

Unusual Presentation of Granulomatosis with Polyangiitis (Wegener's Granulomatosis)

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ABSTRACT

Objective

To present a case of unusual presentations of granulomatosis with polyangiitis (Wegener's granulomatosis) with parotid swelling and intractable otitis externa.

Materials And Method

A 22-year-old male patient presented with left sided otitis externa with left sided parotid swelling and bilateral nasal obstruction for last two weeks. CT scan of paranasal sinuses showed homogenous mass in both maxillary antra and nasal cavities. FNAC from the parotid swelling was suggestive of granulomatous disease and endoscopic biopsy from the nasal mass showed features of granulomatosis with polyangiitis (Wegener's granulomatosis). CT scan of chest revealed multiple cysts within the lung parenchyma; urine examination showed RBC and pus cells. Renal biopsy showed focal segmental glomerulonephritis. c-ANCA was highly positive and thus the diagnosis of Wegener's granulomatosis was made.

Result

Patient received cyclophosphamide and prednisolone immunosuppressive therapy for one year; which showed marked clinical improvement.

Conclusion

Granulomatosis with polyangiitis (Wegener's granulomatosis) is not an uncommon entity for otolaryngologists. Its usual presentation mimics chronic rhinosinusitis; but presentations like otitis externa and parotid swelling are rare for this disease.

Keywords

Granulomatosis with Polyangiitis; Otitis Externa; Nasal Obstruction; Parotid Gland; Endoscopy; Biopsy; Glomerulonephritis; Antibodies, Antineutrophil Cytoplasmic.

Granulomatosis with polyangiitis (Wegener's granulomatosis) is a distinct clinicopathological entity characterised by granulomatous vasculitis of the upper and lower respiratory tracts with glomerulonephritis. It is extremely rare in blacks as compared to whites. Usual presentations of Granulomatosis with polyangiitis (Wegener's granulomatosis) are facial pain over the paranasal sinuses, purulent or blood mixed nasal discharge, proteinuria, and haematuria with or without cough, haemoptysis due to cavitary infiltrates in lungs. In addition to the classic triad of above symptoms, virtually any organ can be involved with vasculitis, granuloma or both. Histopathological hallmark of this disease is vasculitis of small arteries and veins with intravascular and extravascular granuloma formation. Granulomatosis with polyangiitis (Wegener's granulomatosis) presenting with otitis externa and parotid swelling is extremely rare. Biopsy from lung tissue, upper airway and kidney, high c-ANCA and clinical

findings establish the diagnosis of Granulomatosis with polyangiitis (Wegener's granulomatosis). Oral steroid and immunosuppressive drugs have proven therapeutic role in this disease.

Case Report

A 22 year-old young man from eastern part of rural India presented with excruciating left earache, globular mass in left infra-auricular parotid region and nasal obstruction for last 2 weeks. He had also history of purulent and occasionally blood mixed nasal discharge for the same

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duration. Conservative medical treatment did not relieve the symptoms. Otomicroscopy revealed that external auditory canal is completely occluded due to severe oedema of the canal wall skin. Anterior rhinoscopy reveals red congested nasal mucosa, ulcerations over septal mucosa.

CT scan of nose and paranasal sinuses (Fig. 1) revealed a homogenous mass in both maxillary antra and nasal cavities. Nasal endoscopy revealed mass in both nasal cavities; biopsy taken from the mass and endoscopic debridement of the mass was done with the help of a microdebrider. Histopathological examination of the mass suggested necrotising vasculitis and granuloma formation of small vessels (Fig. 2). His routine blood examination revealed anaemia (Hb. 8 gm %), neutrophilic leukocytosis (total count- 23000/



Fig. 1 Coronal CT scan of PNS showing a homogenous opacity in both nasal cavities, maxillary antra, anterior and posterior ethmoidal cells

ml, neutrophils-71%), thrombocytosis (4 lakh/ml) and elevated E.S.R (60 mm/1st hr). His plain skiagram of chest was within normal limits. Urine examination revealed microscopic hematuria and proteinuria. CT scan of chest showed cavitory lesions present in both lung fields (Fig. 3). c-ANCA (Anti Neutrophil Cytoplasmic Antibody) was highly positive (42 Units; Normal <6 Units). CT-guided renal biopsy was performed which showed focal segmental necrotising glomerulonephritis (Fig. 4).

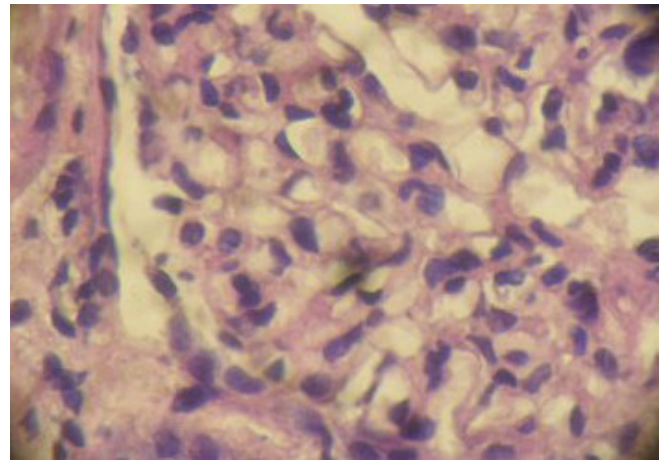


Fig. 2 Histopathology of the mass from nasal cavity showing vasculitis, granuloma formation and mononuclear cell infiltrate (H&E, 400X)

Based on these reports, the patient was diagnosed as a case of Granulomatosis with polyangitis (Wegener's granulomatosis) and prompt treatment with oral Cyclophosphamide (2 mg/kg/day) along with Prednisolone (1 mg/kg/day) started. But he developed skin lesions around lips and lower part of nose (Herpes labialis) with severe abdominal pain. Immunosuppressive therapy had to be discontinued for 2 weeks and conservative treatment was started and these symptoms subsided. A second course of Cyclophosphamide and Prednisolone was started subsequently. He is on immunosuppressive therapy for last one year and showing marked clinical improvement.

Discussion

Granulomatosis with polyangitis (Wegener's granulomatosis) is a distinct clinical entity affecting predominantly Caucasian population with an annual incidence of 3 per 1,00,000. It is extremely rare in blacks as compared to whites. Mean age of occurrence is 41 years (range 9-78 years) with equal sex preponderance. Heinz Klinger first described this disease entity in 1931 and Friedrich Wegener gave detailed description in 1936 and 1939. Churg and Godman carried out the clinicopathological study regarding this disease entity in 1950s and the treatment protocol with cyclophosphamide



Fig. 3 HRCT of chest showing cavitary lesion on left lung parenchyma (white arrow)

and prednisolone was postulated by Wolf and Fauci in 1973.

Granulomatosis with polyangitis (Wegener's granulomatosis) is characterized histologically by necrotising vasculitis of small arteries and veins. The most commonly affected organs are the upper respiratory tract, the lungs and the kidneys. One third of the patients present with locoregional form of this disease,¹ which lasts from few months to maximum few years before it progresses to generalized form, with mean survival of few months if left untreated.² The most common cause of death is renal insufficiency and/or uncontrollable sepsis.³

In Granulomatosis with polyangitis (Wegener's granulomatosis), ear is frequently affected. Otagia, aural discharge, aural polyp, deafness may be the first symptom to bring the patient to medical attention. The otological manifestation of this disease ranges from 19% to 45%. De Remee et al. reported a case series of 50 patients of Wegener's granulomatosis in which the ear was the most frequently affected site followed by lung and kidney.⁴ D' Cruz et al reported 22 cases in which 11 had otalgia as their presenting complaint and 3 had otitis externa.⁵ Common otological symptoms of Wegener's granulomatosis are serous otitis media (90%), 33% of that is bilateral, followed by sensorineural hearing loss (43%), and chronic otitis media (24%).

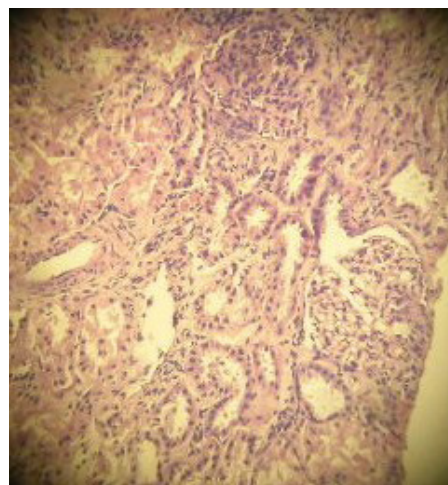


Fig. 4 Histopathology of kidney showing vasculitis (H&E, 100X)

Serous otitis media is the most common cause for conductive deafness whereas cochlear vessel vasculitis, immune complex deposition and/or granulomatous involvement result in sensory neural hearing loss.⁶ Another important but rare otological manifestation is facial nerve palsy, which may be due to cranial neuropathy or due to compression within temporal bone.⁶ Several authors describe a 'limited' type Wegener's granulomatosis, which presents with otological manifestations only.⁷

Parotid gland or any other salivary gland enlargement, as found in this case, as a presenting feature of Granulomatosis with polyangitis (Wegener's granulomatosis) is even rare. Granulomatosis with polyangitis (Wegener's granulomatosis) may present as unilateral parotid gland enlargement,⁸ bilateral parotid and submandibular gland enlargement or a parotid abscess.⁹

The diagnosis of Granulomatosis with polyangitis (Wegener's granulomatosis) should be based on biopsy from suspected lesions of the nose, nasopharynx, kidney, or lungs.¹⁰ Abnormal tissues from external canal, middle ear or mastoid cavity most often does not yield diagnosis.¹

The estimation of c-ANCA titre in the serum with immunofluorescence technique provides a reliable diagnosis, though it is contributory to tissue biopsy but not an alternative to it. Cytoplasmic pattern (c-ANCA)

is highly specific for Granulomatosis with polyangitis (Wegener's granulomatosis), whereas perinuclear pattern (p-ANCA) correlates with other types of vasculitis. Estimation of ANCA can also be done by immunosorbent assay with myeloperoxidase and proteinase³ roughly correspond to p-ANCA and c-ANCA respectively.¹⁰ In cases of locoregional involvement, the sensitivity of this method is 60%, whereas in the generalized form it is 93%.¹ There are reported cases in which estimation of c-ANCA was negative in the beginning and became positive in the course of the disease.¹¹

The treatment of Granulomatosis with polyangitis (Wegener's granulomatosis) with cyclophosphamide and prednisolone provides a 70% to 85% remission rate to an otherwise fatal disease.¹²

Close follow-up and expectation for complete cure should be the routine if there is no histological proof of the disease.¹³ B cell depletion therapy using rituximab, a chimeric anti-CD20 monoclonal antibody, has been shown to be effective for certain refractory Granulomatosis with polyangitis (Wegener's granulomatosis), which causes diminished granuloma and reduced ANCA titers, allowing steroids to be tapered. No adverse effects were detected.¹⁴

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