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RESEARCH ARTICLE

IDIOPATHIC PULMONARY HEMOSIDERIN: ABOUT AN UNUSUAL CASE

Karima EL Fakiri, Ghyzlane Draiss, Noureddine Rada and Mohammed Bouskraoui
Pneumopediatric and Allergology Unit, Paediatric Department A, Children University Hospital Mohammed VI, FMPM Cadi Ayyad Marrakesh, Morocco.

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Abstract

Introduction: idiopathic pulmonary hemosiderosis is a rare disease of unknown etiology.
Case report: This is a 12-year-old boy. The beginning of its symptomatology dates back to one year by several episodes of low-abundance hemoptysis associated with cough and night sweats. He had also a microcytic hypochromic anemia (Hb=4.4) transfused 3 times. . Clinical examination found discoloured skin paleness and conjunctival The count showed microcytic hypochromic anemia with HB:5.7 g/dl. A chest x-ray showed a discrete alveolar syndrome. A tuberculous assessment in particular an IDR tuberculin 3 BK expectorations and Gènexpert were negative. A chest CT scan showed bilateral interstitial syndrome. A bronchoscopy with bronchoalveolar lavage showed the existence of sidephages with a Golde score calculated on 240 Perls staining . A balance of system diseases including antinuclear antibodies were negative. Ac anti transglutaminase IgA were negative. A heart ultrasound did not show a pulmonary hypertension. In fine, the diagnosis was a primary pulmonary hemosidesis. Management consisted of a blood cell transfusion a prenisolone-based corticosteroid 2 mg/kg/d for 3 months. After two months, the improvement was full.
Conclusion: We must think about Hemosidesis idiopathic if we have hemoptysis chronic anemia and pulmonary infiltrates and do a Bronchoalveolar lavage to confirm diagnosis.

Introduction:-

Idiopathic pulmonary hemosidesis (IPH) is a rare cause of alveolar bleeding in children. It is often manifested by anemia ferriprive hemoptysis and infiltrates to the chest x-ray. Bronchoalvlary lavage confirms the diagnosis. Management is poorly codified. Thus we report a rare and unusual case of idiopathic pulmonary hemosidesis after free and informed consent of the parents

Case Report:

This is a 12-year-old boy from an unrelated marriage, vaccinated according to the national immunization program, with no known recent tuberculosis infection or heart disease. The beginning of its symptomatology dates back to one year by several episodes of low-abundance hemoptysis associated with cough and night sweats. All evolving in a context of febrile sensation and alteration of the general state made asthenia, anorexia and weight loss quantified at 3 kg. In addition, it is followed for microcytic hypochromic anemia (Hb=4.4) transfused 3 times. Clinical examination

Corresponding Author:- Karima EL Fakiri
Address:- Pneumopediatric and Allergology Unit, Paediatric Department A, Children University Hospital Mohammed VI, FMPM Cadi Ayyad Marrakesh, Morocco.

found a conscious patient with 20 respiratory frequency =107 heart rate; T= 37; SaO2 = 99% open air TA: 119/65 mmhg Weight = 30Kg

(-2DS) Height = 141cm (-1DS), discoloured skin paleness and conjunctival. He had no digital hippocratism. The pleuropulmonary examination found no rails at auscultation. The cardiovascular examination was normal. The clinical examination was non-specific. The count showed microcytic hypochromic anemia with HB: 5.7 g/dl, VGM:59.7 and TCMH:14.9. A chest x-ray showed a discrete alveolar syndrome (Figure 1). A tuberculous assessment including an IDR tuberculin 3 BK expectorations and Gènexpert were negative. A chest CT scan showed bilateral interstitial syndrome (Figure 2). A bronchoscopy with bronchoalveolar lavage showed the existence of sidephages with a Golde score calculated on 240 Perls staining (Figure 3). Respiratory functional exploration found no restrictive syndrome.

A balance of system diseases including antinuclear antibodies: ANCA (p+c): Antibodies anti MBG Ac anti-ENA (SSB, SSA, Sm) were negative. Ac anti transglutaminase IgA were negative. A heart ultrasound did not show a pulmonary hypertension. In fine, the diagnosis was a primary pulmonary hemosidesis. Management consisted of a blood cell transfusion a prenisolone-based corticosteroid 2 mg/kg/d for 3 months then progressive degression associated with a salt-free diet and adjuvant treatment and inhaled corticosteroid therapy. After a month of retreat, there was a full improvement in clinical symptomatology without any episode of hemoptysis.

Discussion:-

Pulmonary hemosiderosis is a very rare entity, possibly of the immunologic mechanism, causing a defect in the basement membrane of the pulmonary capillary. The estimated incidence of IPH in children is 0.24- 1.23 cases per million, with a mortality rate as high as 50% [1, 2]. In literature there is a female predominance but our patient was male [3]. The diagnosis is done most late; it was one year with our patient. It is probably due to the fact that iron deficiency anemia may be the first and the only manifestation of IPH, preceding other symptoms and signs by several months [4]. Iron deficiency anemia is the most common haematologic disorder seen in childhood, we must think about IPH if we have anemia hemoptysis and respiratory symptoms.

The radiographic appearance of IPH is rather non specific and depends on the acuity and severity of alveolar hemorrhage. During the acute episode, chest X-ray usually shows diffuse alveolar opacity, including consolidation involving the peri-hilar and lower lung zones However, in case of massive hemorrhage, all lung zones may be involved.

High resolution computed tomography of the chest is more sensitive and might show affected areas not previously identified by chest X-rays. It ‘s often shows ground glass opacities[5]. Our patient had bilateral interstitial syndrome .

Idiopathic pulmonary hemosiderosis is a diagnosis made by the exclusion of other causes. It should be confirmed by the presence of many hemosiderinladen macrophages in bronchoalveolar lavage fluid obtained by bronchofibroscopy, which is done in our case

It is possible to find at the coloring of Perls macrophages loaded with hemoglobin called siderophages, whose rate is abnormal when it is greater than or equal to 20%. Quantification of the intensity of sidero phage marking by the coloration of Perls. Each cell is rated from 0 to 4. The score established on 100 sidephages varies between 0 and 400. It is considered as pathological above 100 [6]. It confirm diagnosis in our patient

IPH is in some cases associated with celiac disease, so patients with idiopathic pulmonary hemosiderosis should routinely be tested for gluten intolerance. Recently, other concomitant food allergies have been reported. Our patient didn’ have a caeliac disease.

Treatment is based on first-line monthly intravenous bolus corticosteroid therapy combined with oral corticosteroid therapy. Hydroxychloroquine may be used as a second line [7]. The monitoring is clinical and radiological every 3 to 6 months. prognosis still unknown

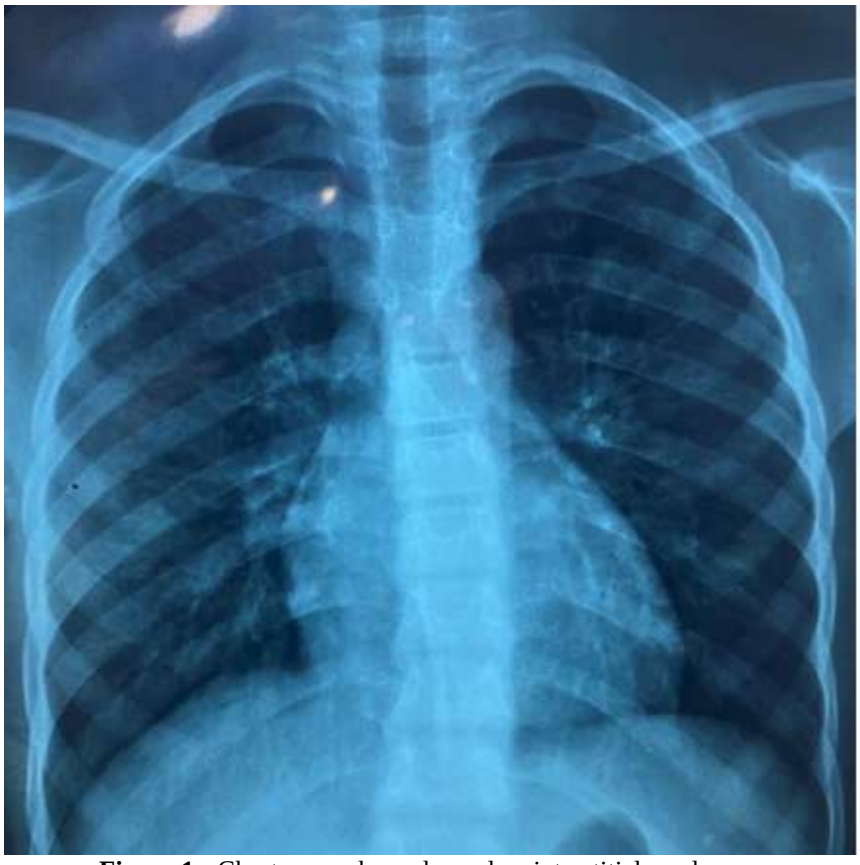

Figure 1:- Chest x-ray showed un alveointerstitial syndrome.

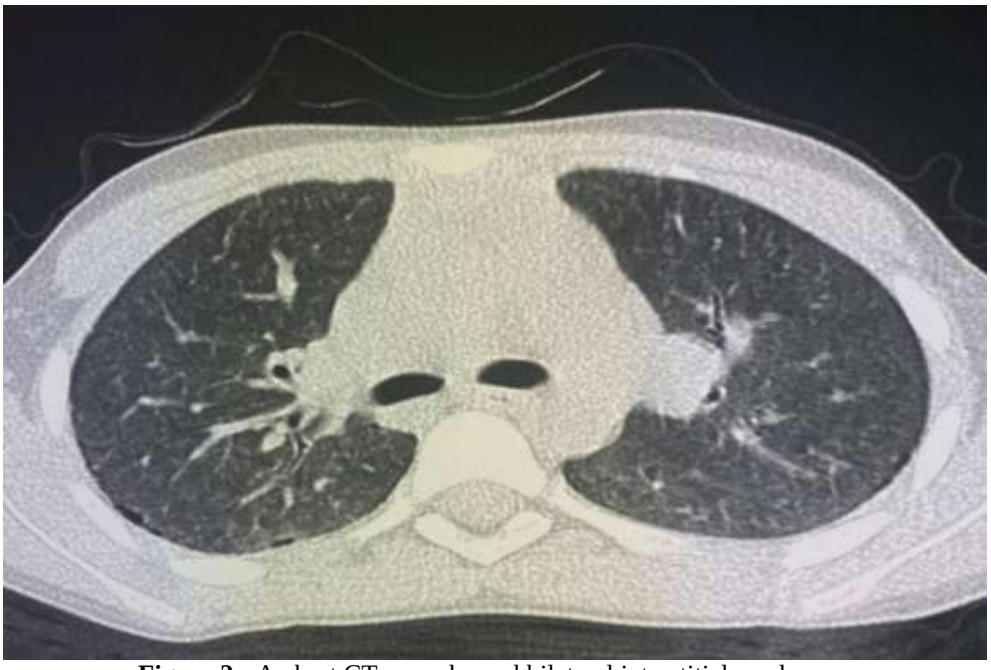

Figure 2:- A chest CT scan showed bilateral interstitial syndrome.

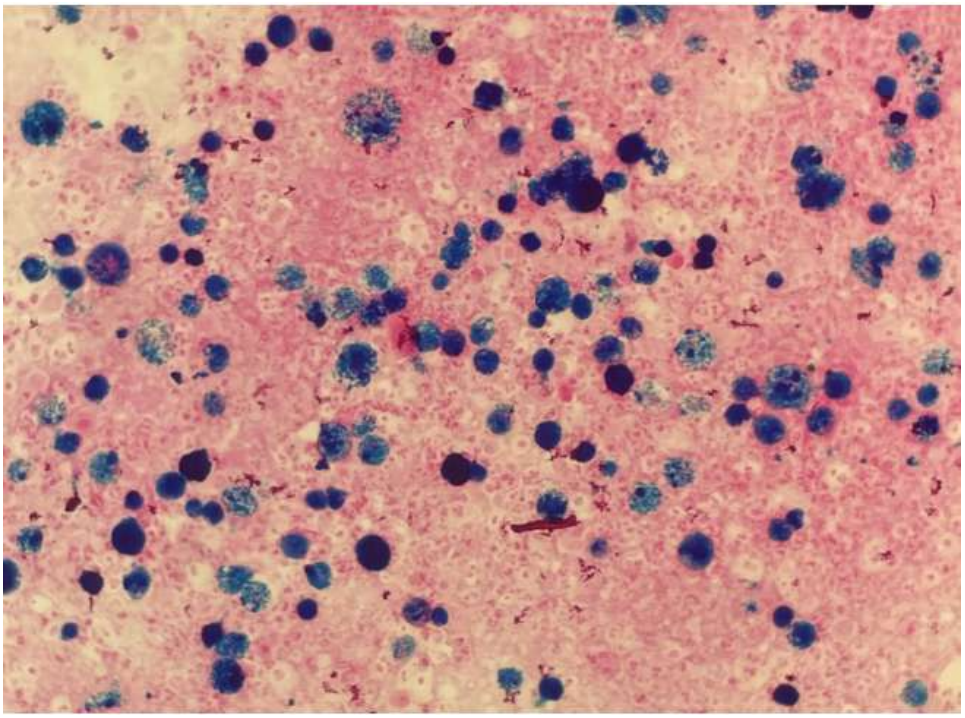

Figure 3 : Bronchoalveolar lavage showed the existence of sidephages with a Golde score calculated on 240 Perls staining.

Conclusion:-

Hemosidesis idiopathic pulmonasia is a rare condition.We has to think about it if we have hemoptysis chronic anemia and pulmonary infiltrates. Bronchoalveolar washing is essential for diagnosis; we must always seek autoimmunity. Finally, corticosteroid therapy is the basis of treatment.

Conflict of interest:
None.

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