

WEGENER'S GRANULOMATOSIS: SKIN DEEP

Inês Pires Silva*, Carla Noronha**, António Panarra**, Nuno Riso***, Manuel Vaz Riscado****

Wegener's Granulomatosis (WG) is an ANCA-associated vasculitis whose clinical triad involves the upper respiratory airway, lungs and kidneys¹. Skin involvement has been observed in 14-47% of patients, either during or at onset of the disease and may develop on unusual sites such as trunk, neck and face².

Necrosis, granulomatous inflammation and vasculitis are histological hallmarks.

Case-report: 60 year-old, diabetic, caucasian male complaining of an eight months' evolution sero-hematic rhinorrhea, nasal obstruction and crusting and a diffuse purplish vesicular rash (Figures 1A, 1B), compatible with leucocytoclastic vasculitis; prednisolone 30 mg/day was then prescribed. A paranasal polypoid mass was excised via rhinoscopy (Figure 2), compatible with a chronic inflammatory process, fibrosis and media thickening of small arteries.

Microhematuria (though normal renal biopsy), polyarthralgia and bilateral recurrent episcleritis were also noted.

Chest X-ray, routine lab and immunological workup (including ANCA) were normal. A small-vessel vasculitis was diagnosed, probably WG. Due to an exuberant skin involvement and refractoriness to corticosteroids, clinical remission was achieved with a 6 months' regimen pulsed cyclophosphamide (1g/m²/month) plus prednisolone (1 mg/kg/day). He relapsed under AZA maintenance therapy (250 mg/day), leading to the use of Mycophenolate Mofetil (MMF- 3g/day), with sustained clinical

improvement (Figure 3).

This clinical case is particular in four keypoints: *an exuberant cutaneous involvement*, resembling pyoderma gangrenosum, a rare manifestation of WG; the uncommon *absence of pulmonary or renal involvement* (20% of cases)¹; *a negative c-ANCA*, possible in limited or inactive GW (65-70%), which, adding to predominant skin and nasal affection,



Figure 1A and 1B. Exuberant cutaneous involvement characterized by lesions in several stages-diffuse purplish papules, pustules, vesicles, nodules, coalescent and sometimes necrotic (Pyoderma Gangrenosum-like)

*Oncology Resident, Oncology Department, Instituto Português de Oncologia de Lisboa Francisco Gentil, Lisbon, Portugal; Programme for Advanced Medical Education

**Attending Physician in Internal Medicine, Autoimmune Diseases' Unit, Medical Department 2, Curry Cabral Hospital, Lisbon, Portugal

***Consultant Physician in Internal Medicine, Autoimmune Diseases' Unit, Medical Department 2, Curry Cabral Hospital, Lisbon, Portugal

****Chief of Department in Internal Medicine, Autoimmune Diseases' Unit, Medical Department 2, Curry Cabral Hospital, Lisbon, Portugal



Figure 2. Polipoid mass in the paranasal sinuses (macroscopic aspect)

favors a limited WG diagnosis in our case; and a *sustained clinical remission under maintenance with MMF without toxicity*.

A high rate of disease relapse (20-45%) after cyclophosphamide's induction therapy prompts the need for additional options¹⁰. Our choice was dictated by MMF safety profile, case series reports³⁻⁷ and satisfactory experience in lupus and small-vessel vasculitis. Nowack⁹ established MMF as well tolerable and effective for maintenance therapy in 9 patients with WG and 2 patients with microscopic polyangiitis, proving to be a promising, but still poorly studied drug in vasculitis.

Correspondence to

Dr. Inês Pires da Silva
Rua Manuel Marques, nr. 10, 9ºB;
1750-171 Lisboa, Portugal
E-mail: inespiresilva@gmail.com

Acknowledgements:

The Programme for Advanced Medical Education is sponsored by Fundação Calouste Gulbenkian, Fundação Champalimaud, Ministério da Saúde e Fundação para a Ciência e Tecnologia, Portugal.

The authors would also like to thank Dr. A. Marta Pimentel for cooperating in the ENT Consult and biopsy



Figure 3. Clinical improvement regarding cutaneous involvement

References

1. Toffart AC, Arbib F, Lantuejoul S et al. Wegener granulomatosis revealed by pleural effusion. Case Report Med 2009;164395. Epub 2010 Feb 4.
2. Le Hello C, Bonte I, Mora JJ, Verneuil L, Noel LH, Guillemin L. Pyoderma gangrenosum associated with Wegener's granulomatosis: partial response to mycophenolate mofetil. Rheumatology 2002; 41: 236-237.
3. Osuna A, Garrido J. Cyclophosphamide-intolerant Wegener's granulomatosis successfully treated with mycophenolate mofetil. Acta Reumatol Port 2008;33:224-228.
4. Nowack R, Birck R, van der Woude FJ. Mycophenolate mofetil for systemic vasculitis and IgA nephropathy. Lancet 1997;349:774.
5. Braasch E, Neumayer HH. Treatment of acute c-ANCA-positive vasculitis with mycophenolate mofetil. Am J Kidney Dis 1999; 34: e9-e9.
6. Woywodt A, Choi M, Schneider W, Kettritz R, Gobel U. Cytomegalovirus colitis during mycophenolate mofetil therapy for Wegener's granulomatosis. Am J Nephrol 2000;20:468-472.
7. Haubitz M, de Groot K. Tolerance of mycophenolate mofetil in end-stage renal disease patients with ANCA-associated vasculitis. Clin Nephrol 2002; 57:421-424.