ECRONICON OPEN ACCESS

EC PULMONOLOGY AND RESPIRATORY MEDICINE

Case Report

A Rare Case Report of Idiopathic Pulmonary Hemosiderosis and Juvenile Idiopathic Arthritis (RF Positive Polyarthritis)

Eman Shhada[1]*, Basheer Khalil[1] and Sawssan Ali[2]

[1]*Rheumatology Paediatrics Department, Children's Damascus University Hospital, Syria*

[2]*Pulmonary Paediatrics Department, Children's Damascus University Hospital, Syria*

***Corresponding Author:** Eman Shhada, Department of Paediatrics, Children's Damascus University Hospital, Damascus University, Damascus, Syria.

Received: September 24, 2019; **Published:** October 23, 2019

Abstract

The concomitant between idiopathic pulmonary hemosiderosis (IPH) and juvenile idiopathic arthritis (JIA) is extremely rare and a few cases were reported in the literature, four cases was published about the association between JIA and IPH. We report a case of 3-years old female admitted for arthritis, refractory anemia and recurrent chest infection. The girl has been diagnosed with IPH JIA. After treatment with prednisolone and MTX, the improvement in pulmonary and joint symptoms were very well during 1 year follow up.

Keywords: *Idiopathic Pulmonary Hemosiderosis; Juvenile Idiopathic Arthritis*

Introduction

Idiopathic pulmonary hemosiderosis (IPH) is extremely rare (the incidence of IPH is 0.24 - 1.23 patient per million children), characterized by the trade if hemoptysis, iron deficiency anemia and diffuse alveolar infiltration on chest imaging [5,6] it's remains idiopathic. There are a few reports of IPH combination with autoimmune diseases such as celiac disease, vasculitis and juvenile idiopathic arthritis [1,8]. In our patient we report a case of a combination between IPH and JIA. Juvenile idiopathic arthritis (JIA) is a chronic arthritis, which involved all forms of arthritis of unknown etiology lasting for at least 6 weeks and with onset before the age of 16 years [7]. Up to date, four cases of IPH associated with juvenile idiopathic arthritis JIA has been reported in the literature previously (seronegative polyarthritis and oligoarticular JIA) [1-4]. Hence that our case is the fifth report about this association in the literature.

Case Presentation

A 3-years old female admitted to hospital because of mild grade of intermittent fever, arthralgia and arthroncus (arthritis), refractory iron anemia without response to iron treatment and recent history of recurrent chest infection.

On physical examination, failure to thrive, the body weight 8 kg and height 77 cm (under - 3 percent), overt pallor, clubbed fingers, polyarthritis involving both knee, proximal interphalangeal joints, right wrist, and both elbow, and reduced range of movement in the same joint. In palpation, liver was palpation 3 cm below the right subcostal margin. In auscultation, bilateral fine crackles heard.

Laboratory tests were performed and revealed microcytic iron deficiency anaemia (Hct: 31%, Hg: 9.7 g/dL, MCV: 70, RDW: 18, Fe: 13 mg/dL), TIBC = 65 (normal 240 - 450 mcg/dL)rheumatoid factor positive, erythrocyte sedimentation rate 106 mm/hour, C-reactive protein (CRP) = 95 mg/L, complement C3 108 mg/dL (normal 90 - 180 mg/dl) and C4 20.7 mg/dL (normal 20 - 60). Ferritin, B12, reticulocyte count, immunoglobulin level, folic acid, and liver functions levels were within normal range.

Peripheral smear showed hypochromic microcyte anemia.

Laboratory tests for extensive workup was performed, including antinuclear (ANA), antineutrophil cytoplasm (ANCA P-C), Anti- DNA (ds) antibody, anti smith, were also negative, Anti-CCP = 150 (normal up to 5 U\L). The tuberculin skin test was negative. The investigation of celiac disease was negative (serum IGA transglutaminase antibodies and endomysial antibody).

Pulmonary function tests (PFTs) were performed and showed that the lung is un compliance.

The radiographic investigation; echocardiography was normal, chest X-ray showed bilateral alveolar interstitial infiltration (Figure 1).

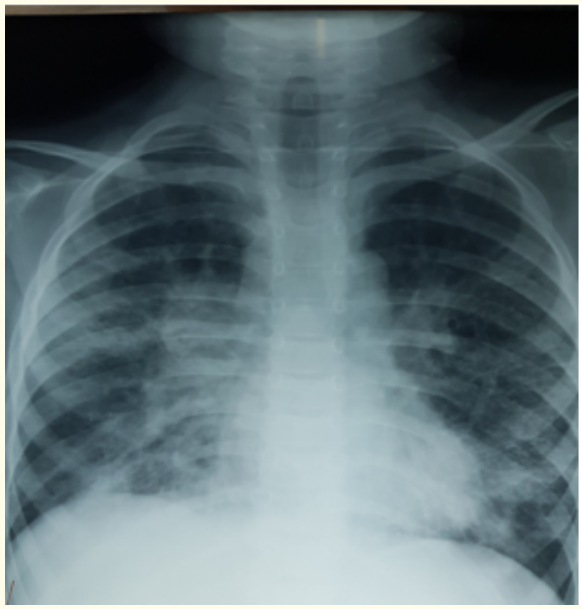

Figure 1

The high-resolution computed tomography (HRCT) showing diffuse bibasilar ground-glass opacities (Figure 2).

Stomach lavage showed a few of hemosiderin macrophage, so an extensive work up was performed and Bronchoalveolar lavage showed numerous hemosiderin-laden macrophage was confirming (IPH) (Figure 3).

The child was treated with prednisolone at a dose of (1 mg/kg/day), MTX (10 mg/m^2/week), with recovery growth, anemia, articular symptoms, joints movement and chest X-ray finding.

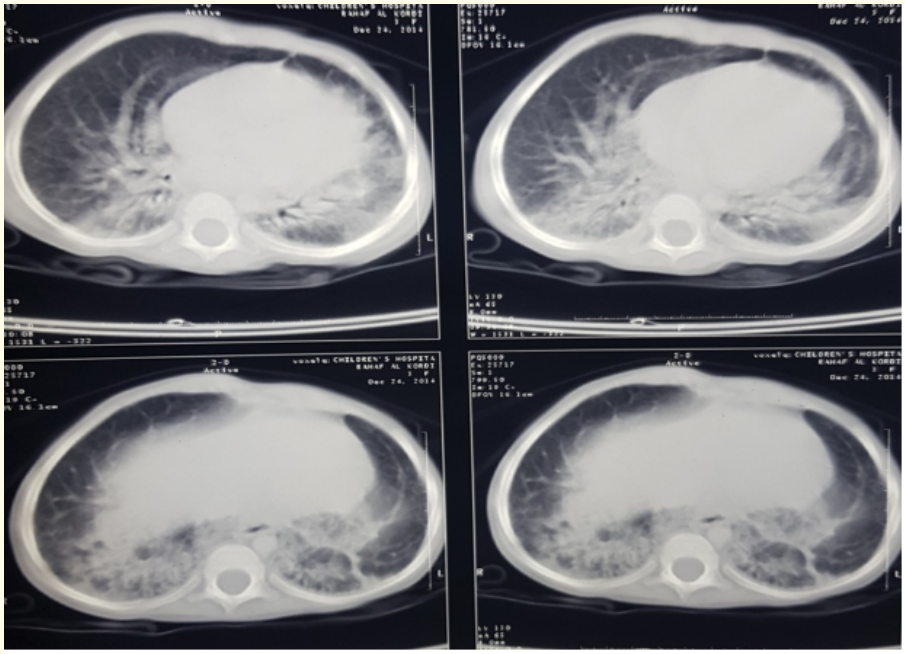

Figure 2

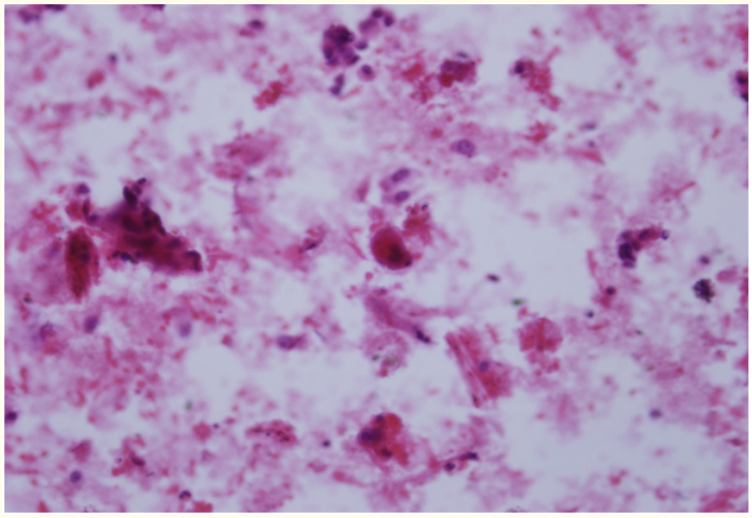

Figure 3

At follow up, within 6 months, the disease has controlled with low dose of steroids. On his last follow up after 1 year of treatment, the child gained weight and height, and improvement the joints movement and the radiography finding CXR and CT.

Discussion

IPH is a rare disorder (the incidence of IPH between 0.24 and 1.26 patient per million children) [2,3].

The trade of hemoptysis, iron deficiency anemia and pulmonary infiltrates on chest imaging suspects diagnosis and the finding of haemosiderin - laden macrophages in bronchoalveolar lavage fluid confirm the disease [4].

JIA is a type of chronic arthritis involving all forms of articulars in the body [7], the presence of arthritis associated with, positive rheumatoid factor and Anti- CCP, highly suggestive of juvenile idiopathic arthritis (RF positive polyarthritis) according to (ILAR Classification). The combination of IPH and JIA has been rarely reported in the literature [1-4], the pathogenetic link between them is not clear, but co-existence of IPH with many autoimmune disease such as celiac disease, systemic lupus erythematosus, and JIA suggest the autoimmunity in etiology of this combination. The previously four report cases [1-4] were classified as polyarticular and oligoarticular arthritis, the association was IPH and JIA same as our case, and to the best of our knowledge, this is the fifth case of IPH and JIA has been reported in the literature and the first one with positive Rheumatoid Factor (RF). Iron deficiency anemia due to both alveolar hemorrhage in IPH and the chronic inflammation in JIA. After confirming diagnosis, we have managed the disease, and she was treated by prednisolone and MTX, however, the same medication give a good results in the previously cases and controlled her pulmonary and joint symptoms.

Conclusion

The association between IPH and JIA is extremely rare, and our case showed that immunological tests should be include in the extensive work up for patients present with IPH. Due to the rarity of this concomitant, diagnosis is usually delayed, but early diagnosis and treatment with corticosteroids is essential for proper management for the patient and improve the outcome (both pulmonary and articular symptom).

Bibliography

1. K Barut., *et al*. "Idiopathic Pulmonary Hemosiderosis In a Child with Recurrent Macrophage Activation Syndrome Secondary to Systemic Juvenile Idiopathic Arthritis". *Case Reports in Pediatrics* (2017): 5693501.
2. A R Wong., *et al*. "Pulmonary Haemosiderosis with Juvenile Idiopathic Arthritis in a Malaysian Child". *Medical Journal of Malaysia* 62.4 (2007): 352-354.
3. R Topaloglu., *et al*. "Pulmonary Hemosiderosis with Juvenile rheumatoid arthritis: a case report". *Turkish Journal of Pediatrics* 42.2 (2000): 148-150.
4. SH Chu., *et al*. "Juvenile idiopathic arthritis with pulmonary hemosiderosis: a case report". *Journal of Microbiology, Immunology and Infection* 35.2 (2002): 133-135.
5. I Bakalli., *et al*. "Idiopathic pulmonary hemosiderosis- a diagnostic challenge". *Italian Journal of Pediatrics* 40 (2014): 35.
6. Kjellman B., *et al*. "Idiopathic pulmonary haemosiderosis in swedish children". *Acta Paediatrica Scandinavica* 73.5 (1984): 584-588.
7. OC Ioachimescu., *et al*. "Idiopathic pulmonary haemosiderosis revisited". *European Respiratory Journal* 24.1 (2004): 162-170.
8. Reading R., *et al*. "Pulmonary haemosiderosis and gluten". *Archives of Disease in Childhood* 62.5 (1987): 513-515.

Volume 8 Issue 11 November 2019