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RELATO DE CASO

REDUCTION OF SPLEEN SIZE IN A CHILD WITH HYPERREACTIVE MALARIOUS SPLENOMEGALY (HMS) TREATED OUTSIDE THE BRAZILIAN ENDEMIC AREA OF MALARIA WITH ONLY ONE COURSE OF QUININE

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We report the clinical picture, treatment and evolution of a child with hyperreactive malarious splenomegaly treated outside the endemic area of malaria. The patient presented gross splenomegaly, proceeded from an area where malaria is endemic, showed increased immunoglobulins levels, high antimalarial antibody titres and hepatic sinusoidal lymphocytosis. The child did not return to an area where malaria is endemic and showed a favorable response to only one course of quinine. The response of this patient to limited antimalarial therapy suggests the importance of reinfection with malaria in the development and maintenance of this syndrome.

Key-words: Hyperreactive malarious splenomegaly (HMS). Chronic malaria. Short antimalarial therapy.

Hyperreactive malarious splenomegaly (HMS) has been described from malarious areas throughout the tropics[1 2 3 4 5 11 14], and is usually characterised by marked hepatosplenomegaly, raised immunoglobulins, a high titre of antimalarial antibodies and hepatic sinusoidal lymphocytosis[7 8 9 10].

The pathogenesis of this syndrome is not clarified[6], but there is strong evidence that malaria play an important role[3 6 7 9]. Genetic factors governing a disregulation of the immune response has been suggested as an explanation of this syndrome[3 7].

In areas of high malaria transmission successful therapy of this syndrome implies that antimalarials must be used regularly and for a prolonged time. The syndrome may recur if the patient stops this treatment in such areas[6 7 9 13].

We describe here the clinical picture, treatment and evolution of a child with this syndrome treated in the University Hospital of Brasília. This child did not return to an area where malaria is endemic and responded to limited antimalarial therapy.

CASE REPORT

The patient was an eleven year old, white girl who arrived from state of Pará, Brazil, with a palpable spleen 8cm from the left costal margin reaching the umbilicus (Hackett grade 4).

The haemoglobin was 10.8g% and the reticulocyte was 1.3%.The white blood cell count was 7000/mm^3, with a normal differencial count. The platelet count was 190500/mm^3, and the erytrocyte sedimentation rate 16mm in the first hour. Haemoglobin electrophorese showed only haemoglobin A1. Hepatic and renal function were normal. The total serum protein was 7.0g%, with 3.0g% of total globulin (alpha 1=0.15g%; alpha 2=0.50g%; beta=0.55g% and gamma=1.8g%). The diameter of the portal vein was normal when evaluated by abdominal ecography. Oesophageal varices were not detected on barium swallow. The serum gammaglobulin estimation showed 2650mg/dl of IgG, 271mg/dl of IgM and 144mg/dl of IgA. Malaria parasites were not identified in multiple peripheral blood films. The indirect immunofluorescent antibody test using *Plasmodium*

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falciparum antigen showed a titre of 320 for IgG and 80 for IgM. Histophatological examination of the liver obtained by percutaneous biopsy identified lymphocytosis in dilated sinusoids (grade II)[8]. The portal tracts also showed lymphocyte infiltration without portal fibrosis (Figure1).

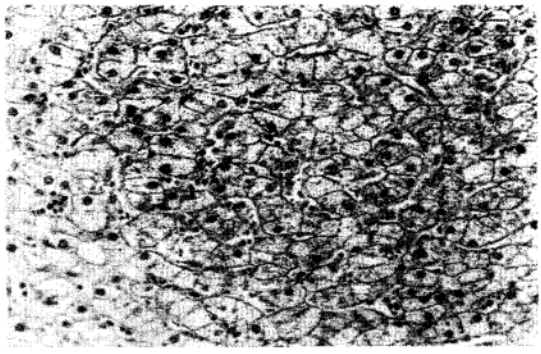

Figure 1 - Mag. 1000X. Liver biopsy histology showing sinusoidal dilatation and Grade II hepatic sinusoidal lymphocytosis. (Ref. 7)

The patient had most of the criteria defined for the diagnosis of the HMS. She presented with a gross splenomegaly, proceeded from an area where malaria is endemic, showed increased immunoglobulins levels, high antimalarial antibody titres and hepatic sinusoidal lymphocytosis. Curiously hepatomegaly was not detected and the IgM level was normal.

As the child was living in Brasília for 3 months and would not return to area where malaria is endemic it was decided to treat the child with a curative dose of ten days oral quinine sulphate, 30 mg/kg/day.

Seventy days after the treatment, the splenomegaly had reduced to 3cm from the left costal margin and the indirect immunofluorescent antibody test to *P. falciparum* showed a titre of 160 to IgG and 40 to IgM. Unfortunately the patient was lost to further following and repeat liver biopsy was not possible.

DISCUSSION

For established HMS short term therapy with antimalarial had been attempted with disappointing results[9]. Watson Williams and Allan in 1968 reported for the first time success with prolonged treatment with antimalarials for patients with this syndrome[15]. HMS usually occurs in endemic areas where malarial transmission is intense and children may be exposed to about 2 or 3 bites from infected mosquitoes per week[6]. Therefore it is clear that treatment must be regular and prolonged[7 9]. In patients living in malarial endemic areas and submitted to this treatment it has needed 6 months to one year of continuous therapy to have a measurable improvement in spleen size and antibody titre[9 12 13].

The case reported here presented the peculiarity that she was emigrating from the endemic area, and this provided the opportunity to evaluate therapy with only one course of quinine in a situation where repeated malarial infection could not occur. Quinine was used because chloroquine resistance is so frequent in Brazilian falciparum infections. We observed after 70 days an important reduction in spleen size and the serum levels of antimalarial antibodies tended to decrease. Unfortunately full follow up proved impossible.

The favourable response of this patient to only one course of quinine suggests the importance of reinfection with malaria in the development and maintenance of this syndrome and attributes an important role to an alteration in immunereactivity to *Plasmodium* in such patients.

RESUMO

Relatamos o caso clínico, tratamento e evolução de uma criança com a síndrome da esplenomegalia hiperreativa da malária tratada fora da área endêmica para a malária. A criança apresentava importante esplenomegalia, era procedente de área endêmica para malária, os níveis de imunoglobulinas e de anticorpos antimaláricos estavam elevados e observou-se linfocitose sinusoidal hepática. A criança não voltaria mais para a área endêmica de malária, pelo que foi tratada com apenas um curso de quinino apresentando resposta clínica favorável. Esta resposta a um único curso de terapia curativa antimalárica sugere a importância da reinfecção com o parasita da malária no desenvolvimento e na manutenção desta síndrome.

Palavras-chaves: Esplenomegalia hiperreativa da malária. Malária crônica. Tratamento curativo antimalárico.

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