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Visceral Leishmaniasis with Secondary Hemophagocytosis in a Sickle Cell Anemia Patient

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Abstract Sickle cell anemia (SCA) is one of the most common hemoglobinopathies, which is characterized by a high level of abnormal hemoglobin called hemoglobin S (Hb S). This abnormal hemoglobin results from changes in amino acids and valine instead of glutamine at the sixth position of the β globin molecule. Two affected genes are needed to develop sickle cell anemia, homozygous Hb S.

Keywords: *sickle cell anemia*

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1. Introduction

Patients with SCA usually suffer from many clinical symptoms, mainly vaso-occlusive crisis, hemolytic crisis, sequestration crisis, acute chest syndrome, and cerebral vascular accident. Rare sequels like leg ulcer, priapism in male patients, sickling nephropathy and retinopathy can also happen.

Patients with sickle cell anemia are prone to bacterial infections (such as streptococcus pneumonia and hemophilusinfluenzae) and special viral infections such as parvovirus B19.

Leishmaniasis is a parasitic infection caused by an obligate intracellular Protozoa Leishmania. The transmission of the Leishmania parasite is through the bite of female sandflies of the genera Phlebotomus and Lutzomyia. Infection happens when sandfly feeds on an infected host's blood. So, amasigote will pass into sandfly gut where it will transform into infective form, promastigote, which will then travel to mouthparts waiting for the next bite. When sandfly feeds on the blood of its next victims, promastigotes exploit the chance to pass into the skin or blood and transform again into amastigotes, which resides and multiply in phagolysosomes.

Leishmaniasis is classified into three clinical diseases: (1) a local cutaneous form, typically self- healing and caused by Leishmania tropica, Leishmania major, or Leishmania mexicana; (2) mucocutaneous disease with progressive destruction of the nasal, oral, or pharyngeal mucous membranes, caused by Leishmania braziliensis and L. aethiopica; and (3) a visceral form, in which the parasite spreads systemically, caused by Leishmania donovani and Leishmania chagasi.

In 2012, Daniel Garcerant et al described a case of cutaneous leishmaniasis in a patient with homozygous for sickle cell disease who developed some adverse effects after starting pentavalent antimony.

Both sickle cell disease and visceral leishmaniasis (VL) are common in Africa. In Africa, more than 200,000 infants are born yearly with SCD [2].

Many parasitic infestations are reported in patients with SCD. Professor Joseph O.A. et al from Nigeria published, on April 1997, a study about parasitic infections in patients with sickle cell crisis, which 150 patients had been involved in [1]. A total of 102 parasitic infections associated with clinical cases of sickle cell crisis were recorded (malaria, 36 [35.3%]; helminths, 49 [48%]; and malaria and helminths together, 17 [16.7%]). Of the 49 helminthic infections, 26 (53.1 %) were due to Ascaris lumbricoides, 15 (30.6%) were due to hookworms, 7 (14.3%) were due to Trichuris trichiura, and 1 (2%) was due to Strongyloides stercoralis. The mean hemoglobin levels during clinical crises were 7.1 g/dL for helminths, 6.4 g/dL for malaria, and 6.1 g/dL for malaria and helminths together. No case of visceral leishmania had been discovered, although SCA and visceral leishmania are quite common in Nigeria.

Up to our information, no case of sickle cell anemia with visceral leishmania had been reported.

2. The Case

Our case is that of a 2 year-old boy. He was a known patient of sickle cell anemia, which was diagnosed by hemoglobin electrophoresis. He was presented to the emergency department in the referring hospital with a history of intermittent fever for the previous two weeks. No history of upper or lower respiratory infections or gastroenteritis symptoms. Because of the anemia and the increased spleen size, patient was treated as spleenic sequestration crisis with packed RBC transfusion. The

doctor discharged the patient when the fever subsided for two days without any further trace of infection after unremarkable sepsis work up.

Two days later, the patient was again presented to the hospital with fever with no clear focus. Therefore, the patient was referred to our hospital for further diagnosis.

As at the time of admission to our hospital, the patient was irritable but conscious. His temperature was 38.8 degrees Centigrade with tachycardia and normal respiratory rate and blood pressure. He had normal chest and cardiovascular examination. The liver and spleen were palpated 2 cm and 6 cm below costal margins, respectively. The hepatospleenomegally abdominal examination was unremarkable. The rest of systemic examination was unremarkable as well.

During admission, white blood cells count dropped to 2.3 ×10^9L, red blood cells count to 2.77×10^9L, and platelets counts to 93×10^9L. Hemoglobin reached 5.9 g/dl. Liver enzymes, serum electrolyte, urea and creatinin all were normal. Triglyceride, serum ferritin and triglycerides levels were 6.83 mmol/l, 4264 and 6.83, respectively.

Urine and blood cultures, brucella titer, parvovirus, malaria, HIV, CMV and EBV studies were all negative. Patient received packed RBC more than once.

Peripheral blood film showed red cells with normocytic normochromic with poikilocytosis. WBC appeared normal with lymphoid predominance and platelets appearing normal in morphology. No abnormal cells were seen.

Because of prolonged fever, hepatospleenomegaly, pancytopenia and no obvious focus for infection, bone marrow aspiration (BMA) were done and showed scanty particles with normal findings. Infectious diseases team was then consulted and they suggested a repeat of BMA. Repeated BMA showed hypercellular marrow with erythroid hyperplasia /dysplasia and moderate extracellular infestation of Leishmania donovanii with secondary hemophagocytosis.

Liposomal amphotericin B started as 3 mg/kg for 5 days, then on day 14 and 21.

After starting liposomal amphotericin B, fever subsided gradually within five days. The patient improved clinically with a noticeable improvement in his mood and appetite. However, hepatomegaly and spleenomegaly size did not regress till the time of discharge, which was seven days after the commencement of treatment administration. Before discharge, WBC, hemoglobin and platelets counts increased to 5.77 ×10^9L, 8.1 g/dl, and 212 ×10^9L, respectively.

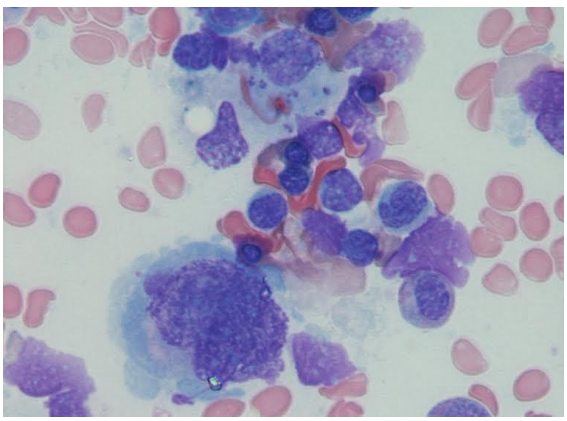

Figure 1. Bone marrow aspiration of the patient. It showed hemophagocytic cells with multiple Leishmania donovanii bodies

Over the frequent visits to the outpatient clinic, patient was stable clinically with normal vital signs. Liver and spleen sizes regressed gradually. However, the liver was just palpable and spleen was around three centimeters below coastal margin, which may be related to his primary disease of SCA. CBCs were normal in spite of accepted low hemoglobin levels in SCA patients. ESR became normal and triglycerides level normalized as well. Serum ferritin dropped gradually to 786, after seven months of illness.

3. Discussion

Both sickle cell anemia and visceral leishmania (VL) are common diseases in southwest area of Saudi Arabia.

In a community-based survey, Al-Qurashi MM et al gave a prevalence of sickle cell diseases in Saudi children and adolescents. Sickle cell disease was detected in 108 of 45,682 children and adolescents with a prevalence of 24 per 10,000. The regional distribution of sickle cell disease showed eastern region dominance with a prevalence of 145 per 10,000, followed by the southern region with a prevalence of 24 per 10,000, the western region with 12 per 10,000, and the central region with 6 per 10,000.

Both SCA and VL could present with fever, pallor, spleenomegally and pancytopenia. This makes clinical differentiation between SCA and visceral leishmania difficult. So, considering VL in SCA patients, who are presented with fever, pancytopenia, spleenomegally with or without hepatomegaly is reasonable and quite important, especially in areas where SCA and VL are common. Other related differential diagnosis, such as epstein barr virus, CMV, parvovirus, brucella infections and malignancy, should be taken into consideration as well.

Secondary hemophagocytosis complicated VL was present in our patient. It is diagnosed by a fever more than 38.8 C, spleenomegally, high ferritin, and high triglycerides, in addition to histological evidence by BM aspiration. After treatment, all indices of secondary hemophagocytosis improved except ferritin level, which dropped from 4264 to 786, which is expected in SCA patients, because of frequent blood transfusions.

This case and maybe the next reported cases may give clearer figures about how VL will manifest in SCA patients.

The recommended treatment with liposomal amphotericin B [4,5] for VL (with dose 3 mg/kg/dose once for three consecutive days, then on day 14 & 21) looks very effective to treat VL in SCA as noticed in our patient.

We recommend physicians following patients with SCA to consider VL, if they have fever and splenomegaly in areas endemic with VL, especially if the fever persisted with no clear focus.

Moreover, We advise them to treat the confirmed VL cases with liposomal amphotericin B.

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