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Wegeners Granulomatosis : A Rare Case Presentation

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ABSTRACT

Wegener's granulomatosis (WG), the most common of the pulmonary vasculitides, typically involving the upper respiratory tract (e.g., sinuses, ears, nasopharynx, oropharynx, trachea), lower respiratory tract (bronchi and lung) and kidney, with varying degrees of disseminated vasculitis commonly known as „Wegener's triad". We report here a case of a 22 year , old case of wegeners granulomatosis.

KEYWORDS :

INTRODUCTION

Wegener's granulomatosis is a systemic vasculitis that primarily involves the upper and lower respiratory tracts and kidneys. Pulmonary Wegener's granulomatosis can present with multifocal lung involvement or solitary lung lesions with no evidence of extrapulmonary disease. Diagnosing Wegener's granulomatosis on the basis of cytological material obtained from fine needle aspiration or sputum may present a challenging problem to the pathologist. A wrong diagnosis may lead to inappropriate treatment for the patient.In practical terms the demonstration of one or more of these in combination with positive serological evidence is now generally accepted as fulfilling the criteria[2]. Although in its classical form Wegener's granulomatosis is a multisystem disease, there can actually be a spectrum of clinical manifestation and the disease may present with limited organ involvement[3]. In so called limited Wegener's granulomatosis, there is no kidney disease and no evidence of systemic vasculitis[1]. Where the clinical findings are typical or consistent and there is a positive c-ANCA, tissue may not be necessary to establish the diagnosis[1]. There are no absolutely diagnostic pathological features of Wegener's granulomatosis[1]. A transbronchial lung biopsy may occasionally support diagnosis, but more tissue from thoroscopic or open lung biopsy may be required if evidence elsewhere is not supportive[1].

Case report

A 22 year old male patient who presented with cough with expectoration, hemoptysis, epistaxis, chest pain and severe headache since 1 month

On admission, the patient was febrile, pulse was 120 beats/min, blood pressure was 110/90 mm of Hg, RR was 18/ min. There were no edema, cyanosis or clubbing, pallor, icterus. A small lymph node measuring 2 × 1 cm was noted in the axilla.

The patient had bilateral red eyes with burning sensation and dryness.

The patient was using accessory muscles of respiration. There was intercostal retraction and tenderness over the chest. Occasional b/l crepts were present. Cardiac examination was normal .Per abdomen examination revealed no organomegaly

CBC

Hb	9.7
WBC	10,120
RBC	4.07
Platelet count	4.96

Urine R/M

Urine appearance	Slightly hazy
protein	Present +
Occult blood	Present ++
Red blood cells	4-6 /HPF

Peripheral smear showed mild hypochromic microcytic anemia with relative neutrophilic leucocytosis with shift to left maturation (myeloid leukemoid reaction) and marked thrombocytosis

CXR showed multiple ,

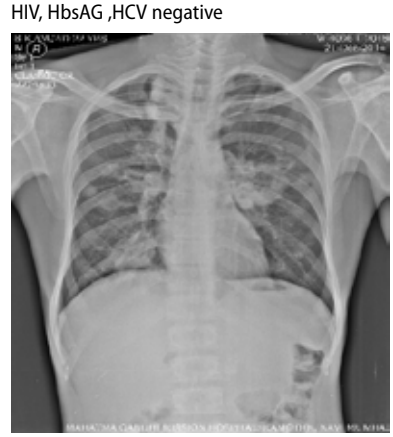
Sputum AFB negative

Sputum culure senstivity –Streptococcus sp susceptible to Augmentin, Cephotaxime, Gentamicin , Ciprofloxacin, Lincomycin, Roxithromycin , Tetracyclin, Llinezolid, Levofloxacin

RFT and LFT were within normal limit

24 hrs urine protein was 323 mg/day . Serum creatinine was 1.09.

HIV, HbsAG ,HCV negative



MDCT scan of chest showing multiple varying sized well defined non enhancing hyposense lesions scattered in b/l lung parenchyma (L>R) predominantly in peribronchovascular and subpleural location .

Nasal swab showed : 1)staphylococcus aureus susceptible to

Cloxacillin ampicillin+sulbactum , cephalexin, augmentin ,cephotaxime, gentamicin , ciprofloxacin, lincomycin , tetracycline, linezolid , levofloxacin

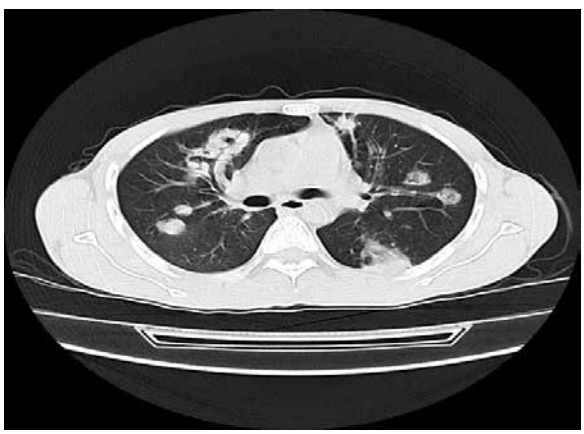
2)Acinetobacter spp susceptible to gentamicin , amikacin, ofloxacin , netiliin

c-ANCA was positive.

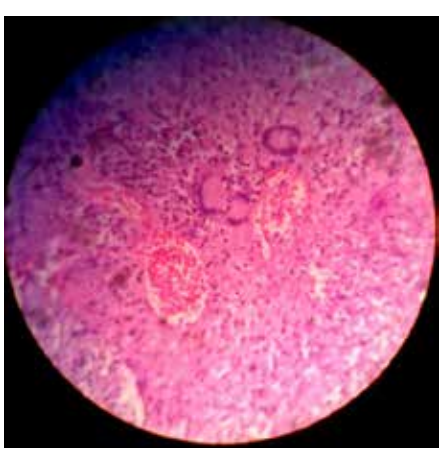
Ophthalmology reference was taken for red eyes. There was evidence of anterior wall filaments and impending perforation of sclera. Fundus was normal.

As patient had severe headache , he was referred to neurologist . Examination revealed no e/o increased intracranial pressure & meningitis. CT scan brain revealed no abnormality.

ENT reference was taken as patient had discharge from left ear. Examination revealed secondary otitis media and retracted tympanic membrane. Nasal mucosal showed congestion and blood clots. Oral cavity examination revealed fibrosis of mucosa. Multiple nodular lesions were seen in the throat and over the soft palate.



Nasal biopsy
H & E stained section studied show stratified squamous epithelium. Underneath seen is lymphoid proliferation , necrosis and neutrophilic infiltrate. Few vessel show changes of vasculitis . Areas of hemorrhage also seen. Histopathologist feature consistent of Wegners's Granulomatosis



The diagnosis of Wegener's granulomatosis was confirmed and patient started on Tab cyclophosphamide 2 mg /kg/day & Tab prednisolone 1 mg /kg /day

Strict ,regular monitoring of blood picture, urine and s. creatinine was done

Patient improved clinically as well as radiologically and was discharged on treatment and advised regular follow up.

Discussion:
WG was first described by Klinger in 1933, followed by other investigators, including Rossle in 1933, Wegener in 1936 and 1939, and Ringertz in 1947.1

the incidence of WG is estimated to be 5 to 12 cases per million, with a slight female predominance Male:female ratio= 2:3

Classic WG is the triad of granulomatous inflammation of the upper and lower respiratory tracts, systemic necrotizing small vessel vasculitis and immune glomerulonephritis.

Symptoms and clinical disease manifestations are the result of necrotizing granulomatous inflammation and small vessel vasculitis that occur in variable degrees of combination.

The use of the term "limited Wegener's granulomatosis" implies that: (a) the pathology is predominantly a necrotizing granulomatous and the vasculitis seen on biopsy is of lesser clinical significance; and (b) there is no immediate threat either to the patient's life or that the affected organ is at risk for irreversible damage . Pulmonary lesions in Limited Wegener`s.Granulomatosis are similar to that of Classical form.[5,6]

Severe wegeners 's granulomatosis is by definition either threatens the patient's life (alveolar hemorrhage) or a vital organ with the risk of irreversible damage (rapidly progressive glomerulonephritis, scleritis, or mononeuritismultiplex).

Over 90 percent of patients with Wegener's granulomatosis first seek medical attention for symptoms arising from either the upper and/or lower airway, the upperor lower respiratory tracts are involved in up to 90% of patientswith WG. Nasal involvement is responsible for presentingcomplaints in greater than 70% of cases .Nasal and sinus disease is characterized by congestion and epistaxis due to mucosal friability, ulceration, and thickening. Patients may also have features of chronic sinusitis and recurrent or chronic serous otitis. Perforation of the nasal septum and/or saddle nose deformity may result from ischemia of the nasal cartilage . Oral manifestations include gingival hyperplasia and oropharyngeal ulcerations. Subglottic steno sis occurs in approximately 20 percent of patients and can cause life-threatening compromise of the airway. Subglottic stenosis may occur in the absence of other features of active Wegener's granulomatosis, and its symptoms may be non- specific, e.g., dyspnea, hoarseness, cough or stridor.

Wegener's granulomatosis involving the lower airways can affect the pulmonary parenchyma, the bronchi, and rarely the pleura. Presenting features of parenchymal involvement may include cough, dyspnea, chest pain, or hemoptysis. How- ever, some patients may be completely asymptomatic. Patients with diffuse alveolar hemorrhage usually present with progressive dyspnea and anemia . Hemoptysis is absent in about one-third of patients. Patients with diffuse alveolar hemorrhage may deteriorate rapidly and experience respiratory failure, which has a mortality rate of 50 percent.

The most common form of pulmonary involvement in Wegener's granulomatosis is that of nodules or mass lesions, which may cavitate Frequently, these lesions are incidental findings on thoracic imaging studies as they cause little symptoms and do not result in significant abnormalities of pulmonary function. These lesions are caused by necrotizing granulomatous inflammation. Prominent air-fluid levels can be seen when the necrotic center of the inflammatory lesion gets superinfected . These necrotizing granulomatous lesions are a disease-defining feature of Wegener's granulomatosis

Inflammation and stenosis of the tracheobronchial tree occurs in at least 15 percent of patients with lung involvement. Endobronchial disease may be an incidental finding on bronchoscopy or present with cough, hemoptysis, wheezing, dyspnea, or symptoms related to parenchymal collapse or post- obstructive infection. Spirometry including inspiratory and expiratory flow-volume loops may show characteristic abnormalities indicative of degree and location of airway narrowing. Subglottic stenosis represents a fixed airway obstruction resulting in flattening of both the inspiratory and expiratory loops. If the intrathoracic trachea, or more commonly, one or both mainstem bronchi are affected, flattening of the expiratory curve can be found. Pleural effusions may occur, but are usually small, asymptomatic, and

incidental findings . Other thoracic manifestations of Wegener's granulomatosis include inflammatory pleural pseudotumors or hilaradenopathy. The latter should raise the suspicion of infection, sarcoidosis, or lymphoma. Glomerulonephritis is among the most concerning disease manifestations of Wegener's granulomatosis as it can progress to complete renal failure in the absence of symptoms. It is usually detected by the presence of abnormal labo- ratory results such as active urine sediment with microscopic hematuria and red cell casts, proteinuria, and declining renal function.

Continued vigilance for glomerulonephritis is essential as it is present at diagnosis in less than half of all patients. However, over the course of their disease, the kidneys are affected in 80 percent of patients. A renal biopsy is useful to establish a diagnosis of ANCA-associated vasculitis and to determine the renal prognosis .Other organ systems commonly affectedin WG include musculoskeletal (70%), ocular (30–60%), neurologic (20–50%) and gastrointestinal (5–10%) A wide spectrum of ocular manifestations has been ob- served in Wegener's granulomatosis, which may threaten vision by affecting the eye directly or involving its contiguous structures. Manifestations may include conjunctivitis, episcleritis, scleritis, keratitis, corneal ulceration, uveitis, and retinalvasculitis. Involvement of the lacrimal system may result in epiphora, dacryocystitis, and fistula. Retro-orbital inflammatory pseudotumors may affect one or both eyes, threaten the vision, and represent the most difficult challenge in the management of Wegener's granulomatosis.

The contribution of ANCA to the pathogenesis of Wegener's.Granulomatosis is unclear, but they appear to be responsible for some portion of the immune response that leads to disease.[4]

Any patient with Wegener's granulomatosis who presents with eye pain or redness, proptosis, change in visual acuity, diplopia, or loss of visual field should be referred for emergent ophthalmologic consultation. Nervous system involvement may occur in up to one- third of patients.

Mononeuritis multiplex of the peripheral nervous system caused by inflammation of the vasa nervorum as well central nervous system vasculitis and pachymeningitis represent severe disease manifestations with substantial risk of irreversible damage, persisting even after the acute inflammation is adequately controlled . Cardiac involvement may be occult. Regional wall motion abnormalities with a noncoronary distribution pattern are frequent echocardiographic findings. It is unclear whether this type of cardiomyopathy is the result of small vessel disease or inflammatory infiltration of the cardiac muscle. Pericarditis, valvulitis, and inflammatory pseudotumor have also been described.

A wide spectrum of cutaneous manifestations may be observed in Wegener's granulomatosis. Leukocytoclastic vas- culitis presenting as palpable purpura is most common, followed by pyodermagangrenosum like lesions and so called Churg-Strauss granulomas. Mucocutaneous involvement occurs in approximately 40% of patients with WG, and it can be thepresenting sign in 10% .The most common lesions are palpablepurpura, followed by oral ulcers. Skin changes that resemblepyodermagangrenosum can also be seen.Most common gastrointestinal manifestations include:abdominal pain, nausea/vomiting, diarrhea, hematochezia or melena.

Open lung biopsy is, however, usually necessary for the definitive diagnosis of pulmonary Wegener's granulomatosis.[7,8]

Such cases emphasise that Wegener's granulomatosis must be considered when assessing multiple pulmonary lesions in the absence of other clinical signs. Antineutrophil cytoplasmic antibody titre should be tested. Close communication between the cytopathologist and the clinician is essential to avoid an erroneous diagnosis in the presence of equivocal cytological test results. This is necessary to ensure that Wegener's granulomatosis is diagnosed early so that lifesaving treatment can be started promptly.

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