SBR 2019
36º CONGRESSO BRASILEIRO DE REUMATOLOGIA
CENTRO DE EVENTOS DO CEARÁ
04 A 07 DE SETEMBRO
FORTALEZA 2019
PROMOÇÃO
70 anos Sociedade Brasileira de Reumatologia
REALIZAÇÃO
SCR SOCIEDADE CEARENSE DE REUMATOLOGIA

GRANULOMATOSIS WITH POLYANGIITIS WITH BILATERAL FACIAL PALSY AND ORAL ULCERS - AN ATYPICAL PRESENTATION

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BACKGROUND

Granulomatosis with Polyangiitis (GPA) is a systemic vasculitis of autoimmune etiology, characterized by the triad: necrotizing granulomatous inflammation of the respiratory tract, disseminated vasculitis and glomerulonephritis. Early diagnosis requires a high level of suspicion, being essential the anatomopathological examination and the presence of antineutrophil cytoplasmic antibodies directed against proteinase 3 (c-ANCA). Among the ANCA - associated vasculitis, peripheral neuropathy is more commonly observed in eosinophilic granulomatosis with polyangiitis (60 – 80%) than in microscopic polyangiitis (40 – 50%) or GPA (20 – 25%). Neuropathy is not usually associated with life threatening manifestations, usually affects motor nerves and is long-lasting.

CASE REPORT

A 34-year-old woman was admitted with severe holocranial headache and a history of pruritus in both ears 6 months ago, followed by otalgia and otorrhea, culminating with the need for drainage. Cranial computed tomography (CT) showed bilateral mastoidopathy, without bone erosion. Submitted to several courses of antibiotic therapy, all unanswered. It evolved with bilateral peripheral facial paralysis, nasal obstruction, odynophagia and two oral ulcers on the hard palate, as well as hematuria. With the suspicion of GPA, tests were requested to confirm the diagnosis: c-ANCA reagent with 1/40 titer, non-reactive p-ANCA, proteinuria and erythrocytic dysmorphism in urine summary,chest CT showed cavitations, areas of atelectasis, and small right pleural effusion. Therapy with intravenous methylprednisolone was performed, resulting in a substantial improvement in neurological deficit and in palatal lesions, in addition to remission of the pulmonary condition.

CONCLUSION

When one of the first clinical manifestations of GPA is of neurological nature, its diagnosis becomes more difficult. Bilateral facial paralysis is a rare manifestation and may signal serious diseases, being mandatory the differential diagnosis. Diagnostic evaluation should focus on signs and symptoms indicative of an underlying systemic vasculitis, although when neuropathy is the initial manifestation of the disease and / or there is no definitive evidence of vasculitis elsewhere, nerve biopsy is necessary for diagnosis. The prompt recognition of these clinical and pathological features is important to better recognize and treat patients with peripheral nerve vasculitis.