Rare coexisting diagnoses of Ulcerative colitis in a child with sickle cell disease: A case report.

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Abstract— Sickle cell anaemia is a disease is characterized by sickle shapes of its red blood cells, attracting RBCs to each other, and polymerization when in a deoxygenated state.Pain is the hallmark of this disease, and it is always the major issue of the patients and their caregivers. Abdominal pain is the most irritating form of sickle cell pain. The abdominal vaso-occlusive crisis might be clinically difficult to distinguish from acute abdominal pain from other causes and can be a diagnostic dilemma. We report an ulcerative colitis case in a patient with SCA which is a rare coexistence.

Index Terms— SCD, Ulcerative colitis, AL-Ahsa, Saudi Arabia, Sickle cell, Anemia.

1 INTRODUCTION

Sickle cell anemia is an autosomal recessive hemoglobinopathy leading to the synthesis of abnormal β globin chains resulting in the production of HbS instead of HbA.(1) This disease is characterized by sickle shapes of its red blood cells, attracting RBCs to each other, and polymerization when in a deoxygenated state. Hemoglobin polymerization, leading to rigid RBCs which cause profound multi-organ effects by occlusion of blood vessels, decreasing blood flow to these organs, and affecting the immunity.(2) Pain is the hallmark of this disease, and it is always the major issue of the patients and their caregivers. Abdominal pain is the most irritating form of sickle cell pain. The abdominal vaso-occlusive crisis might be clinically difficult to distinguish from acute abdominal pain from other causes and can be a diagnostic dilemma.(3) We report a case of the concurrent diagnoses of sickle cell anemia and ulcerative colitis. Coexisting presentation of ulcerative colitis and sickle cell disease is a highly rare incident.(4)

2 CASE REPORT

A 2-year-old white boy from Al-Ahsa, Saudi Arabia, a known case of sickle cell anemia was admitted to children and maternity hospital with the chief complaint of high fever reaching up to 40 degrees not controlled by antipyretics. After a week of the admission, his mother noticed that he has diarrhea mixed with blood and mucous, started suddenly in an amount of ten times per day. It is occasionally disturbed his sleep at night. This diarrhea is associated with mild intermittent periumbilical colicky abdominal pain for the same duration, lethargy, and night sweet. He also has a history of tenesmus and urgency. His mother denied any history of loss of appetite, weight loss, nausea, vomiting, abdominal distention, jaundice, purities, joint pain or swelling, skin rash, mouth ulcer, exposure to animals. His medical records are clear from any chronic disease (DM –HTN –asthma). The patient has a history of frequent admissions in the past due to the vaso-occlusive crisis; therefore, he received many blood transfusions. His quality of life is markedly decreased. He is the first child for sickle cell trait healthy non-consanguineous parents. He was born at full term, delivered with normal spontaneous vaginal delivery with no obstacles. He has two brothers and one sister, the younger brother has sickle cell disease, and the others are well with good health. His grandfather is diagnosed with ulcerative colitis. On physical examination, he looked well and alerted. He is not pale, jaundiced or cyanosed. His weight was 9.8 kg, and height was 85 cm, both below the third percentile for his age. His vital signs were within normal ranges. Normal skin examination, and no lymphadenopathy. He has mild abdominal tenderness (periumbilical) and mild splenomegaly. The rest of physical examination was unremarkable. His laboratory workup showed hemoglobin 7.1 g/dl, RDW 22.5 %, with a reticulocyte count of 13.43 %, platelets 296 × 109 /L. His total bilirubin was 1.75 µmol/L. The direct bilirubin was 0.4 µmol/L, C-reactive protein (CRP) 34.3 mg/L, IgG1 10.100 g/L. The occult blood in stool was positive. ANA was slightly positive, and his erythrocyte sedimentation rate (ESR) was 55 mm. The white blood cell count, liver and renal function test, stool analysis, abdominal ultrasonography, electrolyte, uric acid, serum immunoglobulins (IgG, IgA, and IgM) all were within normal. Hepatitis (A, B, C), ANCA, ASCA, anti-smooth muscle antibodies, liver kidney microsomal antibodies, anti-mitochondrial antibodies, clostridium, anti-endomysial antibodies test, adenovirus, d Rotavirus, H pylori were negative. The patient underwent upper endoscopic and colonoscopic examinations. The upper endoscopic examinations were normal. Colonoscopy showed inflamed mucosa of the entire colon with a lack of vascularity, edema, pallor and distorted cecum. A biopsy was taken from ascending and transverse colon, Both showed some architectural distortion by moderate to the remarkable increase of lamina propria, in addition to neutrophilic infiltration of the lamina propria and crypt epithelium. Crypt abscess formation is noted focally in both specimens. Foci of goblet cell depletion are noted. The overall findings were consistent with active inflammation with features consistent with ulcerative colitis. The patient was treated as an ulcerative colitis case. Initially, the patient received oral mesalazine (1.5g daily), oral corticosteroid (prednisolone 40mg daily) was added later on due to poor response. He started to improve and show signs of remission. He did great after that with neither diarrhea nor abdominal pain and began to have a better appetite. Prednisolone dose was tapered till stopped. As a maintenance therapy, He was continued with the same dose of sulfasalazine. He was attending the clinic regularly with no complaints up to the present.

3 Discussion

SCA has clinical manifestations in various organs which are considered to be an outcome of structural changes in the hemoglobin molecule. The Gastrointestinal system can be affected, although it is rare.(5) Abdominal pain is one of the manifestations of gastrointestinal affection in SCA. It is usually a consequent of vaso-occlusion, and it is maybe a single complaint or presented with generalized pain crisis in other parts of the body. (6)

Our patient diagnosed with ulcerative colitis on the basis of clinical, radiological, and histopathological results. Although the clinical presentation of our patient has many differential diagnoses, including ischemic and infectious colitis. After multiple cultures and examination of stool, blood in our patient, no particular organism could be isolated.

Ischemic colitis had been reported in the literature which accentuates the liability of SCA patient to vaso-occlusion and ischemia. Yet Ischemic colitis is a rare occurrence in SCA patients, not more than eight cases have reported in literature up to the present. (7) This is likely owing to the fact that the colon has a rich collateral blood vessels and low oxygen needs. The colon can bear up to a two-third reduction of the blood supply for half a day with no ischemic changes. (8) Classic characteristics of ischemic colitis include ischemic changes in mucosal, mural or transmural that can be detected by colonic biopsies, and a presentation with acute abdominal pain and bleeding. Our particular patient presented with chronic diarrhea which is not a feature of ischemic colitis. Also, the endoscopic examination showed an inflammation that involves the entire colon, which is not in favor with ischemic colitis. The best explanation of all these features goes toward ulcerative colitis. The competition now is what is the best investigation to detect the ulcerative colitis in CSD patients. Usually, in a similar condition, we use CBC, LFT, inflammatory markers, the antibody test, barium meal and enema, colonoscopy, chromoendoscopy, CT scan and stool analysis. A new study suggests other tools for determination of illness such as monocytosis biomarkers and low lymphocyte/monocyte ration. This tool has low cost and very useful to identify active ulcerative colitis patient from other patients with C. difficile or without IBD. (7) Improper diagnosis can lead to inexpedient treatment and can raise complications; also it will affect child growth and development. Nonetheless, since this coexistence of CSD and ulcerative colitis is rare and the complexity of the CSD (4), we encourage other researchers to do further studies on this rare coexistence to explain more about their relationship.

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