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RECURRENT IRON DEFICIENCY ANEMIA WITH AN UNUSUAL CAUSE

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ABSTRACT

Microscopic polyangiitis presents in unique ways with systemic manifestations. The disease is seen classically in elderly population and has atypical presentation in children and adolescents. We present a case of young adult who presented with recurrent iron deficiency anemia because of microscopic polyangiitis. We present the case for obscure presentation of a rare disease with a common manifestation.

***Key words:** Iron deficiency anemia, microscopic polyangiitis, pulmonary renal syndrome*

INTRODUCTION

Microscopic polyangiitis is a rare disease which presents classically as small vessel vasculitis affecting the arterioles, venules, and capillaries. This is a form of autoimmune disease and shares similar clinical presentation with other pulmonary renal syndromes.[1] Goodpasture's syndrome (GPS), Microscopic polyangiitis (MPA), Wegener's granulomatosis (WG), and Churg-Strauss syndrome (CSS) are the classical pulmonary renal syndromes. They present with hemoptysis, pneumonia, and rapidly progressive renal dysfunction leading to end stage renal disease. Clinical distinction among these diseases is ambiguous, thus relying on the investigations for diagnosis.[2]

Iron deficiency anemia is a common disability in our population due to a wide variety of conditions. The replenishment of iron stores along with correction of the primary etiology lead to significant improvement in hemoglobin without any recurrence of the disease. The common causes for iron deficiency anemia (IDA) are inadequate intake, poor bioavailability, worm infestation, and chronic blood loss.[3] Systemic autoimmune disorders rarely present with iron deficiency anemia and there is only one such report available in the literature of MPA presenting as IDA.[4] We recently encountered a young patient who presented with recurrent iron deficiency anemia with underlying microscopic polyangiitis. We report the case for the unusual presentation of a systemic autoimmune disease.

CASE REPORT

A 23-year-old man came to medical attention for the first time in September 2010 with short febrile illness and easy fatigability. He gave history of occasional hemoptysis without any cough and asthmatic symptoms. He denied history of chronic blood loss and past history of blood transfusions. Clinical examination revealed pallor, no organomegaly and normal vital parameters. Investigations revealed microcytic hypochromic anemia (Hemoglobin 4.1 gm/dL, Mean Corpuscular Volume 66 fl) and no evidence of hemolysis. Other parameters like total iron binding capacity–363 μg/dL (normal 230-400), serum iron 79 μg/dL (normal 80-176) and ferritin 34 ng/mL (normal 30-280) were suggestive of iron deficiency anemia. He had normal upper gastrointestinal endoscopy and bone marrow revealed erythroid hyperplasia with deficient iron stores. Chest X-ray was normal and antinuclear antigen (ANA) was negative along with normal hemoglobin electrophoresis. He was managed with blood transfusion and parenteral iron replacement and hemoglobin (Hb) improved to 13.6 gm/dL. He was discharged with continuous oral iron replacement and was advised regular follow-up.

In February 2011, he reported again with breathlessness, easy fatigability, listlessness, hemoptysis, and polyarthralgia. Examination revealed pallor with normal systemic examination. There was no evidence of arthritis or bony tenderness. Investigations revealed Hb 5.4 gm/dL, microcytic hypochromic anemia and urinalysis showing hematuria with proteinuria. Chest X-ray showed military mottling with a differential diagnosis of military tuberculosis or pulmonary hemorrhage. CT chest showed bilateral extensive ill defined ground-glass haze suggestive of pulmonary hemorrhage [Figure 1]. The presence of microscopic hematuria, nonnephrotic range proteinuria along with pulmonary hemorrhage leads us to pulmonary renal syndrome with a differential diagnosis of MPA, WG, CSS, or GPS.

Further investigations revealed 24-hour urine protein 750 mg, urea 23 mg/dL, creatinine 0.8 mg/dL, and sputum for acid fast bacilli was negative. Immunological tests were negative for anti-glomerular basement membrane (GBM) antibodies, ANA, ds DNA, and antineutrophil cytoplasmic antibody (ANCA) type C. p- ANCA (Perinuclear ANCA)

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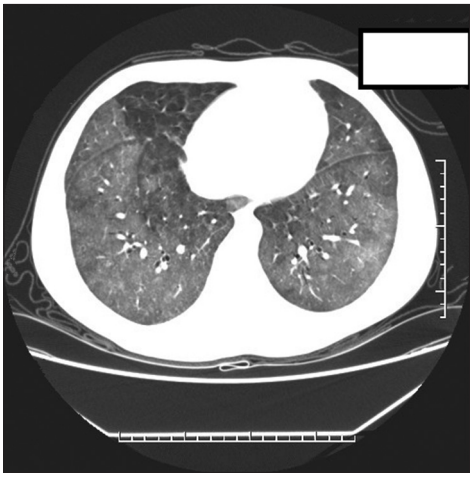

Figure 1: CT chest showing bilateral diffuse ground-glass haze suggestive of pulmonary hemorrhage

was positive by both ELISA and immunofluorescence. Transbronchial lung biopsy showed pulmonary hemosiderosis with diffuse alveolar hemorrhage and renal biopsy showed focal segmental glomerulosclerosis (FSGS). Immunofluorescence of the renal biopsy tissue showed linear, fine, granular deposits of IgG (3+), C1q and C3 (2+) along capillary wall. Ig M and Ig A stains were negative on the renal tissue. Based on the immunological profile a final diagnosis of microscopic polyangiitis was made.

He was treated with intravenous pulse methylprednisolone for 3 days followed by oral prednisolone (1 mg/kg) and Inj Cyclophosphamide (10 mg/kg, 6 cycles at 4 weekly intervals). Further, he was started on tab azathioprine (100 mg/day) with tapering doses of prednisolone (up to 10 mg daily). Immunosuppressive therapy was complicated by steroid-induced glaucoma and diabetes. Clinical symptoms improved and the hemoglobin rose to 14.3 gm/dL with no recurrence of hematuria and proteinuria. He is presently using azathioprine, prednisolone along with Calcium, and vitamin D supplements.

DISCUSSION

Our case is unique in its presentation with iron deficiency anemia in a young adult with apparently good health. Extensive evaluation for etiology did not yield any positive diagnosis and iron replacement resulted in good response in hemoglobin level. The diagnosis was missed at the initial presentation due to occasional hemoptysis with no recurrence and normal chest X-ray ruling out any underlying pathology. Significant improvement was also noted with iron replacement further misleading us about the condition. Repeat presentation with severe anemia coupled with hemoptysis, polyarthralgia, microscopic hematuria, and proteinuria gave a clue to pulmonary renal syndrome as the underlying etiology.

Differentiating between the types of pulmonary renal syndromes is difficult sometimes. GPS is seen in young adults, whereas small vessel vasculitides (WG, MPA, and CSS) are seen commonly in 5th to 7th decade of life.[5] GPS is characterized by rapidly progressive renal failure, hemoptysis, and azotemia. The diagnosis is confirmed by anti GBM antibodies along with kidney biopsy showing linear deposits of IgG and C3 along with IgA and IgM. WG typically has involvement of upper airways and systemic features in addition to pulmonary renal involvement. The diagnosis is confirmed by lung and kidney biopsy and serology shows positive c-ANCA. CSS is characterized by allergic manifestations, atopy and asthma in adult life along with cardiac lesions in majority of patients.

MPA is a rare disease with kidney involvement in 90% and pulmonary disease in 30-40% of cases. Disease profile is similar to that of CSS and WG but the prognosis is better with 10-year survival rate of 70%. The characteristic antibody seen is p-ANCA but our patient had c-ANCA positive and p-ANCA negative serology. The p-ANCA positivity is seen in 75-80% of patients and only 10-15% of patients have c-ANCA positive.[6]

To conclude, we present an interesting case of MPA in a young adult with recurrent severe iron deficiency anemia. Our case highlights the need for keeping pulmonary renal syndromes as a cause of rapidly progressive anemia with no apparent etiology.

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