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Case Report

Multiple Mechanisms Occurring Simultaneously Leading to Severe Anemia -

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ABSTRACT

Pernicious anemia is an autoimmune disease rarely occurring with autoimmune hemolytic anemia. In this article we report the case of a 57 year old woman presented to the internal medicine department with a history of increasing weakness and shortness of breath during 2 weeks. Clinical examination revealed marked pallor, moderate jaundice, fever, tachycardia and enlarged spleen without other lymph nodes. A full blood count showed pancytopenia with a regenerative macrocytic anemia at 3.3 g/ dl. White blood cells were at 3500 elt/ mm^3 and platelets at 27000. Blood biochemistry showed elevated indirect bilirubin, liver cytolysis and high LDH levels. Schizocytes were at 2%. Direct antiglobulin test at 37°C was strongly positive for IgG and C3D. A bone marrow differential count confirmed the diagnosis of megaloblastic anemia due to vitamin B12 deficiency. Intrinsic factors antibodies and parietal cell antibodies were positives. Fever was secondary to urinary infection treated with antibiotics. A diagnosis of warm antibody type auto immune hemolytic anemia complicating pernicious anemia was made. Corticosteroids associated with vitamin B12 were administered with good outcome.

Keywords: Auto immune hemolytic anemia; Pernicious anemia; Schizocytes

INTRODUCTION

Pernicious anemia is an autoimmune atrophic gastritis of the fundus predominantly responsible for a malabsorption of vitamin B12. Despite its association with several autoimmune disorders, few observations have reported an association with Autoimmune Hemolytic Anemia (AIHA). We report a case of this rare association.

CASE REPORT

A 57 year old woman presented to the internal medicine department with a history of increasing weakness and shortness of breath during 2 weeks. Clinical examination revealed marked pallor with moderate jaundice, icteric conjunctivae, fever at 38.5°C, tachycardia and enlarged spleen without other lymph nodes. A full blood count showed pancytopenia with a regenerative macrocytic anemia at 3.3 g/dl and Mean Corpuscular Volume (MCV) = 116.7 fl. Reticulocyte count was 387 740 elements/ mm^3 (Elt/ mm^3) , White Blood Cells (WBC) were at 3500 elt/ mm^3 and platelets at 27000. Blood biochemistry showed renal insufficiency with creatinine clearance at 33.82 ml/min. Bilirubin was raised at 30.4 µmol/ l with elevated indirect bilirubin, liver cytolysis with high levels of ALT and SGPT and serum LDH at 2250 U/ l. Schizocytes were at 2% without other signs of thrombotic thrombocytopenic purpura. Direct Antiglobulin Test (DAT) at 37°C was strongly positive for IgG and C3D. A bone marrow differential count confirmed the diagnosis of megaloblastic anemia. Vitamin B12 level was too low (20 ng/ l). Intrinsic factors antibodies and parietal cell antibodies were positives. Fever was secondary to urinary infection treated with antibiotics. A diagnosis of warm antibody type auto immune hemolytic anemia associated with pernicious anemia was made. Three pulses of methylprednisolone (1g/ day) were administered than oral prednisone (1 mg/ kg/ day) associated with vitamin B12, 1000 gamma/ day for 7 days then 1000 gamma each other day and then monthly with good outcome and improvement of the renal function with rehydration.

DISCUSSION

Pernicious Anemia (PA) is an autoimmune atrophic gastritis leading to vitamin B12 deficiency due to its malabsorption. It's one of cobalamin deficiency causes [1]. Its prevalence is 0.1% in general population reaching 1.9% over the age of 60 [2]. Clinical manifestations of PA consist in anemia with accompanying functional manifestations, neuropsychiatric signs and gastrointestinal disorders. Hematological manifestations are mainly represented by the asynchrony of nucleocytoplasmic maturation with an immature nuclei and an acidophilic cytoplasm leading to hemolysis by ineffective intra medullary erythropoiesis. Associated to that, moderate thrombopenia and leucopenia could be seen with hypersegmented neutrophils in the blood smear. Because of this ineffective intra medullary erythropoiesis, other marks of hemolysis could be seen like elevated LDH levels and low haptoglobin. Other hematological manifestations might be life threatening such as pseudo thrombotic micro-angiopathies with severe hemolytic anemia, severe thrombopenia and schizocytosis such is the case of our patient. This entity was revealing the diagnosis of vitamin B12 deficiency in 2.5% of the cohort of Federici, et al. [3]. This is explained by cytoskeletal fragility which is responsible of erythroblast fragmentation in dyserythropoiesis [4]. Anti-intrinsic factor (anti-IF) antibodies and antigastric parietal cell (anti-GPC) antibodies are the hallmark of autoimmunity of PA. Given that, PA could be associated with other autoimmune diseases in nearly 30% of patients [2]. It can precede the disease or occur after its onset. The most common are Type 1 diabetes, Hashimoto's thyroiditis and vitiligo [5]. Other associations have been described like Sjogren's syndrome, coeliac disease and Addison's adrenal insufficiency [6]. Its association with AIHA, like our patient, is rarely described in the literature. Whether the AIHA occurred simultaneously to PA or after its onset, is not clear and urinary infection in this context could be the triggering agent.

CONCLUSION

Regenerative character of a very macrocytic anemia should raise attention toward associated hemolytic anemia to a megaloblastic one. DAT should be done to confirm the diagnosis of AIHA. Schizocytes could be seen in vitamin B12 deficiency resulting from erythroblast fragmentation in dyserythropoiesis.

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