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General Medicine

ATYPICAL PRESENTATION IN GRANULOMATOSIS WITH POLYANGIITIS :A CASE REPORT

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ABSTRACT Granulomatosis with Polyangiitis (GPA) is an rare multi-systemic disease characterized by necrotizing granulomatous inflammation of the upper and lower respiratory tracts and focal necrotizing vasculitis (commonly known as Wegener's triad). The diagnosis of granulomatosis with polyangitis is from the clinical and laboratory findings and from the presence of circulating anti-neutrophil cytoplasmic antibodies (ANCA) although the absence of ANCA does not exclude the diagnosis. It is characterized by formation of granulomas with fibrinoid necrosis, as well as by ulceration and inflammation of small and medium-sized vessels with the involvement of upper and lower airways and kidneys. The process may also occur in other less typical locations, such as the gastrointestinal tract, heart, and nervous system.We described a case of 21-year-old male with atypical presentation pansinusitis initial respiratory tract symptoms such as foul smelling nasal, ear ache and discharge . Later he developed fever. Quantitative anti PR3 positive. He was treated IV methyl prednisolone pulse therapy followed by oral prednisolone tappering doses and Inj. cyclophosphamide. Patient condition get improve satisfactory and on follow up after 3 months he was reasonably well. In this case report, we wanted to show that Granulomatosis with Polyangiitis (GPA), although rare, should be considered in the above clinical scenario and treatment should be initiated as early as possible.

KEYWORDS : Antineutrophil cytoplasmic antibody-associated vasculitis, granulomatosis with polyangitis, Wegener's granulomatosis

Introduction:

Granulomatosis with Polyangiitis (GPA) is the most common Anti-Neutrophil Cytoplasmic Antibody (ANCA) associated vasculitis and is a multisystem autoimmune disease characterized by a combination of airway disease and glomerulonephritis 1. Incidence of GPA as 11 million, equally distributed amongst males and females2. Patients typically present in age age groups of 40-50 years with various nonspecific symptoms including hemoptysis, stridor, hoarseness and wheezing, Systemic symptoms such as fever, weight loss, myalgia,headache may also be present, usually lasted for several weeks to months 3 . Many Studies found that prompt recognition and early intervention are most important factor in the morbidity and mortality of these patients 4. In case of GPA , early diagnosis and early treatment leads to clinical remission in up to ¾ th of patients, but a rapidly progressive disease process and high rate of mortality may occur if diagnosis is delayed3 .We report an atypical presentation of GPA in a middle-aged man who presented with only ear ache ear discharge and High grade fever without any renal involvement. Our case shows the importance of having a suspicion for GPA in patients with atypical presentations, so that early diagnosis and initiation of treatment will help in the better outcome of the patient.

Case Report

A 21-year-old male h/o both earache and ear discharge since 2 month ,for this he took medications but patient condition deteriorated and presented with high grade fever , sore throat , swelling over face and neck area and nasopharyngeal region, difficulty in swallowing which progressed over a month to cause painful mastication and mouth opening. Also complaining of decreased in hearing in both ear . He went to an ear, nose, and throat (ENT) surgeon for the same and fine-needle aspiration cytology (FNAC) was performed after which he developed a left facial weakness Within a few days, after, he developed redness and discharge from the both eyes .During hospitalization developed myocardial infarction (IWMI) received treatment for same . Also complaining of headache and blurring of vision .On examination, he was found to be pale, with diffuse tender swelling of b/l submandibular, left infra-nuclear facial palsy, bilateral mixed hearing loss. . FNAC from cervical swelling suggested inflammatory cystic lesions .Biopsy from nasopharynx f/s/o Granulomatosis with Polyangitis [Fig2]. Erythrocyte sedimentation rate (70 mm/h), C-reactive protein (23 mg/l) levels were elevated, an automated count revealed anaemia (Hb-9.0 g/dl), leukocytosis (16,700/cumm) and thrombocytosis (5,57,000/cumm), renal and liver functions were normal. Blood culture negative. ANA, P-ANCA , C-ANCA by ELISA were performed were negative . Cultures from the nasopharyngeal overgrowth were sterile. A contrast computed tomography (CT) skull base to diaphragm suggested pansinusitis (b/l maxillary , ethmoid, sphenoid sinus) and soft tissue fluid density seen in b/l middle ear b/l mastoid air cells s/o b/l otomastoiditis. High-resolution CT thorax shows hypoenhancing mass of appx 5.5x3.3x4.7cm in right side of anterior mediastinum, multiple nodule of varying size in b/l lung parenchyma largest of measuring 2.9x2.1cm in posterobasal segment of left lower lobe with central cavitation f/s/o possibility of Granulomatosis with polyangitis. Magnetic resonance imaging brain showed Granulomatous lesions along leptomeningeal lining (granulomatous meningitis) [Figure1] without any bony destruction. Pure-tone audiometry was suggestive of Bilateral severe mixed hearing loss. The renal function test and urine routine examination were within normal limits and no evidence of other systemic involvement. 2D ECHO Suggested IHD (RWMA Present, Hypokinesia in inferior, inferno lateral, inferioseptal.) with moderate LV dysfunction, LVEF-45%. Quantitative PR3-ANCA (C-ANCA) 58.02 AU/mL, MPO-ANCA (P-ANCA) 2.29AU/mL.rapidly progressive organ-threatening ANCA vasculitis a provisional diagnosis of GPA was kept and the patient was given pulse injection methylprednisolone (1 g) for 3 days followed by tapping dose of oral prednisolone. Induction with Intravenous cyclophosphamide according to the European vasculitis group (CYCLOPS) protocol was initiated.

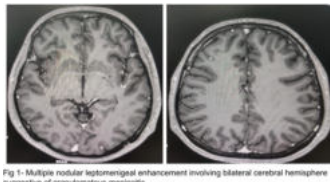
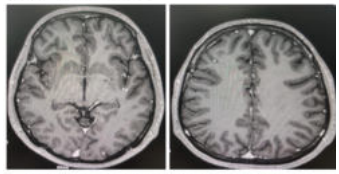


Fig 1- Multiple nodular leptomenigeal enhancement involving bilateral cerebral hemisphere suggestive of granulomatous meningitis



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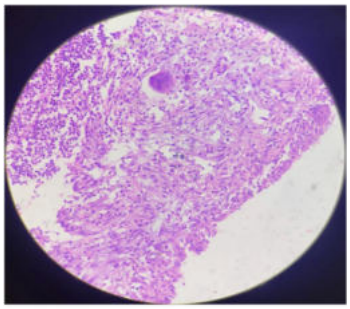

Fig 2 - Biopsy from nasopharyngeal growth f/s/o Granulomatosis with Polyangitis

Discussion :

Granulomatosis with polyangitis (GPA) is an multisystem autoimmune systemic vasculitis. Many factors like Infectious, environmental, chemicals, and toxins have been advocated as etiologic factors in predisposed people3. It is usually associated with high levels of serum antineutrophilic cytoplasmic antibodies (ANCA), even positive ANCA serology is not pathognomonic of GPA, especially in the absence of clinical and histological findings of systemic vasculitis. Clinical manifestations of GPA include constitutional symptoms, , oral ulcers and crusting rhinitis, and subglottic laryngeal stenosis, upper and lower respiratory tract manifestations. Some patients may develop Neurological ,Cardiovascular or renal involvement4. The histological feature of GPA is characterized by inflammatory infiltrate composed by neutrophils, lymphocytes, macrophages, eosinophils, and giant multinucleated cells, plasma cells, focal vasculitis of small veins, arteries, and capillaries and necrosis. HRCT helps in achieving diagnosis, as it shows typical GPA-related pulmonary findings such as nodules, cavitary lesions, tracheobronchial stenosis, and interstitial lung disease5. Early diagnosis plays a main role in outcomes because a proper treatment may induce clinical remission, decrease the morbidity, and improve the survival of patients. Currently, management of GPA is based on immunosuppressant drugs (cyclophosphamide, azathioprine, and methotrexate) and monoclonal antibodies (rituximab). Plasmapheresis has been proposed in addition with cyclophosphamide for patients with aggressive vasculitic involvement of the kidney [6].

Conclusion :

In our case, patient's symptoms are related to Upper respiratory tract region, without systemic involvement. Our patient presented with atypical symptoms, in GPA we usually search for so called triad including upper and lower respiratory tract with glomerulonephritis. As for GPA early diagnosis is crucial, it is important to keep suspicion of GPA even only with upper respiratory tract involvement, it is not necessary to present the triad. This is a case of atypical GPA without lower respiratory tract involvement and renal involvement. If treatment can be initiated at the earliest, thereby improving the survival of patients with GPA.

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