

ANCA-negative limited Wegener's granulomatosis

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ABSTRACT

A 26-year-old man presented with epistaxis, nasal obstruction and a subcutaneous swelling over the left malar region with radiological evidence of a mass in the right nasal cavity. Histology of the lesions showed necrotizing granuloma with evidence of vasculitis. There was no other systemic involvement and the patient was ANCA-negative. Excellent response to systemic steroid and cyclophosphamide therapy was noted.

KEY WORDS: Subcutaneous lesion, Nasal cavity

INTRODUCTION

Wegener's granulomatosis (WG) is characterized by granulomatous inflammation of the respiratory tract, necrotizing vasculitis affecting small to medium sized arteries and necrotizing glomerulonephritis.¹ Although in its classical form WG is a multisystem disease with protean manifestations, there can actually be a spectrum of clinical manifestations and the disease may present with limited organ involvement.² Detection of cytoplasmic antineutrophilic cytoplasmic antibody (c-ANCA) is a valuable adjunct to the diagnosis of WG, but it may be negative in localized forms of the disease.³ A case of ANCA-negative WG limited to the upper respiratory tract and adjoining skin is reported owing to its rarity.

CASE REPORT

A 26-year-old man from rural West Bengal presented with bloody discharge from the nose with nasal

obstruction and pain, and a reddish swelling over the left cheek for three months. The disease started insidiously with a gradually increasing obstruction of the right nasal cavity and a scanty blood-stained serous discharge followed by crusting. Subsequently, the patient developed redness and a gradually increasing swelling over the left malar area which was mildly painful. He did not have any concomitant general illness or any symptoms referable to any other system. Apart from recurrent upper respiratory tract infections, the patient had enjoyed excellent health prior to the illness. No family history of a similar or related illness was reported.

On clinical examination, an indurated subcutaneous lesion with ill defined border over the left malar region extending to the left infraorbital area was seen (Figure 1). It showed a smooth dusky red surface and was tender on palpation. The regional lymph nodes were not enlarged. An otorhinological consultation revealed a granulomatous mass involving the

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Figure 1: A diffuse swelling on the left malar region

osteomeatal complex region of the right nasal cavity.

Routine hemogram and urinalysis were within normal limits. Serum urea, creatinine and glucose estimations revealed no abnormality. An X-ray of the chest was normal, while one of the paranasal sinuses showed a dense homogeneous opacity in the right nasal cavity displacing the septum to the opposite side. Right antral haziness was present. A CT scan of the paranasal sinuses and nasopharynx showed a hypodense shadow in the right nasal cavity extending laterally into the right maxillary antrum (Figure 2). A bony erosion was seen in the inferior part of the lateral wall of the right nasal cavity. Mild mucosal thickening was noted in the right maxillary antrum, ethmoid sinus and the nasal cavity. An abdominal ultrasonography failed to reveal any

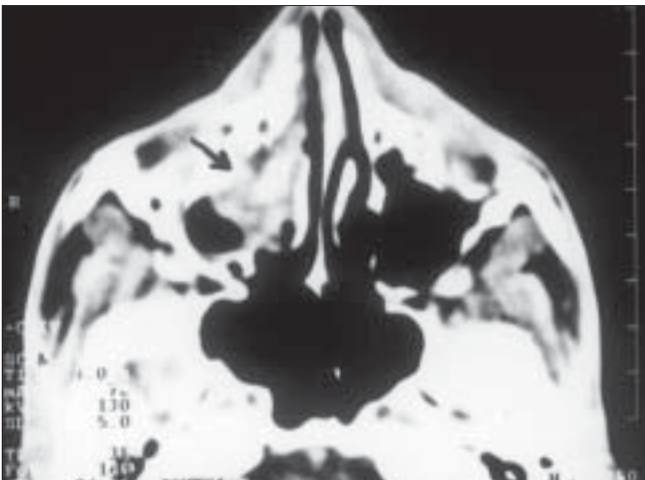


Figure 2: CT-scan of paranasal sinus showing a lesion (arrow) in the right nasal cavity extending into the right maxillary antrum

discernable abnormality.

Histopathological examination of biopsy stained with H & E stain showed features of granulomatous infiltration with foci of necrosis. Evidence of vasculitis with fibrinoid degeneration was also noted in some sections. No fungal elements were seen. An ANCA screen was negative. .

The patient was put on a daily combination regimen of prednisolone 1 mg/kg body weight and cyclophosphamide 2 mg/kg body weight orally which resulted in dramatic improvement of symptoms with rapid clearing of the nasal obstruction and dissolution of the cutaneous lesion within a period of eight weeks.

DISCUSSION

Since Wegener's original description of the disease in 1936, the condition bearing his name has been well characterized. In its complete form, the disease shows multiple organ involvement with necrotizing vasculitis and granuloma; the commonest organ systems affected are the upper and lower respiratory tracts and the kidneys. Other organs commonly involved are the skin, joints, eyes and the nervous system.² Once a rapidly fatal disease, the course of WG has been dramatically improved by daily treatment with cyclophosphamide and glucocorticoids. Nonetheless, the disease and its treatment related morbidity is often profound.

Slightly more common in males, the syndrome typically presents with upper airway disease; sinusitis with nasal obstruction and discharge was the presenting sign in 67% of patients in a large series.⁴ Cutaneous manifestations are reported in 14-50% of cases in different series and may be the presenting feature in a significant proportion of cases.⁵⁻⁷ Palpable purpura was found to be the commonest cutaneous manifestation in a large series, but papular, plaque-like, and ulcerated lesions, vesicles, urticarial lesions, subcutaneous nodules and panniculitis have all been described.⁵⁻⁷ The histopathologic findings of WG, viz. necrotizing vasculitis, granulomatous vasculitis and palisading granuloma, may be seen in the cutaneous lesions in about 25% of cases.⁷

A positive c-ANCA test is of great value in confirming a clinicopathological diagnosis of WG, but these autoantibodies may be absent in a proportion of cases, particularly in the limited variety of the disease. The sensitivity of this test was found to be 67% by immunofluorescence and 60% by ELISA for patients with active local or regional symptomatology.³ Hence, although a positive c-ANCA test is adjunctive evidence for the diagnosis of Wegener granulomatosis, the result must be viewed in the context of the patient's clinical picture and disease activity.⁴

Our patient had a slowly growing subcutaneous mass with nasal obstruction and discharge as the only manifestations of the disease. The characteristic upper respiratory involvement, histopathologic features and the dramatic response to glucocorticoid and cyclophosphamide therapy helped us to confirm the diagnosis.

A study comparing the classical and limited varieties of the disease found that the groups shared many features, particularly their requirement for immunosuppressive therapy, since WG causes major tissue destruction regardless of whether it is a localized or a widespread process.⁸ At the immunopathological level, the two groups appear to be part of a single disease spectrum. Importantly, the non-renal WG group

may change the pattern of their disease to involve the kidney.⁸ Long-term follow-up of such patients is therefore essential.

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