Localised presentation of granulomatosis with polyangiitis: The great masquerade

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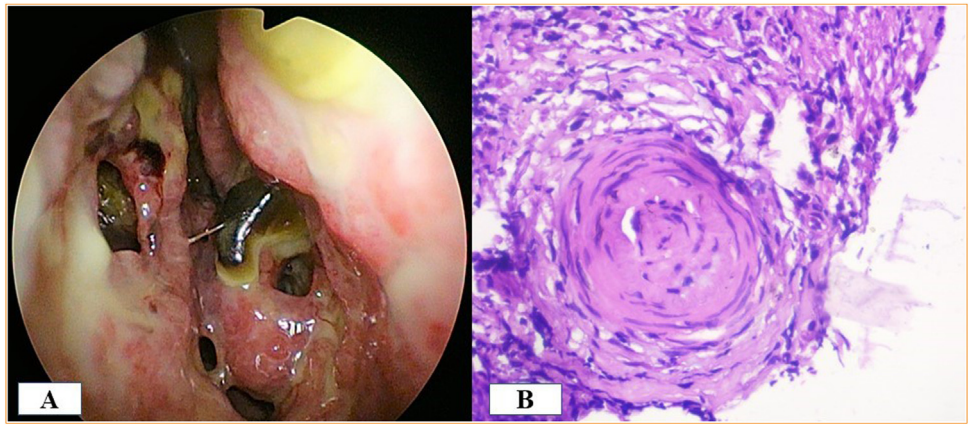

FIGURE 1.

CASE PRESENTATION

A 30-year-old male presented with a history of persistent nasal obstruction for 2 years. There was an associated history of occasional blood-stained discharge. The patient also complained of occasional ear ache and blocked sensation in both the ears without any ear discharge. There was no history of fever or weight loss. Aural examination revealed serous otitis media on both sides. Nasal endoscopy revealed mucopus and crusts in the nasal cavity with congested and granular turbinates. There was a septal perforation in the cartilaginous septum and the edges of the perforation were granular (Fig. 1A). Nasal biopsy taken from the inferior turbinate revealed a mixed inflammatory infiltrate along with leucocytoclastic vasculitis of the medium and small vessels with hematoxyline & eosin staining (Fig. 1B, 40x magnification). The cANCA levels for the patient was positive and the Proteinase (PR3) antibody levels were 60 EU/mL (normal <10 EU/mL). Evaluation of the renal function was unremarkable. HRCT of the chest revealed a small nodule in the left middle lobe.

Granulomatosis with polyangiitis (GPA) is a systemic disease characterised by necrotising vasculitis affecting small to medium sized vessels. The incidence of the condition is 8–10 cases per million and is seen across a wide age distribution without any gender predisposition.[1] It classically involves the pulmonary, renal vasculature and the paranasal sinus. Localised disease can present with only nasal symptoms and may often masquerade other conditions like nasal polyposis, tuberculosis, lymphoma etc. The local presentation can mimic extranodal T-cell lymphoma many times due to the presence of localised destructing lesion. When doubtful, an immunohistochemical analysis of the nasal tissue should be performed. An overdependence on cANCA as the diagnostic test should be avoided as it has been seen that the cANCA levels may be elevated in other conditions as well.[2] It is important to obtain a histopathology sample and also to carry out the specific marker like the PR3 antibody to confirm the diagnosis. Localised disease may have severe disease and uncommon symptoms.[3] Initiation of systemic steroids or intranasal steroids without the definite diagnosis further delay the diagnosis due partial resolution of symptoms.

It is important to actively look for organ involvement in cases presenting with localised symptoms. Treatment of granulomatosis with polyangiitis requires initiation of systemic corticosteroid preferably Prednisolone (1 mg/kg/day) with gradual tapering of doses once a remission is achieved. Methotrexate and azathioprine have been used for maintenance therapy. Cyclophosphamide is added to achieve remission and to prevent the side effects of long-term therapy with steroid. The most recent addition to treatment regimen is rituximab (an anti CD20 inhibitor) which has been found to be effective in severe refractory disease.

DECLARATION OF COMPETING INTEREST

Nil.

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