ATHOLOGICAL DIAGNOSIS

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.

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ener's Granulomatosis (WG) is a rare disease, e incidence of which is not known (1). Most of the s are above 40 years of age and mostly present volvement of upper and lower respiratory tract neys. Other organs involved are eyes, ear and ity including salivary glands, but the incidence is w (2,3). The characteristic microscopic features

- Acute necrotizing granulomas of upper and lower respiratory tract.
- Focal necrotizing vasculitis of medium and small vessels.
- Renal disease in the form of focal and diffuse necrotizing glomerulonephritis.

Some patients of WG who do not manifest the full triad are labelled as "Limited" WG (1). About 15% of the patients show involvement of respiratory tract only and present with rhinorrhea, bloody or purulent nasal discharge, epistaxis, anosmia, mucosal ulcers and sinus pain. Very rarely, swelling or mass is observed (2).

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

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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).

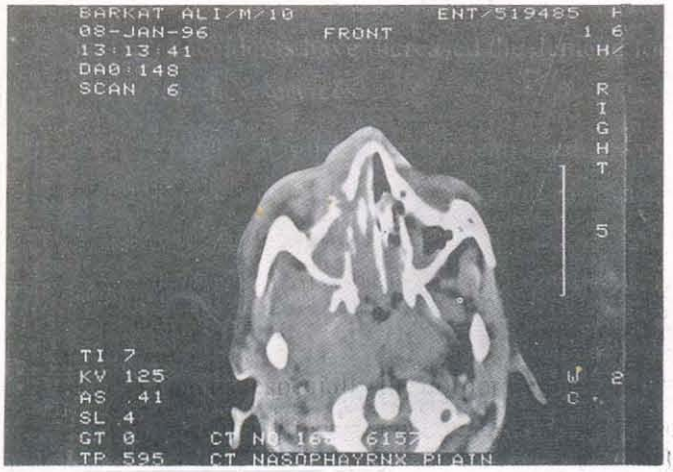

Fig. 1. CT Scan showing mass in left nasal cavity extending into left cheek.

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.

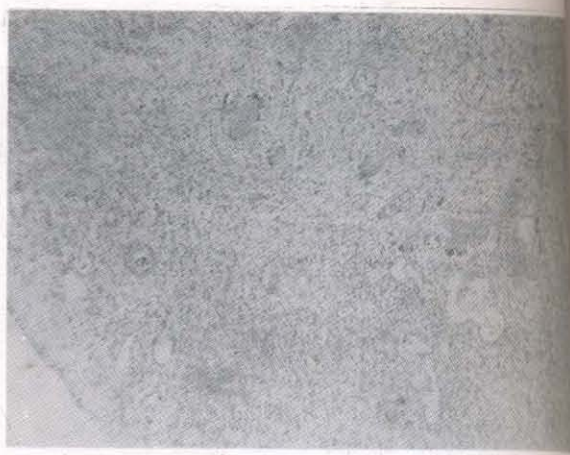

Fig. 2. Low power view showing tiny area of fibrin necrosis (100x).

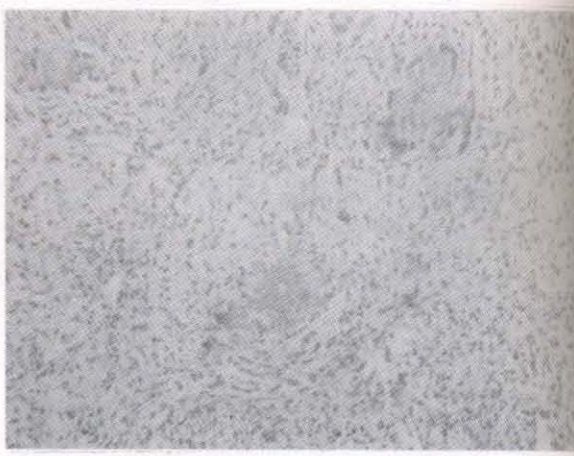

Fig. 3. High power view showing fibrinoid necrosis of ve wall and granuloma (400x).

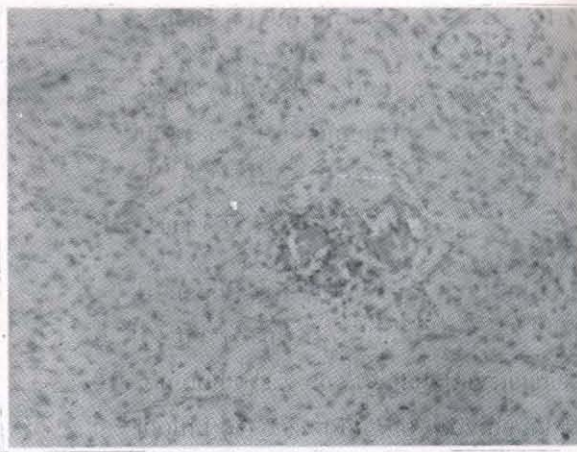

Fig. 4. High power view showing granulomatous destruction of vessel wall with giant cell reaction (400x).

cussion

egener's Granulomatosis is a clinico-pathological y which requires fulfillment of both clinical and ological criteria to establish the diagnosis. All patients t demonstrate the classical triad of disease. Also all sies may not demonstrate classical pathological triad granulomatous inflammation, necrosis and vasculitis In head and neck biopsies, a criteria for diagnosis been suggested and is applied where infectious causes midline granulomatous diseases are excluded (2). ally all three major pathological criteria should be ent in isolated case of head and neck involvement for its demonstration, multiple sections to the extent consumption of whole of the block may be considered. cial strains should be done in all cases. Major entities ich are to be distinguished pathologically from WG clude granulomatous infections, lymphomatoid nulomatosis, idiopathic midline granuloma, sarcoidosis foreign body reaction (2–4). The organisms which likely to be encountered in head and neck regions are one which cause blastomycosis, histoplasmosis, rotrichosis and tuberculosis (2, 4, 6).

In WG, the patients have raised ESR, anemia, cocytosis and mild hypergammaglobulinemia (Ig A), ld increase in rheumatoid factor and positive anti-utrophilic cytoplasmic antibodies–ANCA (1). In our se, the involvement of kidneys and lungs was not seen. atypical or malignant cell was seen on microscopic amination. No special stain for fungus or cobacterium 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 immunoflourescence 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 is differentiating other conditions mimicking WG clinically (2, 5).

Untreated, the course of disease is malignant and 80% patients die within one year. Immunosupression 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).

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