

Hyper-reactive malarial splenomegaly: about a case

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ABSTRACT

Malaria is one of the most severe public health problems worldwide. Hyper-reactive malarial splenomegaly (HMS) is a form of severe malaria. The diagnosis was based on the exclusion of other causes of splenomegaly, immunity to malaria, splenomegaly of at least 10 cm, a high serum concentration of Ig M. We report the case of a 43-year-old male Bangladeshi patient, a missionary in Congo for one year, who had developed a typical clinical feature of HMS.

Keywords: malaria, HMS, splenomegaly.

To Cite This Article

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1. INTRODUCTION

HMS is a relatively rare chronic complication of malaria, with a mortality rate that exceeds 50%, (Hamilton et al. 1974), Crane et al. (1972). Criteria for the diagnosis of HMS were first published in 1979 (Greenwood et al. 1979), with minor revisions in 1981 (Fakunle et al. 1981). The diagnosis was based on the exclusion of other causes of splenomegaly, immunity to malaria, splenomegaly of at least 10 cm, a serum concentration of IgM, and a clinical and immunological response to malaria prophylaxis.

2. CASE REPORT

We report the case of a 43-year-old male Bangladeshi patient, a missionary in Congo for one year, who had developed a typical clinical feature of HMS characterized by massive splenomegaly with hypersplenism, high titers of malarial Ig M antibodies, and a clinical and immunological response to long-term chloroquine therapy. The investigations were not consistent with any other diagnosis. HMS is also briefly reviewed. The ultrasound examination of the abdomen (Figure 1) demonstrated massive splenomegaly (17 cm), with no hepatomegaly, and no radiological evidence of portal hypertension. The ultrasound after treatment showed regression of splenomegaly (Figure 2).

3. DISCUSSION

Malaria is one of the most severe public health problems worldwide. It is a leading cause of death and disease in many developing countries, where young children and pregnant women are the groups most affected. 3.3 billion people (half the world's population) live in areas at risk of malaria transmission in 106 countries and territories (Figure 3). The spleen plays an important role producing antibodies against the malarial parasites. Changes in splenic structure during the course of malaria can result in asymptomatic enlargement or complications such as haematoma formation, rupture, hypersplenism, ectopic spleen, torsion or cyst formation. HMS is a syndrome of massive, unexplained splenomegaly occurring in a malarious region, accompanied by lassitude, fever, weight loss, hypergammaglobulinemia, especially IgM, and cryoglobulinemia. Symptoms of splenomegaly consist primarily of left upper quadrant pain with or without signs of hypersplenism dominating the clinical presentation of HMS. Early in the syndrome, the pain may be episodic and exacerbated by physical activity, which over time progresses in intensity and becomes persistent and debilitating. The HMS which represents an aberrant response to malaria is characterized by high antimalarial antibody titers. The immunologic process stimulates the reticuloendothelial hyperplasia and eventually produces splenomegaly (El-Zayadi et al. 1992; Ali et al. 2004). There are specific criteria for the diagnosis (Table 1).

Our patient presented with the classical symptoms of HMS: abdominal mass, hypersplenism (anaemia, thrombocytopenia and leukopenia) and clinical response to

Cryoglobulinemia:
Cryoglobulins are single or mixed immunoglobulins that undergo reversible precipitation at low temperatures.

Haematoma:
Haematoma refers to a collection of blood outside of the blood vessels, which gathers in body tissues or cavities. Haematoma are most commonly apparent as bruising to the skin, and are caused by internal bleeding into the extracellular space following blunt trauma - this can include accidents, falls and surgery.

Table 1

Diagnostic criteria for hyperreactive malarial splenomegaly (Fakunle 1981)

Major diagnostic criteria	Minor diagnostic criteria
<ul style="list-style-type: none"> - gross splenomegaly in older children and adults - high antibody levels for <i>P. falciparum</i> - elevated serum IgM (at least 2 standard deviations above the mean of the population) - clinical and immunological response to long-term appropriate therapy 	<ul style="list-style-type: none"> - hepatic sinusoidal lymphocytosis (+ 80% of cases) - normal cellular and humoral immune responses to antigenic challenge, included PHA - hypersplenism - lymphocyte proliferation (in some populations) - occurrence within families, tribes



Figure 1
Ultrasound showing splenomegaly



Figure 2
Ultrasound control: regression of splenomegaly

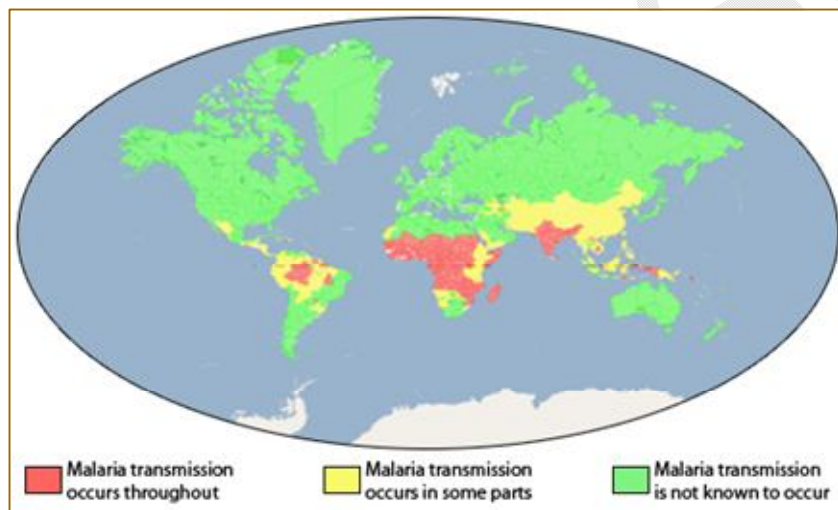


Figure 3
The geographical distribution of malaria

anti malarial drugs. Malaria parasites were not demonstrated in his peripheral blood smear, which could be because malaria antigenemia is not usually high in patients with HMS (Mahapatra et al. 2010). The patient was treated with chloroquine 300 mg weekly in accordance with the standard treatment protocol. We observed after 50 days an important reduction in spleen size. Eradication of parasitaemia seems to be the common pathway for resolution as successful treatment of HMS has been with antimalarial drugs especially, paludrine or chloroquine and in some instances a combination of paludrine and sulphadoxine or chloroquine (Kanwar et al. 2008; Onuigbo et al. 1992). The drugs may have to be given for long periods (years) before a response is noted. The duration of treatment is unknown (Kanwar et al. 2008). The place of splenectomy in the treatment of this disorder is still controversial.

4. CONCLUSION

The unusual immunological features of HMS, might contribute to our understanding of the mechanisms involved in pathogenicity and immunity to malaria.

CONFLICT OF INTEREST

None

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Ali et al. (2004):

Objective:

To find out the relative frequency of clinical conditions associated with splenomegaly that require hematological evaluation in our set up.

Design:

Cross sectional study.

Place and duration of study:

Combined Military Hospital, Quetta, Balochistan, from July 2000 to July 2003.

Subjects and methods:

Patients of either gender and all age groups with palpable spleen were included. Patients with splenomegaly due to liver disease, malarial parasites on thick or thin blood film, positive Widal test, or positive blood cultures were excluded from study.

Patients were initially evaluated with clinical history, microscopic examination of blood smear, and blood counts.

Depending upon provisional diagnosis bone marrow examination or investigations for hemolytic anemia were performed.

Results:

One hundred patients were received. Seventy-eight patients were adults and 22 patients were of pediatric age group. In the adults, hematological malignancies were seen in 37%, malarial parasites in bone marrow in 20.5%, megaloblastic anemia in 13%, bacterial infections in 9%, hemolytic anemia in 9%, tropical splenomegaly in 5%, and positive bone marrow culture for salmonella in 6.5%. In children, hematological evaluation revealed hematological malignancies in 18%, beta thalassaemia in 55%, other hemolytic anemias in 13.5%, congenital sideroblastic anemia in 4.5%, and storage disorder in 9%.

Conclusion:

Hematological workup is informative in most of the cases. Bone marrow examination is the key investigation, hematological malignancies constituted 37% of the adult and 18% of pediatric age group patients. Hemolytic anemia constituted 68% of pediatric age group.

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