Enteric Fever with Severe Pancytopenia in a Four Year Girl

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ABSTRACT

Typhoid fever and paratyphoid fever (also known as enteric fever) are severe systemic illnesses caused by salmonella typhi and S. paratyphi respectively. Enteric fever is prevalent in developing countries including Nepal, where it still remains as a major health problem. There have been reports of pancytopenia with enteric fever which has been attributed to mechanisms like bone marrow suppression, infection associated hemophagocytic syndrome and disseminated intravascular coagulation. We report here a case of severe pancytopenia in enteric fever as a result of bone marrow suppression due to systemic infection.

Keywords: *enteric fever, pancytopenia, bone marrow suppression*

INTRODUCTION

Typhoid fever and paratyphoid fever also known as enteric fever are severe systemic illnesses characterized by sustained fever and abdominal symptoms caused by Salmonella typhi and S. paratyphi respectively.[1] Salmonella infection occurs when organisms ingested via contaminated food or water bypass the gastric defenses and multiply within the submucosal Peyer's patches of the small intestine. Dissemination of S. typhi from the Peyer's patches to the reticuloendothelial system subsequently occurs via the lymphatic system and bloodstream to the liver, spleen and bone marrow and where the organisms reside within monocyte-derived or tissue macrophages. There have been reports of pancytopenia with enteric fever which has been attributed to bone marrow suppression, infection associated hemophagocytic syndrome, disseminated intravascular coagulation.[2-5] However, such cases of severe pancytopenia in enteric fever has never been reported from Nepal.

CASE REPORT

A four year old girl was brought to the pediatrics department of Nepal Medical Collage Teaching Hospital with 10 days history of fever, headache, fatigue and decreased appetite. She had no complaints of diarrhea or vomiting. Her past and family history was uneventful.

On physical examination, she was irritable, ill looking. Her weight and height were 11 kg and 85cm respectively (weight for age and height for age both were below 3rd percentile). Axillary temperature was 99 °F, pulse rate was 110 per minute, regular, respiratory rate was 24 per minute and blood pressure was 90/60 mmHg. The liver was palpable two fingers below the right costal margin which was non tender. Spleen was palpable one finger below the left costal margin. Rest of the physical examination was unremarkable.

Laboratory evaluation on admission revealed a WBC count of 4700/cumm with N 68%, L 26% and E 6%,

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Hemoglobin 10.1 g/dl, platelet count 70000/cumm, ESR 42mm. Her peripheral blood smear showed normochromic, normocytic RBCs with few hypochromic, microcytic RBCs and reduced platelet on smear.

Widal test showed agglutination titre of 1:320 for S. typhi O and S. typhi H antigen. The reports of stool analysis, urinalysis, chest radiography, serum electrolytes, creatinine and BUN were normal. On blood culture, S. typhi was isolated which was reported sensitive to ofloxacin and ceftriaxome. According to her blood culture antibiogram results, ceftriaxone was started which was continued for 12 days.

USG abdomen revealed mild hepatosplenomegaly and mesenteric lymphadenopathy.

Her serial platelet counts showed a decreasing trend reaching lowest levels of 17 ooo/cumm. There was similar decreasing trend of her total WBC count and hematocrit levels. Patient also developed petechiae over limbs, trunk and mucosal surfaces, and hematuria. Patient was evaluated for pancytopenia. Coagulation tests PT and aPTT were done to rule out DIC which were within normal limits. Bone marrow aspiration was carried out to rule out aplastic anemia and leukemia which showed mild hypocellularity with predominance of myeloid cells and suppressed erythroid precursors and increases number of plasma cells without hematophagocytosis. Patient received two units of whole blood and four units of platelet rich concentrate over the next few days. Her followup CBC showed platelet count, WBC count and hematocrit within the normal range. Patient showed gradual improvement with intravenous antibiotic and supportive management. At the end of 14 days, she was in good condition without any other complaints and displayed normal hematologic features.

Two months after the discharge, the patient is doing well.

DISCUSSION

Enteric fever remains a serious public health problem in developing countries like Nepal despite the improvements in living standards.[6,7] Classic reports describe characteristic stages of typhoid fever in untreated individuals :First week of illness — rising ("step ladder") fever and bacteremia; Second week - abdominal pain and rash (rose spots); Third week — hepatosplenomegaly, intestinal bleeding and perforation, may occur with secondary bacteremia and peritonitis[8]. The symptoms, signs, and complications of typhoid fever vary widely in different series, and may be related to age, geographic area, the causative organism, or the time at which patients seek medical care.[9-12] The final diagnosis is based on serology and organism cultures from appropriate sites. Blood culture is considered as the gold standard for the diagnosis.

Our patient had fever, mild hepatosplenomegaly and pancytopenia on admission. It has been previously reported that enteric fever usually produces discernible leucopenia and neutropenia early in the course of the illness which is often associated with thrombocytopenia and anemia.[13-15]

Our patient however developed severe pancytopenia during the course of hospital stay due to which further investigations were done. Her bone marrow aspiration smear revealed mild hypocellularity without hematophagocytosis. In patients with salmonellosis, pancytopenia is mostly due to histiocytic hyperplasia in bone marrow with marked phagocytosis of platelets, leucocytes and red blood cells which is also called IAHS (infection associated hemophagocytosis syndrome).[4] The occurrence of DIC which is rare in enteric fever can also be the cause of pancytopenia.[5] We were able to excluded DIC in our case due to normal PT and aPTT. However, awareness of this potential complication and early diagnosis and treatment may be life saving. Thus after excluding other possible causes, suppression of bone marrow cells due to systemic infection was the probable reason for pancytopenia in our case.

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