Granulomatosis with Polyangiitis Presented as Isolated Destructive Nasal Mass – Case Report

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Granulomatosis with polyangiitis (GPA), previously known as Wegener’s Granulomatosis, is a relatively rare autoimmune disease. It commonly presents with multiple organ symptoms. Isolated nasal manifestation is a rare form of presentation. Herein, we describe the case of a 19-year-old girl who presented with progressive nasal obstruction. Physical examination revealed a nasal mass involving the nasal septum obstructing the nasal cavity, which appeared on the CT-scan as a destructive mass affecting the nasal septum.

INTRODUCTION
Granulomatosis with polyangiitis (GPA) is an autoimmune multi-systemic disease of rare incidence, characterized by necrotizing granulomatous inflammation and pauci-immune vasculitis in small- and medium-sized blood vessels. It has a relatively rare incidence (3/100,000). It usually presents with pulmonary, renal and upper airway manifestations. Most of the cases of generalized disease are associated with the presence of cytoplasmic anti-neutrophil cytoplasmic antibodies (c-ANCA). Some cases presented as a limited disease, in which the c-ANCA is usually negative. It is not uncommon for patients with GPA to have upper respiratory manifestations of the disease, but being presented with isolated nasal mass without any other disease manifestation is rare and may cause the patient to roam among different clinics and take various medications without definite diagnosis.

CASE REPORT
A 19-year-old girl presented with progressive nasal obstruction for 2 months, associated with headache and decreased sense of smell. She had no history of epistaxis, nasal or post nasal discharge, convulsions, cough, chest pain, or hematuria. On presentation, there was a right side nasal mass destructing the septal cartilaginous and bony parts with liquefying necrosis and gray-brown necrotic material obstructing the nasal airway (Fig 1). Initial laboratory workup showed blood urea nitrogen of 27 mg/dL, creatinine of 0.5 mg/dL, erythrocyte sedimentation rate of 90 mm/h, and normal urine analysis. Chest radiograph was normal as well (Fig 2). Computed tomography scan showed a destructive mass in the right nasal cavity and involving the nasal septum, with mild contrast enhancement (Fig 3). Then, an incisional biopsy was obtained, which showed necrotizing lesion with vague necrotizing granulomata and focal vascular changes (Fig 4). Other samples sent for microbiology lab for fungi and mycobacterium were all negative. The c-ANCA was requested based on this finding and came to be positive. Hence, the diagnosis of granulomatosis with polyangiitis was confirmed and the patient was referred to rheumatology team and showed a good response to cyclophosphamide and hydrocortisone.

KEY WORDS: nasal mass, nasal septum, vasculitis, wegener’s granuloma

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DISCUSSION

GPA, previously known as Wegener’s Granulomatosis, was first described by the German pathologist Friedrich Wegener in 1936. In 1931, two patients died from prolonged sepsis with inflammation of blood vessels scattered throughout the body. In 1936, Wegener first described a distinct syndrome in three patients found to have necrotizing granulomas involving the upper and lower respiratory tract. In 1954, seven more patients were described, resulting in a definite criteria. It may occur at any age, although patients typically present in the fifth decade. Patients with GPA usually present with chronic, non-specific constitutional complaints and recurrent respiratory infection in an adult. Therefore, they commonly

![Fig 1](image1.jpg)

**Fig 1:** Endoscopic nasal cavity examination showed a destructive nasal mass involving the septum cartilaginous and bony parts with liquefying necrosis and gray-brown necrotic material.

![Fig 2](image2.jpg)

**Fig 2:** Chest X-ray was normal.

![Fig 3](image3.jpg)

**Fig 3:** Computed tomography scan showed a destructive mass in the right nasal cavity and destructing the anterior part of the nasal septum, with mild contrast enhancement.
present after being evaluated in different clinics where they seek help without definite diagnosis\textsuperscript{7}. It can manifest in two forms: limited disease without renal manifestations and generalized disease with renal involvement. Also, it can present with involvement of eyes, ENT, musculoskeletal, renal, neurological, cutaneous and cardiac symptoms\textsuperscript{8}. According to the modified American College of Rheumatology Criteria for the classification of Wegener’s Granulomatosis, for diagnosis of GPA, patients must have at least two of the following five criteria:

1. Nasal or oral inflammation
2. Abnormal chest radiograph
3. Active urine sediment
4. Granulomatous inflammation and/or necrotizing vasculitis on tissue biopsy
5. Positive enzyme immunoassay for antibodies to serum proteinase 3\textsuperscript{9}

In our patient, the presentation was unusual, in the form of isolated nasal mass with nasal obstruction, without any other disease manifestations. However, the histopathological description is classical for GPA, and therefore c-ANCA was requested and came out positive as further confirmatory clue for the diagnosis.

CONCLUSION

GPA should be considered as one of the differential diagnoses when a patient presents with nasal cavity mass. Early detection of such cases will decrease disease morbidity and eliminate unnecessary intervention and inappropriate management. The workup should include biopsy as well as c-ANCA whenever GPA is to be excluded.

REFERENCES

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