



Case Study

Visceral Leishmaniasis and Chronic Granulomatous Disease in an Infant

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ABSTRACT

Objective of the study is to report a case of an infant with disseminated *Leishmania donovani* (LD) and Chronic Granulomatous Disease (CGD). A report of the clinical and laboratory data on a six months old infant who presented with multiple skin lesions and fever since second week of life. The infant had indolent fever since the second week of life associated with skin lesions on the face, neck and the limbs. He came from an area where both visceral and cutaneous leishmaniasis is endemic. Skin biopsy and the bone marrow aspirate showed *Leishmania donovani* bodies and the culture revealed *Staphylococcus aureus* and *Serratia marcescens* on two different occasions. His immune work up confirmed CGD and living related bone marrow transplantation was successful but complicated with cerebro vascular accident. Although few case reports had been reported regarding this subject but up to my knowledge this is the first case to be reported in infant with CGD and disseminated VL. As the prognosis of CGD is poor, with high morbidity and mortality and infantile leishmaniasis also adds on high rate of morbidity and mortality if not treated early. Establishing an early diagnosis has important practical implications in the successful treatment of these patients. The description of this case and a brief review of the current literature are provided to familiarize physicians mainly in the endemic areas with the relatively rare presentations of these two conditions together.

Keywords

Chronic Granulomatous Disease, Infant, Leishmaniasis

Introduction

Visceral leishmaniasis (VL) is an endemic disease in some part of Saudi Arabia and chronic granulomatous disease (CGD) has been reported (Finocchi *et al.*, 2008; Mauri Pont *et al.*, 1988).

Case report

The patient was a six months old infant born

in Al Hasa, an eastern region of the Kingdom of Saudi Arabia (KSA) for consanguineous parents after an uneventful pregnancy and delivery. As per the national immunization protocol in KSA, he received the Bacillus Calmette–Guérin vaccine (BCG) and Hepatitis B vaccines after birth.

He started during the second week of life to have indolent fever reaching 40°C,

associated with skin lesions that were mainly on the face, and later on the neck and the limbs. At the local hospital, the patient was treated with antibiotics without response. Because he was from an endemic area of VL, he was treated with an antimonial drug for four weeks based on bone marrow results with partial response and later on the lesions started oozing pus. He was brought by his parents to our hospital where he was admitted. The mother had a chronic draining abscess in the groin area for many years and had been subjected to multiple surgeries.

On examination, the infant was alert, active and well hydrated but pale with disfiguring lesions on the nose, the cheeks, neck and the feet with pus (Figure 1, 2 and 3). The abdominal examination revealed hepatosplenomegaly.

The lesions were oozing pus, which was cultured and grew methicillin susceptible *Staphylococcus aureus* (MSSA) and *Serratia marcescens* on two occasions and was treated with the appropriate antibiotics with good response.

The complete blood count showed white blood cells of 26,000 per μl and a neutrophil count of 3800 per μl , Hemoglobin concentration was 8 g/dl and the platelet count was 250000 per μl . Erythrocyte Sedimentation Rate was 100 mm/h, with normal Liver Function Tests, serum Immunoglobulins and lymphocyte markers. Computed tomography (CT) of the ankle and the feet revealed no bony involvement. Bone marrow aspirate not repeated because of the one done at the local hospital and showed *Leishmania donovani* bodies.

Skin biopsy revealed a granulomatoid inflammation with many histiocytes and rare cellular inclusions consistent with

leishmaniasis. The oxidative burst test was zero confirming CGD. He was treated for leishmaniasis with 5 mg/kg of intravenous liposomal amphotericin B as per protocol and later received living related donor bone marrow transplantation (BMT), which engrafted very well. He had a cerebrovascular accident that caused hemiparesis. He was seen in the clinic for two years post BMT, with normal blood counts but residual neurological deficits.

Discussion

The incidence of the CGD is one in 200,000 live birth, usually present with severe and recurrent bacterial and fungal infection (Soler-Palacín *et al.*, 2007). The defect is in the nicotinamide adenine dinucleotide phosphate-oxidase (NADPH oxidase) and absence of microbicidal oxidants.

It can be X-linked (66%) or autosomal recessive (33%). The common infectious agents are *Staphylococcus aureus*, *Aspergillus*, *Nocardia*, *Pseudomonas* and *Serratia* but in 55 %, no organism can be identified.

The Leishmaniasis is a protozoal disease that caused by *Leishmania* parasites, which is transmitted by the female sandfly and it is of three types; cutaneous leishmaniasis, muco-cutaneous and the visceral (Kala-azar).

Its severity depends on many factors mainly: the immune response of the host, virulence of the infecting species and the parasite burden. It is well known that children are at greater risk for severe infection. It is usually manifested with intermittent fever, paleness, anorexia, weight loss, abdominal distension, splenomegaly, hepatomegaly, lymphadenopathy, pancytopenia and hypergammaglobulinemia.

Figure.1 Showed the lesions on the face and the thumb of the patient



Figure.2 showed the lesions on the left side of the neck



Figure.3 Showed the lesions on the right foot



It can be associated with hemophagocytic disease (Martín *et al.*, 2009), and can lead to death. It modulates the immune response, also inhibit the macrophage functions and inhibit the macrophage functions (Olivier *et al.*, 2005; Passwellet *al.*, 1994). The killing of *Leishmania* parasite by human mononuclear phagocytes is oxygen-independent and therefore not affected in the CGD patient (Murray and Cartelli, 1983).

I'm not aware of previous reports linking the CGD to VL in a young infant. But in this case as the mother was diagnosed to be a CGD carrier, and the dissemination of the parasite which is unusual for the *Leishmania donovani* parasite to cause cutaneous

infection may add evidence to what have been published in the literature that CGD patients are can undergo to this fatal infections if not treated early and promptly.

To my knowledge, this is the first case to be reported in this age group with both conditions presenting together.

Reporting it is very important to give more awareness about this category of patients who did not respond to the usual treatment when they present with fever and to keep *Leishmania* infection in their differential diagnosis especially if they are from an endemic areas.

Reference

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