Italian Journal of Medicine (2011) 5, 274—277

Available online at www.sciencedirect.com

SciVerse ScienceDirect

journal homepage: www.elsevier.com/locate/itjm

CASE REPORT

Rare splenic complications and specific serology: decisive diagnostic tools in two cases of visceral leishmaniasis

Rare complicanze spleniche e diagnosi sierologica: strumenti diagnostici decisivi in due casi di leishmaniosi viscerale

Andrea Celestini [a,*], Federica Paglia [b], Orlando Dell' Unto [c], Riccardo Guarisco [c], Claudio Puoti [c]

[a] *UOC Cardiologia, Ospedale San Paolo Civitavecchia, Roma*
[b] *UOC Medicina d'Urgenza, Ospedale Sandro Pertini, Roma*
[c] *UOC Medicina, Ospedale San Giuseppe, Marino, Roma*

Received 19 November 2010; accepted 18 July 2011
available online 13 September 2011

KEYWORDS
Visceral Leishmaniasis;
Splenic infarction;
Splenic rupture;
Serological tests.

Summary
Introduction: Visceral leishmaniasis (VL) is a major endemic vector-borne disease in Southern Europe. We present two cases of VL, both characterized by splenic complications.
Methods and results: Case 1: A 47-year-old female presented with effort angina, hepatosplenomegaly and pancytopenia. The clinical course was complicated by splenic infarction. Although bone marrow biopsy failed to show amastigotes, diagnosis was performed by a fast agglutinating screening test (FAST) and confirmed by a direct agglutinating test (DAT). The patient was treated successfully with AmBisome. *Case 2:* A 22-year-old male who had undergone a splenectomy to treat splenic rupture related to a minor trauma four months earlier presented with fever, nocturnal sweats and weight loss. The lack of pancytopenia was due to the absence of the spleen. The first biopsy did not identify parasites, but because the FAST had been positive, another bone marrow biopsy was performed, which demonstrated leishmaniasis. This patient was treated with the same schedule of AmBisome infusion.
Discussion: 1) The clinical presentation of VL can be atypical, 2) splenic complications can characterize this disease, and 3) specific serology may be an important tool to reach a diagnosis.

* Corresponding author: Via Reggio Calabria 6 - 00161 Roma, Italy.
E-mail: andrea.celestini@gmail.com (A. Celestini).

1877-9344/$ — see front matter
doi:10.1016/j.itjm.2011.07.005

Introduction

Visceral leishmaniasis (VL) or Kala-azar, an infectious disease caused by the bite of *Phlebotomus sp.*, is a major endemic vector-borne disease in Southern Europe. VL affects populations from 70 countries across Southeast Asia, East Africa, South America and the Mediterranean region; the worldwide incidence is estimated to be approximately 8.3/100,000, with approximately 51,000 deaths per year [1]. Although the reported annual incidence in Southern Europe ranges by country from 0.02/100,000 to 0.49/100,000 [1], these numbers are misleading given the number of misdiagnoses and asymptomatic infections. More than 20 pathogenetic species of *Leishmania* have been identified in the world. Most of these species can cause only the cutaneous form of leishmaniasis; *Leishmania infantum* is the only agent of VL in Europe, whereas *Leishmania donovani* is responsible for VL in the Indian sub-continent and in Africa [1,2].

After inoculation, the flagellated parasite (promastigote) transforms into an amastigote after being phagocytosed by dermal macrophages, localizing in the macrophagic lysosomes. The initial immune response against VL and the parasite subtype are fundamental to the subsequent development of systemic disease: a Th-1 type response is necessary for a protective reaction, whereas Th-2 differentiation usually induces the development of VL. The incubation period ranges from weeks to months; humans are considered incidental hosts, and dogs are the most frequent animal reservoir [2]. Patients generally exhibit cachexia with recurrent high fever, anorexia and night sweats; hepatosplenomegaly and hypersplenism-related pancytopenia are other common features. VL may coexist with other infectious diseases, and currently, HIV—VL coinfection is an emerging problem in developing countries. Because HIV and *Leishmania sp*. share the same target immune cells, they exert a synergistic damaging effect on the immune response and may induce atypical clinical manifestations, such as the visceralization of cutaneous leishmaniasis [1].

We report two cases of VL characterized by rare splenic complications.

Case 1

A 47-year-old Italian HIV-negative female (body weight 52 kg), who lived in a farm in South Latium, was admitted to our department presenting with wasting and anorexia over the last month, accompanied in the last two weeks by effort-related chest pain. No history of smoking or alcohol abuse was reported. Physical examination showed significant hepatosplenomegaly without appreciable lymphoadenopathy. Laboratory values revealed pancytopenia with a prevalent reduction in red blood cells and polyclonal hypergammaglobulinemia (for more details, see Table 1). A chest X-ray, an ECG and a cardiac ultrasound were normal. During hospitalization, epigastric tenderness developed, and the patient underwent an abdominal ultrasound that revealed a sub-capsular hypoechogenic area around the inferior wall of the spleen. This sign was confirmed by magnetic resonance imaging, which showed intrasplenic hyperintensity, consistent with a hemorrhagic infarction. No acquired (antiphospholipid antibodies, oral contraceptives) or congenital thrombophilic factors (factor V leiden, prothrombin G20210A or methylenetetrahydrofolate reductase mutations or a lack of antithrombin III, protein C or S) were evidenced. Moreover, tests for viral hepatitis markers, the Wright test and the tine test were negative. Although peripheral blood and bone marrow specimens failed to show amastigotes, a fast agglutinating screening test (FAST) and a direct agglutinating test for *Leishmania infantum* (DAT) (dilution of 1:4000) revealed the presence of anti-*Leishmania* antibodies. A splenic fine needle aspirate sample (FNAS) was not taken because taking such a sample was considered dangerous in the presence of a splenic hemorrhage, and polymerase chain reaction (PCR) was unavailable in our hospital. The patient was treated with liposomal amphotericin B (AmBisome) (4 mg/kg i.v for 5 days and on days 14 and 21). After treatment, a progressive improvement of the patient's anemic status and abdominal symptoms was observed.

Case 2

A 22-year-old Albanian male (body weight 72 kg), who had worked in Greece for a while and then had moved to Italy (South Latium), was admitted to our department. The patient, who had no history of alcohol abuse, presented with asthenia, weight loss, irregular fever and night sweats. Four months earlier, the patient had undergone a splenectomy to treat a splenic rupture resulting from a minor trauma. Macroscopic examination of the spleen had documented marked splenomegaly (21x19x7 cm) and lymphadenomegaly of the splenic hilum (2.2 cm). The histological diagnosis was primary hypersplenism.

Laboratory tests showed normal levels of platelets and white cells, slight anemia and increased levels of hepatolysis markers (for more details, see Table 2). Hepatotropic

Table 1 Case 1: lab values before and after treatment.

	date	r.b.c. ($x10^6/\mu l$)	w.b.c. ($x10^3/\mu l$)	hemoglobin (g/dl)	hematocrit (%)	platelets ($x10^3/\mu l$)	iron (μg/dl)	proteins (g/dl)	albumin (g/dl)
Before treatment	April, 28th	3.60	2.00	9.30	26.0	112	24	5.4	2.8
Before treatment	May, 05th	3.10	2.00	7.60	23.0	77			
After treatment	June, 09th	3.49	2.29	9.26	26.8	80			
After treatment	June, 14th	3.61	2.50	9.70	32.2	87		6.1	3.57
After treatment	June, 25th	4.40	2.80	11.70	34.8	189	75		
After treatment	July, 05th	4.46	3.03	12.00	34.8	161	70	7.2	4.3

Legend: r.b.c.: red blood cells, w.b.c: white blood cells.