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

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/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); a negative c-ANCA, possible in limited or inactive GW, which, adding to predominant skin and nasal affection,
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\textsuperscript{10}. Our choice was dictated by MMF safety profile, case series reports\textsuperscript{5,7} and satisfactory experience in lupus and small-vessel vasculitis. Nowack\textsuperscript{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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