Joint Bone Spine 86 (2019) 273–274

Available online at
ScienceDirect
www.sciencedirect.com

Elsevier Masson France
EM|consulte
www.em-consulte.com/en

Letter to the Editor

Angioimmunoblastic T-cell lymphoma mimicking eosinophilic granulomatosis with polyangiitis (Churg-Strauss)

ARTICLE INFO

Keywords:
Angioimmunoblastic T-cell lymphoma
Eosinophilic granulomatosis with polyangiitis, cryptococcosis

Eosinophilic granulomatosis with polyangiitis (EGPA, formerly called Churg-Strauss syndrome) is a rare systemic small-vessel vasculitis, associated with asthma and blood and tissue eosinophilia [1]. Unmet needs remain in this disease, but refractory disease always raises the question of misdiagnosis.

A 55-year old woman, breeding wood pigeons, complained about a one-year history asthenia, dry cough and dyspnea evolving by acute exacerbation, considered as asthma. Initial blood tests revealed eosinophilia (2190/mm^3), increased C-reactive protein (CRP) 17 mg/L, positive antineutrophil cytoplasmic antibodies (ANCA) with a perinuclear pattern (1/1280) without any specificity. Chest CT-scan showed bilateral reticulonodular and ground-glass opacities, bilateral nodules, left lower lobe subpleural consolidation (Fig. 1A) diagnosis of EGPA was made. Oral glucocorticoids (1 mg/kg/day) were initiated with partial improvement, but disease relapsed after 2 months of follow-up and BALF analysis showed 275,000 cells per mm^3 with predominant eosinophils (74%). Treatment with 3 pulses of 1 gram methylprednisolone followed by oral prednisone led to overall improvement, but disease relapsed below 20 mg/day of prednisone. Because of relapsing EGPA, intravenous cyclophosphamide (CYC) was administered in combination with glucocorticoids, but the disease remained refractory.

Then, the patient complained of severe headaches with nausea and photophobia related to neuromeningeal cryptococcosis (Fig. 1B), treated with intravenous amphotericin B and flucytosine, followed by oral fluconazole. Severe lymphopenia was noted at that time (CD4$^+$ 140/mm^3, CD8$^+$ 53/mm^3, CD19$^+$ 35/mm^3), as before starting CYC. PET/CT-scan showed homogeneous bone marrow and thyroid 18F-FDG uptake, cervical, diffuse hypermetabolic lymph nodes (Fig. 1C). Lymph node and bone marrow biopsies finally revealed angioimmunoblastic T-cell lymphoma (AITL) (Fig. 1D), involving thyroid and lungs, associated with reactive eosinophilia. The patient received 6 courses of high-dose polychemotherapy leading to clinical, radiological, biological remission and to glucocorticoids discontinuation. After three months, the patient relapsed through the same phenotype. Second-line-chemotherapy, including 3 courses of bendamustine, was fatally complicated by acute respiratory distress syndrome, CMV reactivation and infectious pneumonia.

Constitutional symptoms and pulmonary or skin involvement in the setting of B-cell or T-cell lymphoma may mimic EGPA, with sometimes histological evidence of granulomas and/or necrotizing vasculitis and similar radiological pattern. AITL is a rare and aggressive T-cell lymphoma that accounts for 1–2% of all non-Hodgkin lymphoma, commonly presenting with constitutional symptoms, skin rash, autoimmune manifestations [2], and with eosinophilia in 39% of cases of AITL [2]. Also, ANCA positivity without any specificity remains consistent with AITL diagnosis as autoantibodies are frequently detected [3].

Neuromeningeal cryptococcosis is unusual in the setting of EGPA and occurs as the consequence of profound immunosuppression, and is most likely related to the underlying hemopathy in the context of occupational exposure. Although initial clinical and CT-scan findings were compatible with pulmonary cryptococcosis [4], microbiological features and and improvement using polychemotherapy strongly suggest a specific AITL involvement.

This case highlights the multifaceted clinical presentation and the wide spectrum of autoimmune manifestations that AITL may mimic. Underlying T-cell lymphoma should be considered in patients with suspected EGPA, in particular in the absence of asthma or in case of unusual opportunistic infection in this context.

https://doi.org/10.1016/j.jbspin.2018.06.002

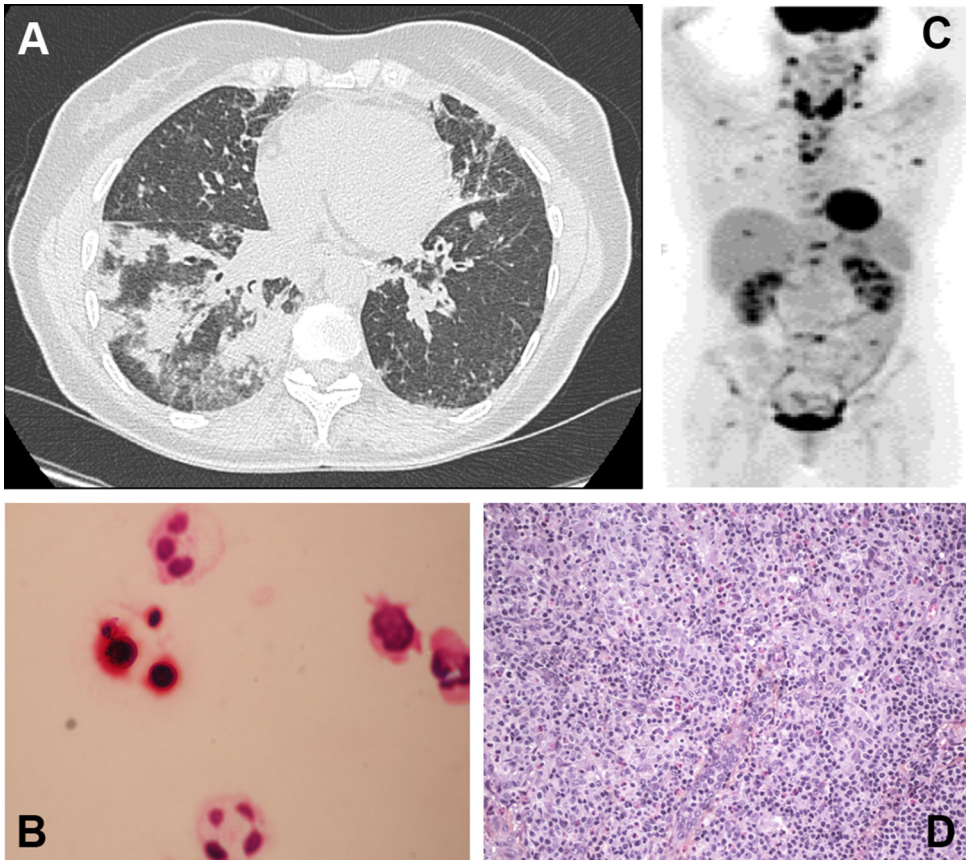

Fig. 1. A. Chest CT-scan showing lesions mimicking EGPA with reticulonodular and ground-glass opacities, left lower lobe subpleural consolidation and bilateral nodules. B. Microscopic examination of cerebrospinal fluid (Gram coloration): in the centre, two yeasts (one on the left budding from another one) of *Cryptococcus neoformans*. C. PET/CT-scan showing disseminated hypermetabolic lymphadenopathies, diffuse homogeneous bone marrow and thyroid 18F-FDG uptake. D. Microscopic examination of a lymph node (HE, magnification × 200): cell infiltrate with eosinophils, plasmocytes, medium and large atypical cells with vascular hyperplasia, consistent with the diagnosis of angioimmunoblastic T-cell lymphoma.

Funding

No specific funding was received from any bodies in the public, commercial or not-for-profit sectors to carry out the work described in this article.

Disclosure of interest

The authors declare that they have no competing interest.

Acknowledgements

The authors want to acknowledge Dr. Marie-Cécile Parrens and Dr. Philippe Blanche, for their participation of the management of the patient.

References

[1] Jennette JC, Falk RJ, Bacon PA, et al. 2012 revised International Chapel Hill Consensus Conference Nomenclature of Vasculitides. Arthritis Rheum 2013;65:1–11.
[2] Lunning MA, Vose JM. Angioimmunoblastic T-cell lymphoma: the many-faced lymphoma. Blood 2017;129:1095–102.
[3] Pautier P, Devidas A, Delmer A, et al. Angioimmunoblastic-like T-cell non Hodgkin's lymphoma: outcome after chemotherapy in 33 patients and review of the literature. Leuk Lymphoma 1999;32:545–52.
[4] Chang W-C, Tzao C, Hsu H-H, et al. Pulmonary cryptococcosis: comparison of clinical and radiographic characteristics in immunocompetent and immunocompromised patients. Chest 2006;129:333–40.

Audrey Le Roy[a,b,c]
Benjamin Terrier[a,b,*]
Jonathan London[a,b]
André Paugam[d]
Luc Mouthon[a,b]
François Lifermann[e]
[a] *Department of Internal Medicine, Cochin hospital, centre de référence maladies systémiques et autoimmunes rares, Assistance publique–hôpitaux de Paris (AP–HP), 27, rue du Faubourg-Saint-Jacques, 75014 Paris, France*
[b] *Université Paris Descartes, 75006 Paris, France*
[c] *French Military Health Academy, École-du-Val-de-Grâce, 75005 Paris, France*
[d] *Department of Parasitology, Cochin hospital, Assistance publique–hôpitaux de Paris (AP–HP), 75014 Paris, France*
[e] *Department of Internal Medicine, 40100 Dax, France*

[*] Corresponding author at: Department of Internal Medicine, Cochin hospital, centre de référence maladies systémiques et autoimmunes rares, Assistance publique–hôpitaux de Paris (AP–HP), 27, rue du Faubourg Saint-Jacques, 75014 Paris, France.
E-mail address: benjamin.terrier@aphp.fr (B. Terrier)

Accepted 20 June 2018
Available online 30 June 2018