Fever, splenomegaly and lymphopenia in sarcoidosis

**CLINICAL PRESENTATION**

A 42-year-old woman was referred to our department with a 5 months history of intermittent fever and fatigue. Her past medical history included a consolidated diagnosis of sarcoidosis (obtained 7 years before with a sub-carinal lymph node biopsy) and in the previous years she had been treated with steroids, hydroxychloroquine, methotrexate and azathioprine with persistent and progressive enlargement of mediastino-hilar adenopathies and bilateral nodular infiltrates. Her symptoms progressed despite a course of antibiotics prescribed for presumed community-acquired pneumonia and a course of corticosteroids. The patient lived in Italy and there was no history of travels or other additional risk factors for infections.

At the time of admission, the patient had a temperature of 38.5°C and the physical examination was only notable for hepatomegaly and splenomegaly. A white blood cell count showed lymphopenia (0.39×10^9/l, CD4 0.22×10^9/l) and anaemia (Hb 10.4 g/dl, MCV 78.7 fl).

CT images of the thorax revealed bilateral nodules of variable sizes predominantly in the upper lung fields and a positron emission tomography showed several areas of increased metabolism in the liver, spleen, lymph nodes (above and under the diaphragm), D3 vertebra and bilateral nodular infiltrates (figure 1).

CT/PET showed numerous areas of increased metabolism in the liver, splenic parenchyma, thoracic and abdominal lymph nodes.

**QUESTION**

What is the diagnosis?

See page below for the answer
Claudia Ravaglia,1 Carlo Gurioli,1 Gian Luca Casoni,1 Silvia Asioli,2 Venerino Poletti1

1Department of Diseases of the Thorax, Pulmonology Unit, Pierantoni-Morgagni Hospital, Forlì, Italy; 2Department of Pathology, Pierantoni-Morgagni Hospital, Forlì, Italy

Correspondence to Dr Venerino Poletti, Pulmonology, Department of Thoracic Diseases, GB Pierantoni—L Morgagni Hospital, via C. Forlanini 34, 47100 Forlì, Italy; venerino.poletti@gmail.com

Contributors All authors have made important contributions in the discussion of the clinical case and drafting the article.

Competing interests None.

Patient consent Obtained.

Ethics approval Ethics approval was provided by Area Vasta Romagna Review Board.

Provenance and peer review Not commissioned; externally peer reviewed.

This paper is freely available online under the BMJ journals unlocked scheme. see http://thorax.bmj.com/site/about/unlocked.xhtml.
From the question on page above

The pathological analysis of the bone marrow revealed diffuse interstitial infiltration of foamy macrophages containing *Leishmania* protozoa. The conclusive diagnosis was visceral leishmaniasis in immunocompromised patient. The patient was treated with liposomal amphotericin (3.5 mg/kg for 5 days and then with a maintenance dose); treatment was well tolerated and resulted in immediate regression of the fever and improvement in general state of health; C-reactive protein decreased from 87 mg/l to 24 mg/l and lymphocytes increased from 0.39 mg/l to 0.95 mg/l.

Fever, splenomegaly and lymphopenia may arise from a large range of infectious, haematological or systemic diseases and therefore represent a difficult diagnostic challenge.¹ In this case, the previous history of sarcoidosis was an apparent clue; however, we know from literature that sarcoidosis and lymphoma, mainly non-Hodgkin’s lymphoma, may occur together, with sarcoidosis usually preceding lymphoma² and the coexistence of sarcoidosis and opportunistic infection, even in the absence of any immunosuppressive therapy, has previously been documented.³ A possible infectious cause of fever, splenomegaly and lymphopenia is visceral leishmaniasis (VL, kala-azar), a systemic infection of the reticuloendothelial system caused by protozoa of the genus *Leishmania*. The definitive diagnosis of kala-azar requires demonstration or isolation of parasites from samples collected by invasive organ aspiration.⁴ VL is endemic in areas bordering the Mediterranean Sea; even though leishmaniasis is seen relatively infrequently in connection with sarcoidosis, our case presentation demonstrates that VL must be taken into consideration in the differential diagnosis of febrile splenomegaly in patients living in areas endemic for such protozoa.⁵

REFERENCES

Fever, splenomegaly and lymphopenia in sarcoidosis

Claudia Ravaglia, Carlo Gurioli, Gian Luca Casoni, Silvia Asioli and Venerino Poletti

Thorax  published online June 2, 2012

Updated information and services can be found at: http://thorax.bmj.com/content/early/2012/06/01/thoraxjnl-2011-201408

These include:

References
This article cites 5 articles, 1 of which you can access for free at: http://thorax.bmj.com/content/early/2012/06/01/thoraxjnl-2011-201408#BIBL

Open Access
This is an open-access article distributed under the terms of the Creative Commons Attribution Non-commercial License, which permits use, distribution, and reproduction in any medium, provided the original work is properly cited, the use is noncommercial and is otherwise in compliance with the license. See: http://creativecommons.org/licenses/by-nc/2.0/ and http://creativecommons.org/licenses/by-nc/2.0/legalcode.

Email alerting service
Receive free email alerts when new articles cite this article. Sign up in the box at the top right corner of the online article.

Topic Collections
Articles on similar topics can be found in the following collections

- Open access (257)
- Screening (oncology) (407)
- Tropical medicine (infectious diseases) (26)
- Cardiothoracic surgery (676)
- Lung infection (97)
- Pneumonia (infectious disease) (579)
- TB and other respiratory infections (1273)
- Lung cancer (oncology) (670)
- Lung cancer (respiratory medicine) (670)
- Lung neoplasms (608)
- Chemotherapy (183)
- Drugs: infectious diseases (968)
- Radiology (diagnostics) (812)
- Pneumonia (respiratory medicine) (562)

To request permissions go to: http://group.bmj.com/group/rights-licensing/permissions

To order reprints go to: http://journals.bmj.com/cgi/reprintform

To subscribe to BMJ go to: http://group.bmj.com/subscribe/