CASE REPORT

PCP: thinking beyond HIV

Alexei Gonzalez-Estrada, James Fernandez

Department of Allergy and
Clinical Immunology, Cleveland
Clinic, Cleveland, Ohio, USA

Correspondence to

Dr Alexei Gonzalez-Estrada,
gonzala5@ccf.org

SUMMARY

A 40-year-old man with a previous AIDS-defining opportunistic infection and five negative HIV tests presented to our outpatient clinic. The laboratory test was relevant for less than 300 total lymphocytes on two separate occasions. He was diagnosed with idiopathic CD4 lymphocytopenia and was started on antibiotic prophylaxis for *Pneumocystis carinii* pneumonia and *Micobacterium avium-intracellulare* infection (MAI). This case report summarises the importance of further immunological characterisation in patients presenting with opportunistic infections and decreased cellular immunity.

BACKGROUND

Idiopathic CD4 lymphocytopenia (ICL) is a rare disease, characterised by a decreased number of circulating CD4T lymphocytes (<300 cells/ μ L or <20% total lymphocytes) on a minimum of two separate time points at least 6 weeks apart. Because it is a diagnosis of exclusion, other laboratory evidence of infections with HIV should be ruled out. This entity is often overlooked by clinicians. This case report was written with the purpose of informing the medical community to be suspicious of patient with AIDS-defining disease and multiple negative HIV tests.

CASE PRESENTATION

A 40-year-old man presented for evaluation of low CD4 count. He initially presented to his primary care physician when he was 30 years old with worsening shortness of breath. He was diagnosed with bronchitis and given antibiotics. He showed a poor response to the oral antibiotics and eventually required hospitalisation and intravenous antibiotics. At that time a chest X-ray showed bilateral infiltrates; a CT of the chest confirmed bilateral infiltrates with a right pneumothorax which required chest tube placement. His clinical course was not improving and an open lung biopsy was performed which revealed *Pneumocystis carinii* pneumonia (PCP). A laboratory test at the time was relevant for total lymphocyte count of 8 cells/ μ L and a negative HIV test on ELISA and RNA testing. The patient completed a course of intravenous bactrim and was discharged home on PCP prophylaxis. He was evaluated by multiple physicians after discharge with no clear diagnosis. He had five negative HIV tests, repeatedly denied all HIV risk factors and the remainder of his medical history was deemed unremarkable.

INVESTIGATIONS

A CT of the chest and abdomen was negative for granulomas and lymphoproliferative disease. The laboratory tests were relevant for negative HIV, normal complete blood count, complete metabolic panel, antinuclear antibody, erythrocyte sedimentation rate, C reactive protein, immunoglobulins, adequate response to vaccines, low total T cells 235 (range 958–2388 cells/ μ L) and low CD3/CD4 T cells 63 (range 533–1674 cells/ μ L). Bone marrow biopsy was normal.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis of this condition in adults compromises primarily HIV, sarcoidosis, common variable immunodeficiency, neoplastic disease, acute respiratory distress syndrome and various autoimmune disorders. This patient had multiple negative HIV tests in the past and other testing which made an alternative diagnosis unlikely.

TREATMENT

There is no standardised treatment for ICL in general. A prompt treatment of acute infections along with appropriate prophylaxis is necessary. Therefore, azithromycin was added to his prophylactic regimen. A possible more aggressive therapy such as bone marrow transplant or interleukin (IL) 2 therapy in future was discussed with the patient.

OUTCOME AND FOLLOW-UP

The patient remains on prophylaxis for opportunistic infections. He has remained stable with no further infections.

DISCUSSION

ICL is a rare disease, characterised by a decreased number of circulating CD4T lymphocytes (<300 cells/ μ L or <20% total lymphocytes) on a minimum of two separate time points at least 6 weeks apart,¹ with no laboratory evidence of infections with HIV. The clinical presentation of ICL varies from asymptomatic laboratory abnormality to life-threatening complications that mimic HIV-infected patients.² Cryptococcosis is the most commonly described infection. The treatment of ICL entails treatment and prophylaxis of secondary infections. In cases of severe refractory infections, alternative approaches exist to increase CD4 lymphocytes; these include IL-2 therapy³ or allogeneic bone marrow transplant.⁴

CrossMark

To cite: Gonzalez-Estrada A, Fernandez J. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2013-201687

Learning points

- ▶ Idiopathic CD4 lymphocytopenia (ICL) is very rare. It is diagnosed with a decreased number of circulating CD4T lymphocytes (<300 cells/ μ L or <20% total lymphocytes) on a minimum of two separate time points at least 6 weeks apart.¹
- ▶ Further investigation is required in patients with opportunistic infections and negative HIV testing.
- ▶ There is no standard therapy for ICL but prophylaxis of opportunistic infections should be considered based on CD4 count, similar to patients who are HIV positive.
- ▶ For severe cases of ICL where infections cannot be controlled on prophylactic antibiotics, interleukin 2 therapy or allogenic bone marrow transplant should be considered.

Competing interests None.

Patient Consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Walker UA, Warnatz K. Idiopathic CD4 lymphocytopenia. *Curr Opin Rheumatol* 2006;18:389–95.
- 2 Smith DK, Neal JJ, Homberg SD. Unexplained opportunistic infections and CD4+ T-lymphocytopenia without HIV infection. An investigation of cases in the United States. The Centers for Disease Control Idiopathic CD4+ T-lymphocytopenia Task Force. *N Engl J Med* 1993;328:373–9.
- 3 Cunningham-Rundles C, Murray HW, Smith JP. Treatment of idiopathic CD4T lymphocytopenia with IL-2. *Clin Exp Immunol* 1999;116:322–5.
- 4 Peterson EJ, Rozenberg-Arska M, Dekker AW, *et al.* Allogenic bone marrow transplantation can restore CD4+ T-lymphocyte count and immune function in idiopathic CD4+ T-lymphocytopenia. *Bone Marrow Transplant* 1996;18:813–15.

Copyright 2013 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow