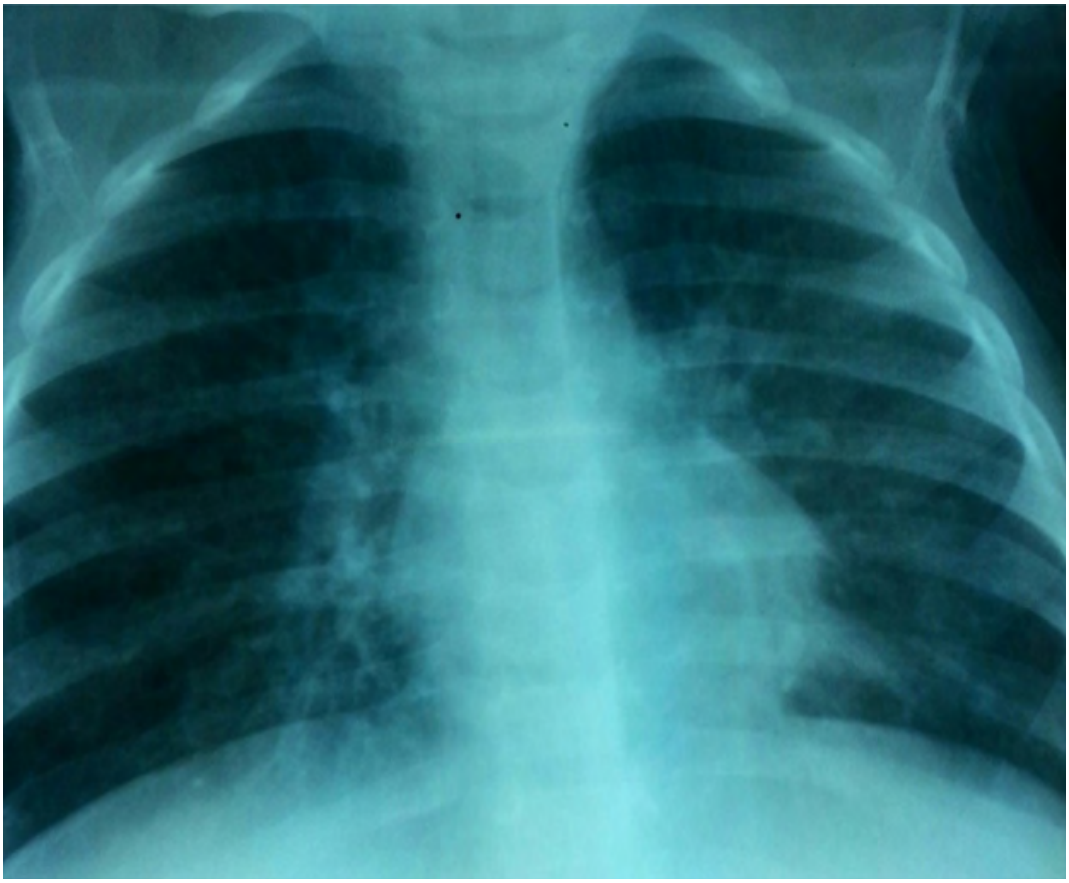

Figure 1: X-ray of the lung showing several discrete reticulomicronodular opacities.

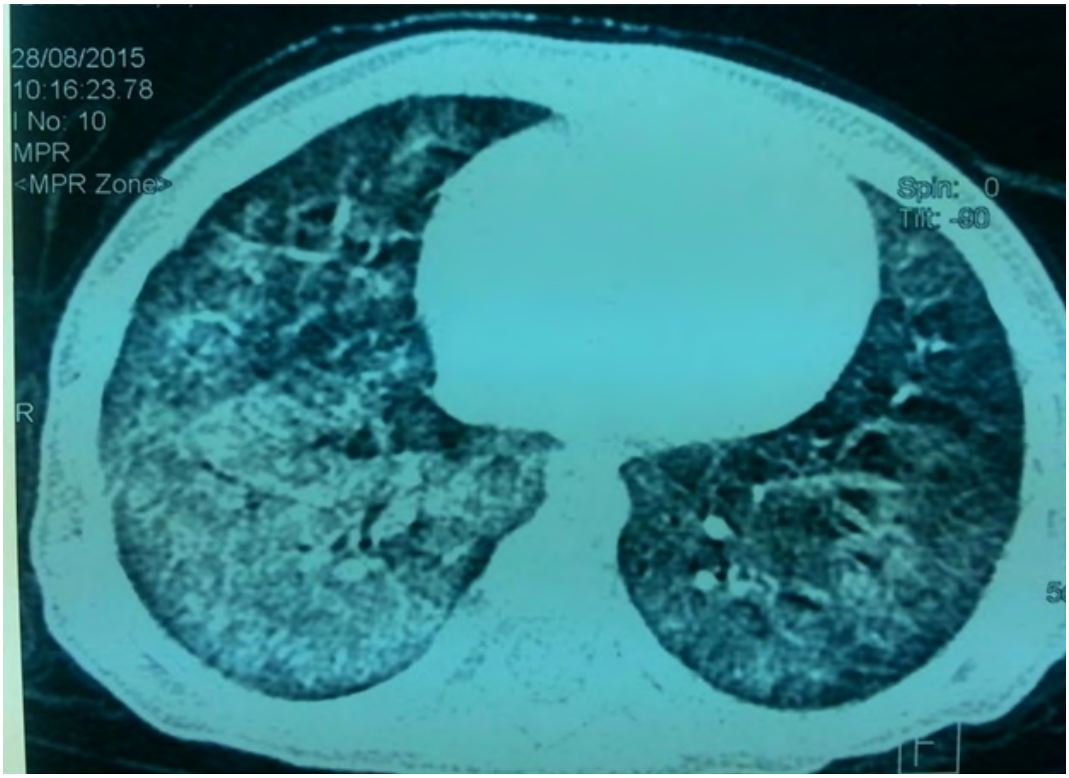

Figure 2: CT-scan revealed bilateral and diffuse ground-glass appearance with homogeneous mediastinal adenopathy.

3. Discussion

Idiopathic pulmonary hemosiderosis was first described by Virchow in 1864. Its incidence and prevalence are undetermined, about 300 cases have been reported in the literature. It is a disease of children, often beginning before the age of 10 years. Diagnosis is based on the triad; hemoptysis, diffuse pulmonary alveolar-interstitial infiltrates and iron deficiency anemia, evolving in «relapses-remissions». The BAL confirms the diagnosis of IPH by showing the presence of siderophages, with a Golde score > 80. EFR shows at a late stage, a restrictive syndrome with a decrease in DLCO. The etiology is uncertain but likely to be multifactorial. Possible associations include toxic insecticides (epidemiological studies in rural Greece), premature birth, and fungal toxin exposure. It may be associated with cow's milk protein allergy or celiac disease defining Lane-Hamilton syndrome and Heiner syndrome respectively.

Treatment is based on the presentation and acute versus chronic nature of the patient. There have been trials/studies done, including elective splenectomy, high dose systemic steroids, inhaled steroids, and immunosuppressant drugs. Blood should be transfused to correct severe anemia and shock. A study by de Jongh et al. recommended screening for hemosiderosis in all patients receiving multiple transfusions. Of the many approaches to idiopathic pulmonary hemosiderosis (IPH) treatment, systemic steroids, and immunosuppressant therapy have shown the most favorable outcomes when used independently or in combination. Current recommendations for an acute phase IPH call for 0.5 to 0.75 mg/kg per day (up to 60 mg/day) initial dosing of prednisolone, which is tapered once chest radiograph shows improvement in new opacifications. Lung transplantation seems to be problematic because of the risk of recurrence on the graft [1-3]. Evolution is variable, ranging from chronic anemia and progressive respiratory insufficiency (due to pulmonary fibrosis) to massive pulmonary hemorrhage, putting the vital prognosis at stake.

4. Conclusion

IPH is a rare disease characterized by repeated recurrent episodes of diffuse alveolar hemorrhage due to abnormal accumulation of hemosiderin in the pulmonary tissue. Early recognition and prompt diagnosis can help reduce the high morbidity and mortality associated with this condition.

5. Acknowledgments

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6. Conflict of Interest

The authors declare that they have no conflict of interest.

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