Wegener's Granulomatosis Presenting as Nasal Polyp

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Abstract

Wegener's granulomatosis (WG) is characterised by aseptic granulomatous inflammation and vasculitis which classically affects kidneys, upper and lower respiratory tract. Although majority of patients present with symptoms of head and neck region, rarely this region is the sole site of involvement. The interpretation of biopsy for the diagnosis of this disease needs precision. We report a case who presented clinically as nasal polyp. However, the microscopy confirmed the diagnosis of WG.

Key Words

Wegener's granuloma, Vasculitis, Midline granuloma.

Case Report

A ten years old boy presented with epistaxis, pain and mild swelling left side of nose of one month duration. There was also history of foul smelling yellowish discharge with difficulty in breathing with no significant past history. General physical examination was unremarkable. Per speculum examination revealed a...
polypoidal ulcerated mass in posterior part of left nostril extending to nasopharynx. It was covered with whitish exudate.

A provisional diagnosis of chronic inflammatory polyp was made with a differential diagnosis of rhabdomyosarcoma. Multiple biopsies were taken. X-ray chest, routine urine analysis, renal function tests and ultrasound of abdomen were within normal limits.

The CT scan of the nasopharynx and nasal cavity showed a mass in left nasal cavity destroying the medial and anterolateral wall of left maxillary sinus and antrum. The pterygoid plate was intact. The mass was seen extending to left cheek (Fig. 1).

Pathological Findings

The microscopic examination revealed fragments lined partly by respiratory epithelium with underlying tissue exhibiting focal areas of fibrinoid necrosis as well as necrosis of vessel walls (Fig. 2, 3). Granulomatous inflammation with giant cells was seen around many vessels (Fig. 4). There was no evidence of malignancy. No atypical lymphoid cells were seen. Other granuloma producing conditions were excluded by special stains.

![Fig. 1. CT Scan showing mass in left nasal cavity extending into left cheek.](image)

![Fig. 2. Low power view showing tiny area of fibrinoid necrosis (100x).](image)

![Fig. 3. High power view showing fibrinoid necrosis of vessel wall and granuloma (400x).](image)

![Fig. 4. High power view showing granulomatous destruction of vessel wall with giant cell reaction (400x).](image)
Wegener's Granulomatosis is a clinicopathological disease which requires fulfillment of both clinical and pathological criteria to establish the diagnosis. All patients must demonstrate the classical triad of disease. Also all cases may not demonstrate classical pathological triad for which head and neck biopsies, a criteria for diagnosis has been suggested and is applied where infectious causes of midline granulomatous diseases are excluded [2].

Ideally all three major pathological criteria should be present in isolated case of head and neck involvement for its demonstration, multiple sections to the extent of consumption of whole of the block may be considered. Special stains should be done in all cases. Major entities which are to be distinguished pathologically from WG include granulomatous infections, lymphomatoid granulomatosis, idiopathic midline granuloma, sarcoidosis and foreign body reaction [2-4]. The organisms which are likely to be encountered in head and neck regions are those which cause blastomycosis, histoplasmosis, cryptococcosis and tuberculosis [2, 4, 6].

In WG, the patients have raised ESR, anemia, eosinophilia and mild hypergammaglobulinemia (Ig A), and increase in rheumatoid factor and positive antineutrophil cytoplasmic antibodies—ANCA (1). In our case, the involvement of kidneys and lungs was not seen. A typical or malignant cell was seen on microscopic examination. No special stain for fungus or acid fast bacterium was positive.

Ideally, ANCA should be performed for diagnosis and estimating the activity of disease. Major antibody is directed against proteinase-3, a neutrophilic serine protease which can be seen on indirect immunoflorescence in the cytoplasm of neutrophils. ANCA is positive in 67% patients with active limited disease and approaches 96% if disease is generalized. The test is helpful in differentiating other conditions mimicking WG clinically [2, 5].

Untreated, the course of disease is malignant and 80% patients die within one year. Immunosuppression with cyclophosphamide improves survival in 90% patients. The presence of immune complex in glomeruli and vessel wall and granulomas in upper respiratory tract and lungs suggest some form of hypersensitivity reaction possibly to an inhaled infectious or other environmental agent. Highest relapse rate has been seen in Staph aureus nasal carriers. The reason for this is unknown [1-3].

References