WEGENER'S GRANULOMATOSIS: SKIN DEEP

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Wegener s Granulomatosis (WG) is an ANCA-associated vasculitis whose clinical triad involves the upper respiratory airway, lungs and kidneys[1]. 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 face[2].

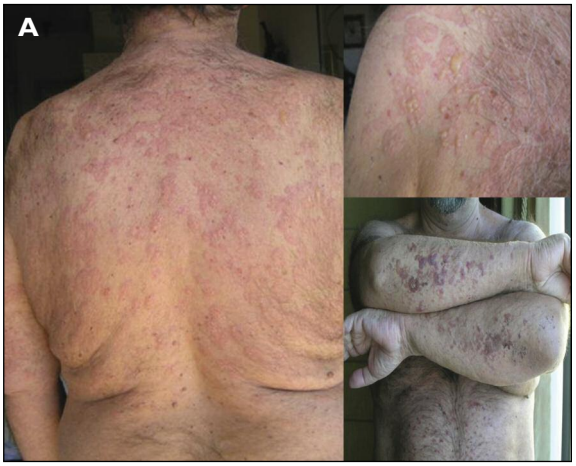
Necrosis, granulomatous inflammation and vasculitis are histological hallmarks.

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 polipoid 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 refractoriety to corticosteroids, clinical remission was achieved with a 6 months' regimen pulsed cyclophosphamide (1g/m^2/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,



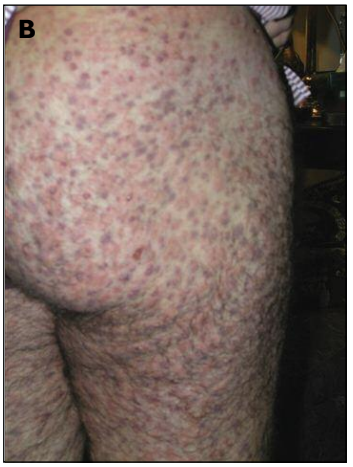


Figure 1A and 1B. Exuberant cutaneous involvement characterized by lesions in several stages-diffuse purplish papules, pustules, vesicles, nodules, coalescent and sometimes necrotic (Pyoderma Gangrenosum-like)

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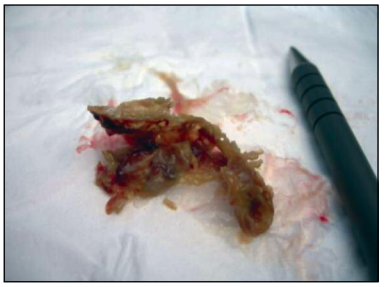

Figure 2. Polipoid mass in the paranasal sinuses (macroscopic aspect)

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 options[10]. Our choice was dictated by MMF safety profile, case series reports[3-7] and satisfactory experience in lupus and small-vessel vasculitis. Nowack[9] 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.

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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

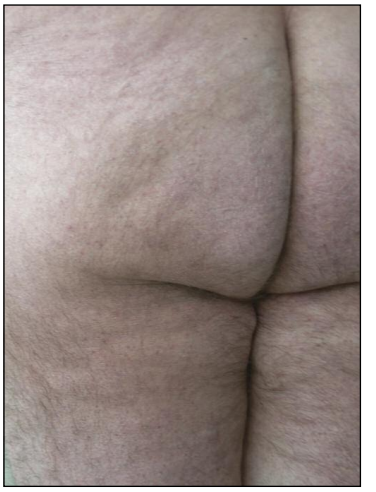

Figure 3. Clinical improvement regarding cutaneous involvement

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