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WEGENER'S GRANULOMATOSIS AS A DIFERENCIAL DIAGNOSIS OF MUCOCUTANEOUS LEISHMANIASIS: A CASE REPORT

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Abstract: The case report had its data kept confidential, in accordance with the provisions of Resolutions 466/12, 510/16 and 580/18 of the National Health Council, and supported by the General Law for the Protection of Personal Data 13.709/2018. It should be noted that data collection began after the application of the Informed Consent Form, with the knowledge and signature of the participant of the case report. The approving review board was the Ethics Committee of Associação Beneficente Santa Casa de Campo Grande (CEP/ABCG Santa Casa), located at Escola de Saúde da Santa Casa, Mato Grosso Avenue, 421, Center, Campo Grande; Postal code 79002-230, e-mail cep@santacasacg.org.br and phone number **+55 (67) 3322-4252.**
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INTRODUCTION

Granulomatosis with polyangiitis is a rare form of vasculitis affecting small vessels. Hallmark features include necrotizing granulomas and pauci-immune vasculitis, most commonly affecting the upper respiratory tract, lungs, and kidneys. (1). The diagnosis is based on clinical, laboratory, radiological findings and biopsies (2).

The upper respiratory system is affected in 70 to 100% of cases. It usually affects the nose and paranasal sinuses with manifestations such as chronic nasal obstruction, ulceration and edema of the nasal mucosa, parosmia, epistaxis and headache. The deformity of the "saddle" nose, caused by damage and collapse of the nasal cartilage and vomer, occurs in 3.5 to 7% of cases (3,4,5).

Among the differential diagnosis that must be considered, especially in endemic areas, is mucocutaneous leishmaniasis. Other differential diagnoses that should be considered include malignancies, paracoccidiodomycosis, tuberculosis, and sporotrichosis (6).

CASE REPORT

Female patient, 38 years old, with history of periorbital edema with hyperemia and purulent rhinorrhea intermittently for one year before admission. In May 2023, she developed a destructive lesion with nasal septum perforation, sinus erosion and nasolacrimal fistula – figure 1, in addition to bilateral marked periorbital edema, em tongue ulcer and mucositis. A nasal biopsy was performed with the presence of multinucleated giant cells and necrosis, without evidence of well-formed granuloma or vasculitis; PCR for tuberculosis and baar were negative; serologies for leishmaniasis and culture for fungi were negative, and direct mycology of a nasal fragment demonstrated conidia. The patient started treatment for Cutaneomucosal Leishmaniasis and Mucormycosis after infectology consultation, with leishmaniasis being the first diagnostic hypothesis as it is an endemic area for the disease.

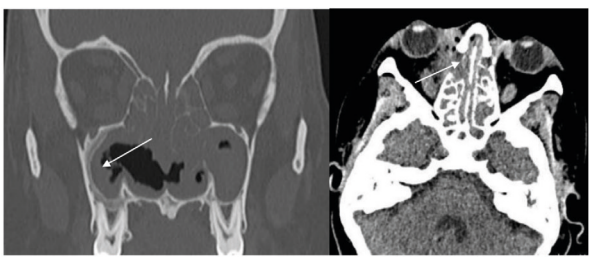

FIGURE 1. ON THE LEFT: MARKED MUCOUS THICKENING OF ALL PARANASAL SINUSES, GREATER ON THE RIGHT WITH SCLEROSIS OF THE BONE WALLS (WHITE ARROW) COMPATIBLE WITH CHRONIC SINUSOPATHY. RIGHT: CONTINUITY SOLUTION IN THE LAMINA PAPYRACEA OF THE ETHYMOID WITH A SMALL COLLECTION NEXT TO THE NASOLACRIMAL DUCT ON THIS SIDE (WHITE ARROW).

The patient developed, while in treatment for leishmaniosis, acute respiratory failure requiring mechanical ventilation, acute renal dysfunction and alveolar hemorrhage, diagnosed by hemoptysis and compatible tomographic image concomitant with a drop in hemoglobin – figure 2.

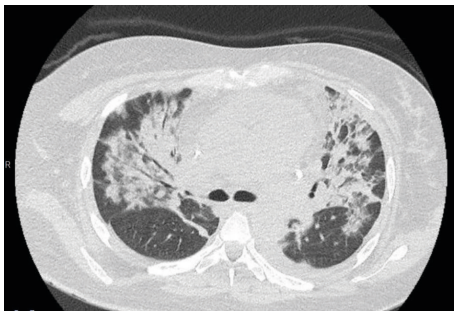

FIGURE 2. CONFLUENT CONSOLIDATIVE OPACITIES IN BOTH LUNGS, PREDOMINATING IN THE CENTRAL REGIONS OF BOTH SIDES. SMALL BILATERAL PLEURAL EFFUSION. IN THIS CASE, ALVEOLAR HEMORRHAGE.

C-ANCA was measured, which resulted positive (1/20); Patient met the 2022 classification criteria for Wegener's Granulomatosis of the American College of Rheumatology at 14 points. The BVAS (Birmingham Vasculitis Activity Score) resulted in a score of 12 points. She started a treatment regimen with methylprednisolone in pulse therapy associated with cyclophosphamide according to the CYCLOPS protocol, with good clinical evolution. The renal function recovered, and the patient was discharged. Control tomography demonstrated scarring lung lesions, with some retractile and fibrotic areas in the lung parenchyma. The patient also kept a collapsed nasal septum, with a saddle nose.

DISCUSSION

Since both GPA and cutaneous mucosal leishmaniasis affect the nasal mucosa and can be difficult to diagnose, it is important to make an accurate diagnosis based on clinical characteristics and supported by laboratory tests (6).

GPA rhinosinusitis can lead to perforation of the nasal septum, saddle nose deformity, serous otitis, hearing loss and nasocutaneous fistulas (7). Suspicion should be greatly increased if there is laboratory detection of antineutrophil cytoplasmic autoantibody (ANCA) (8). Approximately 82 to 94 percent of patients with GPA have a positive ANCA, depending on the severity of the disease (9).

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