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Granulomatosis with Polyangitis Presenting As Sphenoidal Sinus Mass Lesion

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Abstract- *A 26 year male presented with complaints of epistaxis, purulent nasal discharge, serous ear discharge, since 6 month. Chest complaints were cough and chest pain since 3 month. Chest Skiagram revealed bilateral multiple nodular lesions. CT paranasal sinus revealed mass lesion in sphenoid sinus along with bony erosion. Biopsy of mass lesion and nasal mucosa revealed necrotizing granulomatous inflammation. c-ANCA was positive. Patient was discharged on oral prednisolone and cyclophosphamide. We report here a case of Granulomatosis with polyangitis in young male presenting with sphenoidal sinus mass lesion.*

Key Words-
GPA- Granulomatosis with polyangitis
IIF-Indirect Immunofluorescent assay
ANCA- Anti Neutrophilic Cytoplasmic Antibodies
ANA-Anti nuclear antibody
PR3- Proteniase 3
MPO- Myeloperoxidase

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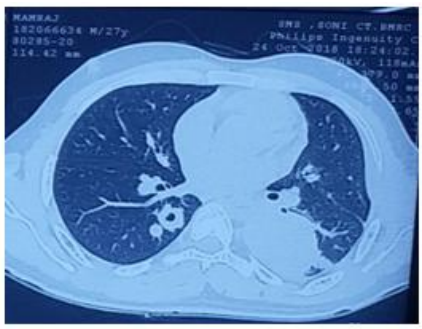
I. Introduction

Granulomatosis with polyangitis **(GPA)** encompasses necrotizing granulomatous inflammation involving the respiratory tract, and necrotizing vasculitis affecting small to medium-sized vessels.**[1]** Most frequently involved sites are upper airways, lungs, and kidneys.**[2]** Maxillary sinus is most frequently involved in GPA whereas sphenoid sinus is least commonly involved. Nasal and sinus disease is characterized by congestion/epistaxis due to mucosal friability, ulceration, and thickening .Patients may also have recurrent or chronic serous otitis. GPA involving the lower airways can affect the pulmonary parenchyma,the bronchi and rarely the pleura. The most common airway abnormality in GPA consists of mucosal edema, erythema, thickening, and granularity of mucosal surface.**[3]** Presenting features of parenchymal involvement may include cough, dyspnea, chest pain, or hemoptysis. However, some patients may be completely asymptomatic.

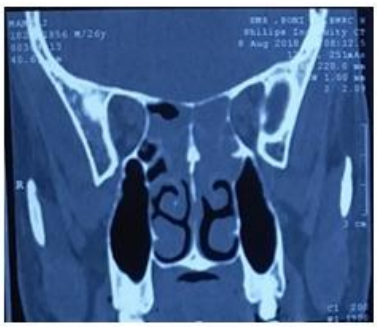
II. Case Report

A 26 year old male, engineer by occupation presented with complaints of epistaxis, foul smelling purulent nasal discharge along with bony fragments and serous ear discharge since 6 month.Patient also complained of loss of appetite and weight since 4 month along with cough and left sided chest pain since 3 month. He had already taken multiple courses of oral antibiotics since last 6 months and since 2 months he was on anti-tubercular treatment with no improvement. On physical examination saddle nose deformity was appreciated along with the crusting of nasal mucosa and there were decreased breath sounds in left interscapular and infrascapular area on auscultation. Complete blood counts, liver function and renal function tests were within normal limits. HIV status was Non-reactive and Random blood sugar was 83mg/dl. Urine Analysis was normal. His sputum sample was negative for mycobacteria on both smear by Zeihl-Nielson method and culture by BACTEC method.Chest Skiagram revealed bilateral multiple nodular lesions. CECT chest showed bilateral multiple nodules with some nodules undergoing cavitation. CECT paranasal sinus revealed mass lesion in sphenoid sinus also involvement of ethmoid sinus. Nasal endoscopy showed perforation of the bony part of nasal septum, biopsy was taken from the mass lesion and nasal mucosa of inferior meatus which revealed necrotizing granulomatous inflammation. Pure tone audiometry showed Bilateral conductive hearing loss suggestive of Chronic serous otitis media. RF,ANA, p-ANCA were negative whereas c-ANCA was positive(>100) by IIF method .Further, PR3 came out to be positive as well. Thus final diagnosis of GPA was

made in view of compatible clinical, radiographic, histological and serological profile. Patient was discharged on oral prednisolone and cyclophosphamide and was advised for follow up after 1 month. On follow up patient had dramatic improvement in clinical status and radiological features.



(Fig-1)



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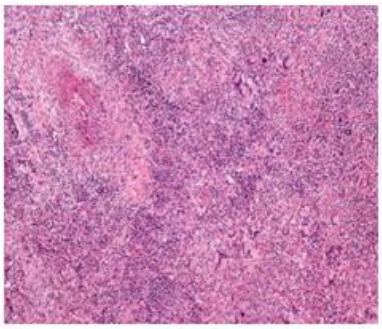
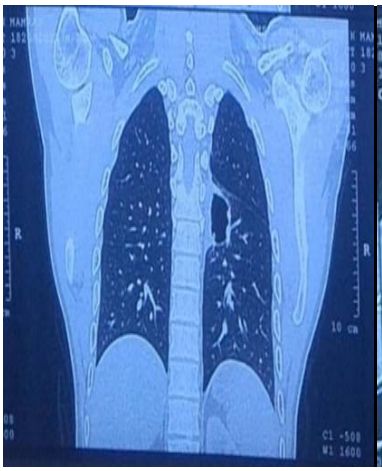


(Fig-3)

Fig-1 CT chest shows bilateral pulmonary nodules with some cavitary nodules and soft tissue lesion in left lower lobe
Fig-2 CT paranasal sinus showing mass lesion in sphenoid sinus and ethmoid sinus
Fig-3.Biopsy from mass lesion showing necrotising granulomatous lesion.



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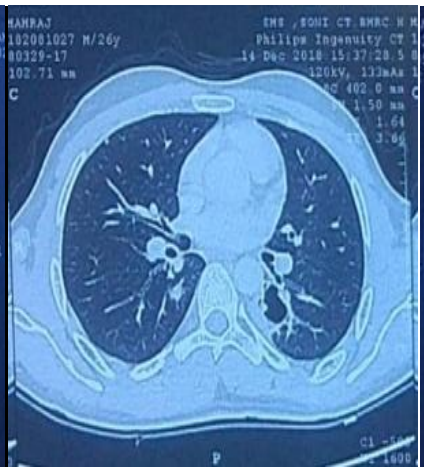

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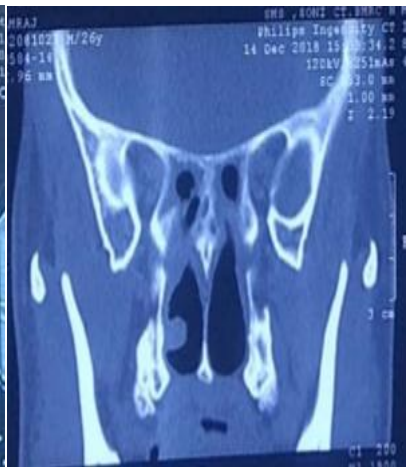


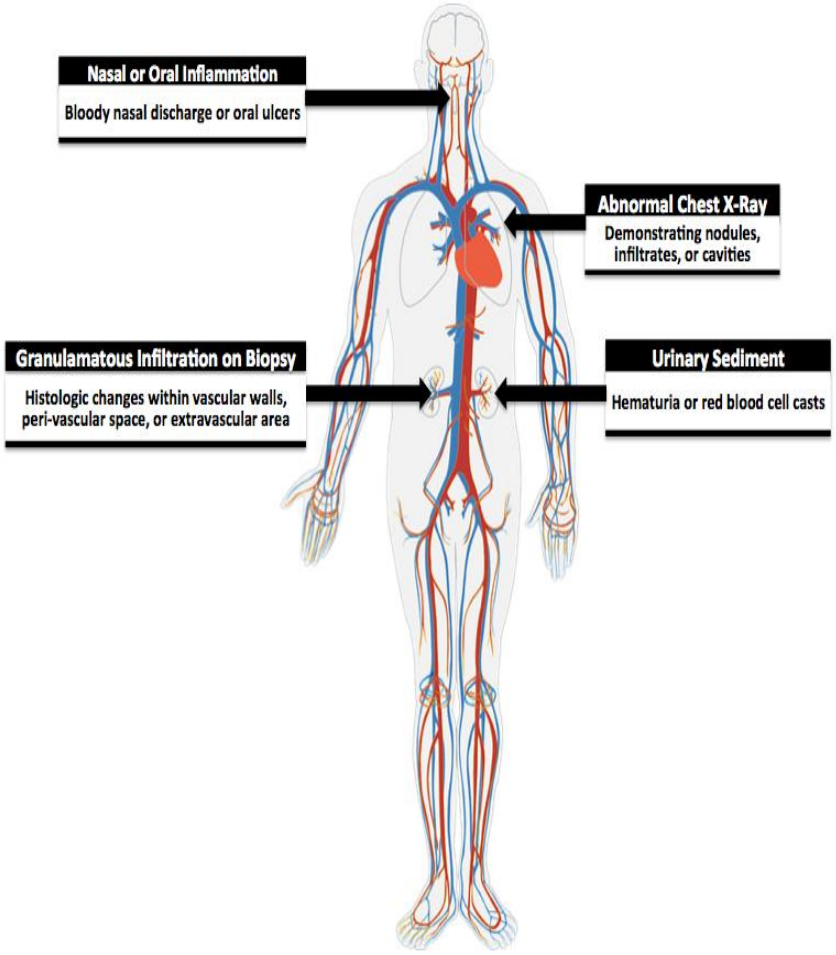
(Fig-6)

Fig-4 and 5.CECT chest (follow up) showing marked diminution in nodular lesions
Fig-6.CT Paranasal sinus (follow up) showing involvement of sinus and mucosal edema

III. Discussion

GPA is part of a family of syndromes that are commonly referred to as ANCA-associated vasculitides (AAV), which include GPA, microscopic polyangiitis (MPA) and Eosinophilic granulomatosis with angitis(EGPA). It is the most common form of vasculitis to involve the lungs.[1]It was first described by Klinger in 1933, followed by other investigators, including Rossle in 1933, Wegener in 1936 and 1939, and Ringertz in 1947.[4] Earlier it was known as Wegener Granulomatosis.GPA has a broad clinical spectrum that ranges from limited disease with predominantly necrotising granulomatous inflammatory lesions affecting the respiratory tract to severe, life-threatening necrotizing vacuities affecting many organs. It is a type of primary systemic vacuities. Over 90% patients with GPA first seek medical attention for symptoms arising from either upper and/or lower airway. The etiology of GPA and other AAV remains unclear. Many theories have been proposed, and it is probably the result of a combination of both permissive genetic and triggering environmental factors. ANCA panels can confirm the presence of an ANCA associated vasculitis and reveal a distinct diagnosis.Enzyme linked immunosorbent assay (ELISA) and staining by indirect immunofluorescence (IIF) are used to detect anti-PR3 and MPO ANCA.[5] ELISA is the newer method and its advantages include the speed at which one can obtain results, ease of performance, cost, efficiency and portability.[6] The disadvantages of ELISA include low sensitivity, especially in regards to p-ANCA related disease.[7] By combining ELISA and

IIF, both a high specificity (98%) and sensitivity (75%) can be reached. Studies suggest that an initial screening of the patient using ELISA, followed by a confirmatory IIF test maximizes the efficiency of these tests.[8]



(Fig-7):

Criteria for GPA. The American College of Rheumatology classifies a patient to have GPA if they present with at least 2 of these 4 symptoms (sensitivity is 88.2% and specificity 92.0%).**(Fig-7)**[9]

Other differential diagnosis of cavitating lung nodules are tuberculosis, fungal infections, septic emboli,sarcoidosis, rheumatoid lung disease,langerhans cell histiocytosis, cavitating metastasis, tracheobronchial papillomatosis and benign metastasizing leiomyoma. A positive biopsy is strongly supportive of a diagnosis of vasculitis and biopsies are recommended to assist in establishing a new diagnosis and for further evaluation for patients suspected of having relapsing vasculitis as per EULAR/ERA-EDTA/EUVAS joint recommendations.[10] No renal involvement was seen in this case although it can be seen in upto 50% of patients at the time of presentation and in upto 80% during the natural course of disease. Glomerulonephritis does subsequently develop in 77 to 85 percent of patients, usually within the first two years of disease onset. Accordingly, normal renal function at presentation cannot rule out GPA as in our patient with chronic sinusitis and pulmonary symptoms without renal involvement. Granuloma formation is a key pathologic finding in two of the ANCA-associated vasculitides: GPA and EGPA.It is suggested that PR3 (and possibly also MPO) displays features of an endogenous "danger signal" inducing autoinflammation. Although diagnostic sensitivity and specificity of PR3-ANCA and MPO-ANCA for the ANCA associated vasculitis are very high, a minority of patients with GPA has MPO-ANCA,which indicates the diagnosis of MPA. It is known that PR3-ANCA was found in 70% to 90% of patients with active GPA, and MPO-ANCA was observed in only 5% to 10% of patients with GPA. The racial difference may affect the selection of PR3-ANCA and MPOANCA in the incident of vasculitis. Fujimoto et al. showed that MPA and MPO-ANCA were the predominant subtypes in Japan, while GPA and PR3-ANCA were predominant in the UK. Chen et al. also reported that patients with MPO-ANCA positive GPA were not rare in Chinese subjects. However, it is reported that MPO-ANCA positive GPA patients showed less organ involvement than PR3-ANCA positive GPA patients. Therapy of GPA is comprised of two components which are induction of complete remission and maintenance therapy. Induction phase treatment includes combination of glucocorticoids plus cyclophosphamide for three to six months to achieve remission.

For remission-induction of non-organ-threatening AAV , treatment with a combination of glucocorticoids and either methotrexate or mycophenolate mofetil is recommended .Whereas for remission-induction of new-onset organ-threatening or life-threatening AAV treatment with a combination of glucocorticoids and either cyclophosphamide OR rituximab is recommended .[10] Plasma exchange therapy is suggested in the presence of diffuse pulmonary hemorrhage, rapidly worsening kidney function or overlap syndrome of AAV and anti-glomerular basement membrane antibody glomerulonephritis as in Goodpasture syndrome.[10] Remission is defined as a clearing of active lesions or resolution of organ dysfunction. For remission-maintenance of AAV recommended treatment is a combination of low-dose glucocorticoids and either azathioprine, rituximab, methotrexate or mycophenolate mofetil. [10] Recommendations are that remission-maintenance therapy for AAV should be continued for at least 24 months following induction of sustained remission. Prompt initiation of treatment for AAV can achieve remission at 6 months in >90% of patients, however, relapse rates approach 50%. Without treatment, the average survival rate is 5 months with one and two year mortality rates of 82% and 90% respectively. This case emphasizes the importance of early diagnosis and management to prevent permanent organ damage.

IV. Conclusion

Any patient presenting with recurrent sinonasal symptoms should be evaluated for GPA. Biopsy of an affected organ is paramount for the definitive diagnosis. A high index of suspicion is crucial for early diagnosis, which can substantially reduce morbidity and mortality.

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