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Massive Splenomegaly, Pancytopenia and Leucoerythroblastosis as Presentation of Megaloblastic Anemia

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Abstract:

Megaloblastic anaemia is commonly a disease of nutritional deficiency. Its usual presentations are fatigue, paleness and recurrent infections. Megaloblastic anemia rarely presents with splenomegaly and moreover there is only mild to moderate splenomegaly. There are few cases reports of megaloblastic anaemia with splenomegaly as a presenting feature. We report an unusual presentation with massive splenomegaly and leuco-erythroblastosis in a child with megaloblastic anemia as cause.

Key words: Megaloblastic Anaemia, Splenomegaly, Anemia, Pallor, Hematologic Diseases, Humans.

Introduction

Megaloblastic anemia is uncommon condition in childhood generally seen in nutritional deficiency or gastrointestinal disease [1]. Many symptomless patients are detected through the raised mean corpuscular volume (MCV) on a routine blood count. The main presenting feature in more severe cases are those of anemia. Anorexia is usually marked and there may be lethargy, weight loss, diarrhea, or constipation. Glossitis, angular cheilosis, fever, jaundice (unconjugated), and reversible melanin skin hyperpigmentation may also occur with deficiency of either folate or cobalamin. Thrombocytopenia sometimes leads to bruising, and this may be aggravated by vitamin C deficiency or malnutrition. The anemia and low leukocyte count may predispose to infections, particularly of the respiratory or urinary tracts.

Megaloblastic anemia rarely presents with splenomegaly and moreover there is only mild to moderate splenomegaly. We report a child with megaloblastic anemia who presented with massive splenomegaly and leuco-erythroblastosis.

Case Report

A 14 years old child was admitted to our hospital with history of on and off fever from last 3-4 years along with abdominal mass, increasing fatigability and increasing paleness from last 1 year. Abdominal mass gradually progressed to present size. No history of bleeding manifestations, blood transfusion, jaundice, bony or joint pain was noted.

On physical examination child was having severe

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pallor, grade 2 clubbing, with no evidence of icterus or significant lymphadenopathy. Splenic enlargement (19cm) below the left costal margin with no apparent fluid in abdominal cavity was present. The rest of her systems' examination was normal.

Investigations revealed child had severe anaemia with pancytopenia. TLC-1500/mm^3 with neutrophil: 37.5%, lymphocyte: 53%, monocyte: 9.5%, platelet count: 91000/mm^3, MCV: 117.8 fl, MCH: 38 pg, MCHC: 32.0%. Liver function test showed total serum bilirubin 0.78 mg/dL, indirect bilirubin 0.5 mg/dL, alanine aminotransferase 34 IU/L, aspartate aminotransferase 21 IU/L, alkaline phosphatase 31 IU/L, total serum protein 5.5 gm/dL and serum albumin 3.3 gm/dL. Peripheral smear was suggestive of macrocytic anaemia with pancytopenia and no evidence of malarial parasite for consecutive 3 days. Ultrasound abdomen was suggestive of hepatosplenomegaly with spleen of 19 cm with no thrombus and normal portal colour flow and spectral wave form with normal velocity on colour Doppler. On bone marrow aspiration erythroid hyperplasia with erythroid precursors showing distinctive maturation arrest suggestive of megaloblastic anemia was reported. Serum vitamin B12: 115 pg/mL (200-800 pg/mL) was low.

Although there are previous rare reported cases of megaloblastic anaemia with splenomegaly but such massive enlargement of spleen is rare as in our case.

Discussion

Cobalamin deficiency is manifested clinically through its effect on rapidly proliferating tissue, particularly the bone marrow, lining of the intestinal tracts and nervous system [2]. This can give rise to three clinical pictures in which megaloblastic anemia, gastrointestinal symptoms, or neurologic degeneration predominates. Among the gastrointestinal symptoms frequently seen are loss of appetite with weight loss, nausea, constipation, occasional diarrhoea, and soreness of the tongue (glossitis) or 'cankers' of the tongue. Commonly, macrocytic anemia occurs and is often accompanied by neutropenia and thrombocytopenia. The MCV is 120 fl or greater, unless the increase is balanced by a decrease due to coexisting iron deficiency. The blood smear contains oval macrocytes and multilobar neutrophils. The bone marrow is usually megaloblastic. The concept of hypersplenism was proposed in 1907, and criteria for the diagnosis includes: splenomegaly, compensatory bone marrow hyperplasia, cytopenia and improvement or resolution in these findings after splenectomy [3].

In adults 13% cases of megaloblastic anemia are associated with splenomegaly [4]. A study from north-west India by Sarode R *et al*. have found 34% of megaloblastic anemia are associated with mild splenomegaly [5]. Few studies have shown association of pernicious anemia with giant splenomegaly [6]. A study by Khan FS *et al*. demonstrated the different aetiologies of pancytopenia based on bone marrow examination and found 13.2% cases of megaloblastic anemia as aetiology and 37.2% splenomegaly as clinical association [7].

In above mentioned case, child was having all symptoms of chronic hemolytic anemia with pancytopenia (hypersplenism) with no history of frequent blood transfusions, peripheral smear was not suggesting any evidence of hemolysis rather showing macrocytes predominantly. Splenomegaly was not associated with any lymphadenopathy. Ultrasound has ruled out portal hypertension as the cause of splenomegaly. Dilemma for considering him as a megaloblastic anemia was due to presentation of massive splenomegaly with anemia. Further bone marrow aspiration was done, which revealed megaloblastic anemia with leuco-erythroblastosis.

Conclusion

Splenomegaly with pancytopenia is an unusual association of megaloblastic anemia as most of the literature suggest only severe anemia as predominant symptom of megaloblastic anemia. We report an atypical clinical observation of megaloblastic anemia, which is especially interesting in that it illustrates the difficulty in approaching a case of massive splenomegaly.

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