Clinical expression of autoimmune hepatitis in a nine-year-old girl with visceral leishmaniasis

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Autoimmune hepatitis (AIH) is a chronic disease of unknown aetiology, which usually progresses to cirrhosis if not diagnosed and treated promptly. In childhood, autoimmune hepatitis prevalently presents with non-specific indications. The cause of entrance to hospital of the 9-year-old child was fever for 6 days (39°C) and intense cough, with other clinical symptoms and signs: abdominal distention, paleness, lack of appetite. In our case we report experience with AIH presenting with atypical clinical features of visceral leishmaniasis.

Key words: child, autoimmune hepatitis, visceral leishmaniasis.

Introduction

Autoimmune hepatitis (AIH) is a chronic disease of unknown aetiology, which usually progresses to cirrhosis if not diagnosed and treated promptly. Without treatment, AIH often progresses to cirrhosis and in more severe case, carries a high incidence of mortality and low rate of spontaneous remission [1]. In childhood, autoimmune hepatitis prevalently presents with non-specific indications, including hepatomegaly, stepping up serum transaminase and hypergammaglobulinemia, and it is characterized by the presence of autoantibodies [2, 3].

We report our experience with AIH presenting with atypical clinical features of visceral leishmaniasis.

Case report

A 9-year-old child from a cattle farm in Greece entered the paediatric section of the University Hospital in Thessaly. The patient's clinical symptoms and signs during the time of admission were fever, abdominal distention, paleness, lack of appetite and non-productive cough.

During the clinical checking there was hepatosplenomegaly and laboratory analysis showed anaemia, leucopoenia, hypergammaglobulinaemia, (IgG), and raised levels of AST (96 IU/l) and ALT (95 IU/l). The clinical picture is consistent with that of malaria, typhoid fever, miliary tuberculosis, schistosomiasis, brucellosis, amoebic liver abscess, infection mononucleosis, lymphoma and leukaemia [3-11]. The most common causes of infection and metabolic liver disease, such as viral hepatitis, were excluded.

Cause of entrance to hospital of the 9-year-old child was fever for 6 days (39°C) and intense cough.

Despite the presence of hepatosplenomegaly associated with mild anaemia and hypergammaglobulinaemia, the fact that the patient came from a region endemic for visceral leishmaniasis raised the suspicion of leishmaniasis, but laboratory tests showed an erythrocyte sedimentation rate of 125 mm in the first hour, CRP 1.8 (+), white blood cells 49 000/mm^3, platelets 328 000/mm^3, Hb 9.8 g/dl, Ht 27.5%, Glucose 79 mg/dl, γ-GT 8 IU/L, ALP 89 IU/l, LDH 218 IU/l, CPK 22 IU/l, AST 77 (248) IU/l, ALT 59 (211) IU/l, IgG 6720 mg/sl, IgA 122 mg/dl, IgM 177 mg/dl, IgE 30.4 mg/dl and Coomb's-positive autoimmune haemolytic anaemia and serum autoantibodies for (antinuclear antibody ANA, smooth muscle antibody SMA and A-ANCA) were seropositive just as one more factor that hypergammaglobulinaemia with selective elevation of serum IgG which is responsible and characteristic of autoimmune hep-

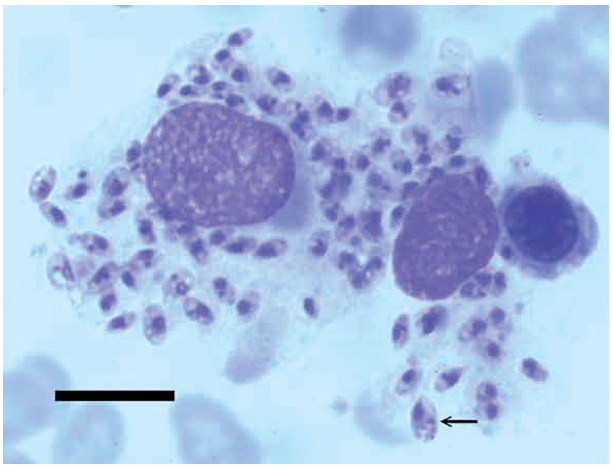

Fig. 1. Light micrograph of bone marrow, showing macrophages containing amastigotes characteristic of Leishmania. Amastigotes appear as oval organisms 2–6 μm in length that contain two darker-staining organelles (a nucleus and a kinetoplast – arrow). Scale bar = 10 μm

atitis. Bone marrow aspiration performed on day 2 to investigate the cause of the pancytopenia showed cellular bone marrow with relative erythroid hyperplasia. Multiple blood cultures gave negative results. We started treatment with azathioprine intravenous 5 mg/kg/24 h, but after new laboratory findings for identified detection Leishmania DNA-PCR test, the results confirmed the diagnosis of visceral leishmaniasis. Furthermore, a second bone marrow biopsy at this time revealed occasional macrophages containing amastigotes (the resting intracellular stage of Leishmania, formerly known as Leishman-Donovan bodies; Fig. 1). Review of the first bone marrow biopsy specimen failed to show any protozoa.

Visceral leishmaniasis is a systemic illness caused by intracellular protozoa of the *Leishmania* bonovi complex [3-5, 10]. Classic visceral leishmaniasis is characterized by irregular fever, pancytopenia, hepatosplenomegaly, hypergammaglobulinaemia, and the production of a wide spectrum of autoantibodies [1, 2, 4].

Liposomal amphotericin B and prednisolone treatment was started. A total dose of 4 mg/kg/24 h and tablets 5 mg/24 h of prednisolone was administered with complete regression of clinical and laboratory abnormalities. The patient's blood count returned to normal 2 months after starting treatment with amphotericin B.

Conclusion

Visceral leishmaniasis must be considered in the differential diagnosis of hepatitis (mimicking the clinical picture of autoimmune hepatitis), associated with anaemia and hypergammaglobulinaemia, especially in children from endemic regions.

Moreover, visceral leishmaniasis should be suspected in patients with fever, hepatosplenomegaly and cytopenia, especially if they reside in the Mediterranean region.

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