Wegener Granulomatosis: Operated Nasal Septal Perforation

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Abstract

Wegener's granulomatosis is an idiopathic necrotizing granulatous vasculitis that can hold small and medium arteries, upper respiratory tract, lung and kidney involvement. It's an ANCA associated vasculitis. c-ANCA positivity is considered very sensitive. In our case, a 40-year-old female patient was admitted to our hospital with complaints of nasal bleeding and crusting. The patient was operated due to idiopathic nasal septal perforation, was seen saddle nose deformity and 3x4 cm in size perforation in anterior septum and 1x2 cm in size perforation in posterior septum. Nasal biopsies of the patient with C-ANCA positivity were taken and the result of pathology came in harmony with Wegener's granulomatosis. The patient was directed to the related clinic for the treatment of Wegener's granulomatosis. Wegener Granulomatosis, which enters the diagnosis of idiopathic nasal septal perforation, is a disease that should be kept in mind.

Keywords: c-ANCA, septal perforation, wegener's granulomatosis, vasculitis

INTRODUCTION

Wegener granulomatosis is a systemic inflammatory disease of the small and medium arteries; it usually has an unknown etiology. It is a type of vasculitis that produces systemic necrotizing granulomas by holding the upper respiratory tract, lungs and kidney (1). Its incidence in males and females is approximately the same and usually presents during the sixth and seventh decades of life (2). c-ANCA is used to diagnose Wegener granulomatosis. Here, we present the case of a patient with Wegener granulomatosis who was operated owing to idiopathic nasal septal perforation and continuous complaints.

CASE PRESENTATION

A 40-year-old female patient was admitted to our outpatient clinic with complaints of nasal obstruction, crusting, and bleeding. Approximately 3 years ago, she was operated in another hospital because of idiopathic nasal septal perforation; saddle nose deformity was also observed. Physical examination revealed two nasal septal perforations in the nasal septum: 3×2 cm in the anterior and 1×2 in the posterior and hemorrhagic crust in the nasal passage (Figure 1. a, b). The patient was referred to the Rheumatology outpatient clinic for further examination and diagnosis. The c-ANCA test was positive, and the patient demonstrated a high sedimentation rate. Biopsy was performed from the posterior edge of the perforation and the lower cone. Biopsy results revealed inflammatory granulation in accordance with Wegener granulomatosis, and the patient was referred to the Rheumatology outpatient clinic for treatment.

Written informed consent was obtained from the patient.

DISCUSSION

Wegener granulomatosis is a rare idiopathic vasculitis of the small and medium vessels; it presents with necrotizing granulomatous inflammation of the glomerulonephritis and respiratory tract (1). It is estimated to affect 3/100,000 individuals (2). Nongranulomatous and granulomatous inflammatory changes may occur histologically be seen together. Although it occurs at all ages, it is most frequent in the sixth and seventh decades of life (3). Symptoms related to Wegener granulomatosis occur due to the involvement of the upper respiratory tract, lungs, and kidneys. Patients most commonly present with symptoms related to the lungs. Approximately 80%-95% of patients with Wegener granulomatosis initially present with sinonasal symptoms (4). These may include nasal congestion, malodorous discharge, recurrent epistaxis, nasal crusting, and septal perforation (4). The rate of septal perforation in such patients is reportedly 38.7% (5).

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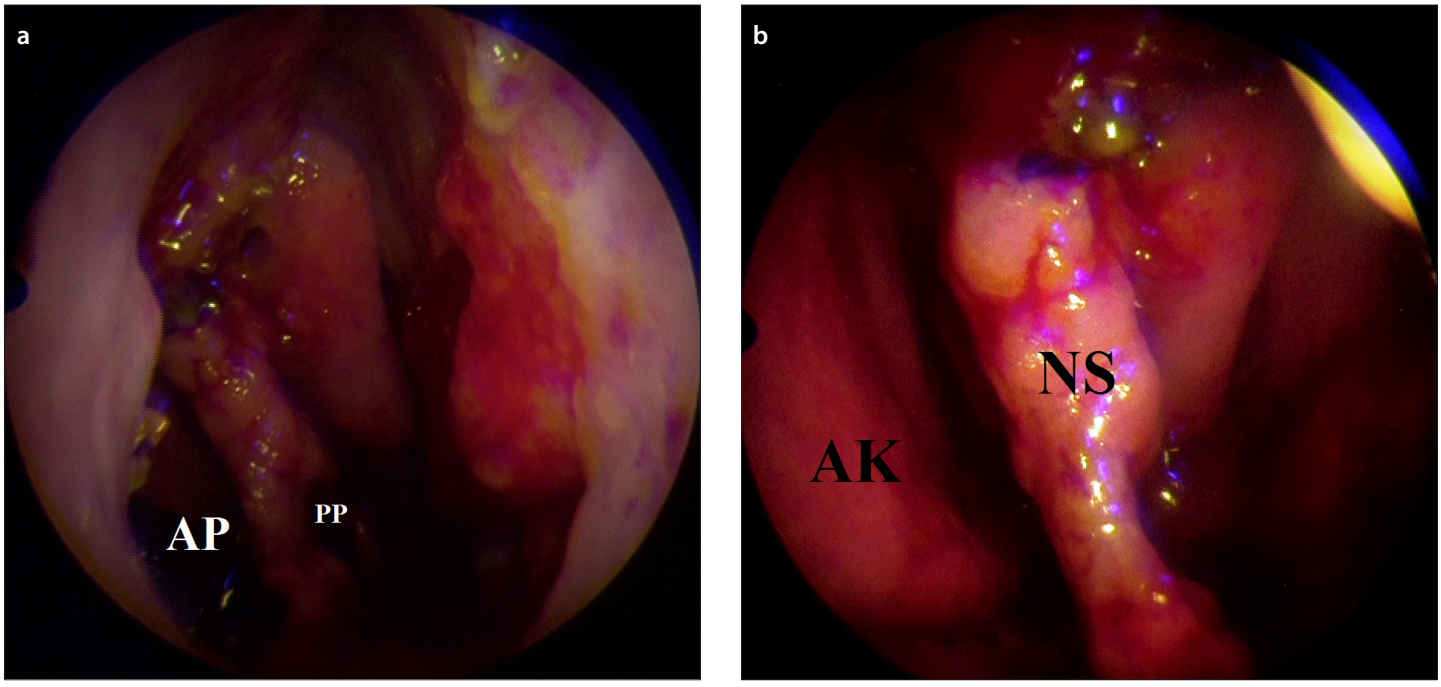

Figure 1. a, b. Nasal passages and anterior and posterior nasal septal perforation
AP: anterior perforation, PP: posterior perforation, AK: lower concha, NS: nasal septum

Patients with Wegener granulomatosis have a history of nasal obstruction, nasal crusting, and recurrent nosebleeds. Erythematous appearance with the presence of nasal crust, septal perforation, and granulation tissue may be observed during examinations (6). The c-ANCA test was considered both sensitive and specific in the diagnosis (7). The c-ANCA test is detected in 80% of patients with systemic disease (8). Rheumatoid factor is increased in 50% of patients (9) and may lead to mild renal changes, such as urinary sediment abnormalities and renal pathologies (e.g., acute kidney injury, etc.) requiring renal replacement therapy. Proteinuria may be present in 57% of patients (10). Leukocytosis, anemia, thrombocytosis are nonspecific in Wegener granulomatosis. There may be an increase in the IgE level owing to elevations in the sedimentation rate and CRP level (9).

Imaging modalities are often more diagnostic for lung and renal involvement. Computed tomography as well as sinonasal tract, sclerosing osteitis, and bone structure thinning findings can be observed (11). Suspicious nasal lesions may be biopsied; however, nonspecific results, such as inflammatory granulation, may occur.

Immunosuppressant therapy is used in the treatment. Systemic corticosteroids, cyclophosphamide, rituximab, and azithiopyrin can be used (12). Nasal irrigation and moisturizing nasal ointments can be used in patients with sinonasal involvement. Endoscopic sinus surgery can be performed in cases resistant to medical treatment.

CONCLUSION

Wegener granulomatosis is a disease that should be considered in the differential diagnosis of patients with nasal symptoms, such as nasal bleeding and crusting. Wegener granulomatosis should definitely be considered in patients with long-term nasal complaints despite medical therapy.

Informed Consent: Written informed consent was obtained from the patient who participated in this case.

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