Case Report

Lung Nodules with a Saddle nose-A happy ending to a stormy course

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Introduction

Granulomatosis with polyangiitis (GPA), formerly known as Wegener's granulomatosis (WG), is a systemic disorder that involves both granulomatosis and polyangiitis. It is a form of vasculitis that affects small- and medium-size vessels in many organs. Here we report a case of a middle aged female who presented to us with cough, fever and dyspnoea on exertion and was diagnosed with Wegener's granulomatosis

Case Report

A 50 year old female, housewife, diabetic on oral hypoglycaemic agents presented to our department with complaints of cough with scanty mucoid expectoration, intermittent low grade fever of 2 months duration and insidious onset, progressive grade I to grade II dyspnoea on exertion(DOE) of 1 month duration. There was associated myalgia and eye pain and redness. She had loss of appetite with no documented loss of weight. Past history revealed history of recurrent episodes of mucopurulent discharge from nose with 2 episodes of epistaxis for the last 6 years. Recurrent episodes of eye pain and redness were present since 1year, the recent episode since 1 month.

On examination she was moderately built and nourished. There was no pallor, icterus, cyanosis, clubbing, lymphadenopathy, pedal oedema or elevated JVP, afebrile with stable vitals. Both eyes showed conjunctival congestion with thinned sclera and a bluish hue with developing staphyloma (Figure 1). On auscultation, bilateral fine mid to late inspiratory crackles in mammary, infra axillary and infra scapular areas were heard.

Her CXR (Figure 2) revealed bilateral nodular shadow predominantly in right mid and lower zone with both hemidiaphragms at same level. Her hemogram showed leucocytosis-12200/cumm, hemoglobin- 12.1gm% with elevated ESR 120/1st hr. She was referred to our department with a contrast enhanced computerised tomography(CECT) thorax (Figure 3) which showed bilateral nodules in the middle and lower lobes with feeding vessel sign, with no pleural effusion or mediastinal adenopathy and was referred as a case of angioinvasive metastasis/ septic emboli.

Patient was admitted and started on parenteral antibiotics, and she was worked up for connective tissue disorder and sarcoidosis. An opthalmology and ENT opinion were obtained and CT Paranasal sinus (PNS) was ordered. Her Antinuclear antibody (ANA) and Creatine phosphokinase (CPK) were negative, Rheumatoid factor was borderline elevated (47IU/mL), serum calcium – 8.4mg/dl, serum Angiotensin converting enzyme(ACE)-65U/L. There was no evidence of proteinuria, with normal renal function tests (RFT) and liver function tests (LFT). She could not raise phlegm for testing and her mantoux was 22 mm. Her DLco was 56.9ml/min/mm Hg. Fiberoptic bronchoscopy showed normal bronchial tree and transbronchial lung biopsy(TBLB) was done from Right lower lobe basal segments.

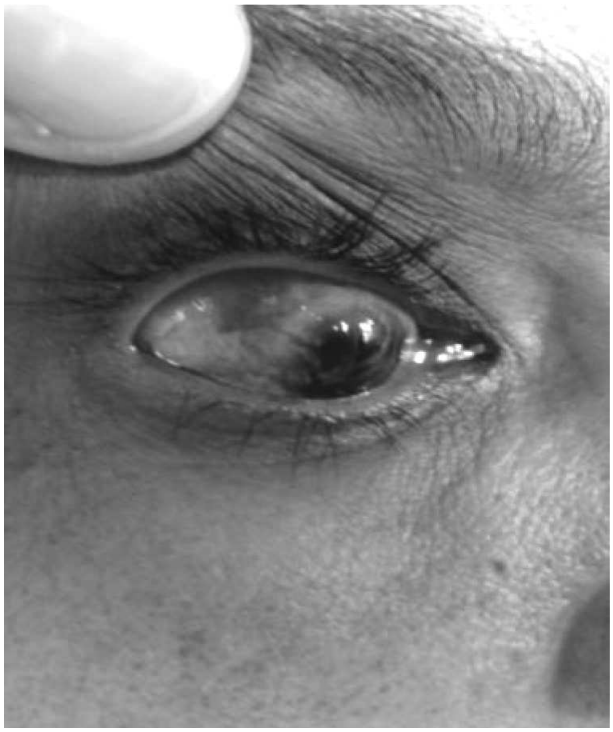

Figure 1 : Episcleritis with staphyloma

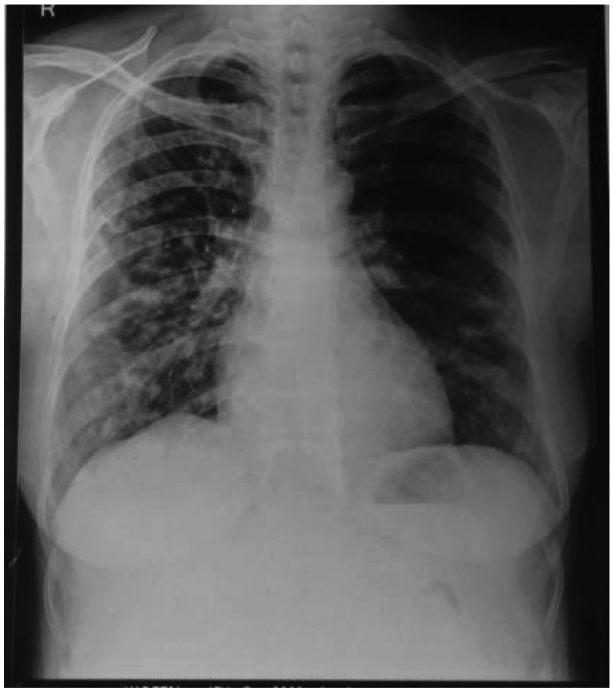

Figure 2 : Chest X ray showing multiple nodular shadows

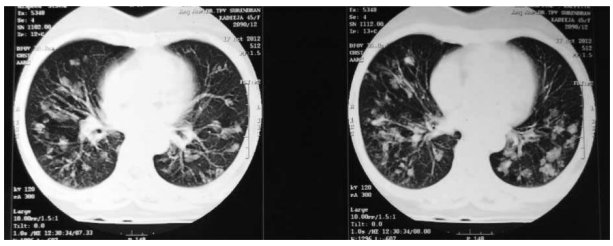

Figure 3 : CECT Thorax-Lung window

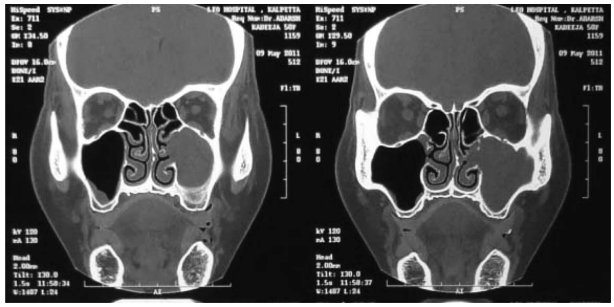

Figure 4 : CT Paranasal sinus

Ultrasound abdomen was within normal limits. Ophthalmologist suggested the possibility of connective tissue disorder in view of recurrent episcleritis. CT PNS (Figure 4) showed bilateral maxillary sinus polyps with sinusitis, bilateral frontal sinusitis and hypoplastic nasal bone. She had noticed her nasal deformity (Figure 5) for the last one and a half years.

Her recurrent episcleritis, pan sinusitis and nasal deformities with nodular shadows in the lung prompted us to do an ANCA evaluation. The reports revealed a positive c- ANCA (6.37u/ml) and negative p ANCA(1.93u/ml). TBLB showed lymphocyte infiltrate with no definite granuloma. The otolaryngologist did a Direct nasal endoscopy which showed irregular granulation like area in the posterosuperior part of nasal septal mucosa, with histopathology of the biopsy specimen revealing granulomatous inflammation.

Our patient who had nasal mucosa ulcers, saddle nose deformity, bilateral sinusitis with nasal polyp, scleritis, bilateral lung nodules with granulomatous inflammation on nasal mucosa biopsy and positive c ANCA was diagnosed to have Wegeners granulomatosis. As there was life threatening organ involvement in the form of vision impairment it was a severe form.

As she had severe life threatening disease, she was started on high dose steroids, Cotrimoxazole 960 mg OD thrice a week and pulse Cyclophosphamide infusion was suggested. Dramatically as the patient was being hydrated for cyclophosphamide infusion she collapsed in the ward with bradycardia and hypotension. Noting an elevated JVP with no increase in the distribution of crackles a right heart infarction was clinically diagnosed with the Electrocardiogram (ECG) showing inferior wall myocardial infarction (IWMI) with Right Ventricular Infarction and Complete heart block (CHB). All emergency and supportive treatment was started and in view of a short window period, she was immediately considered for primary Percutanous transluminal coronary angioplasty(PTCA) by

the cardiologists. A coronary angiogram showed mid total occlusion of Right coronary artery(RCA) and PTCA was done with Bare metal stent (BMS). Post procedure period was uneventful, with normal sinus rhythm in ECG. Considering vasculitis as a precipitant for coronary stenosis, she was given Pulse cyclophosphamide therapy along with Prednisolone from the cardiology department. Thereafter patient was discharged with dual antiplatelets, Methotrexate and Prednisolone with a final diagnosis of Severe Wegeners Granulomatosis, Acute coronary syndrome- ST elevation Myocardial infarction- IW with CHB and Type II Diabetes Mellitus.

Patient was under our follow up, Prednisolone was tapered after 6 months and she is presently on maintenance dose Prednisolone. Patient improved symptomatically and radiologically with disappearance of the nodules completely on follow up skiagrams.

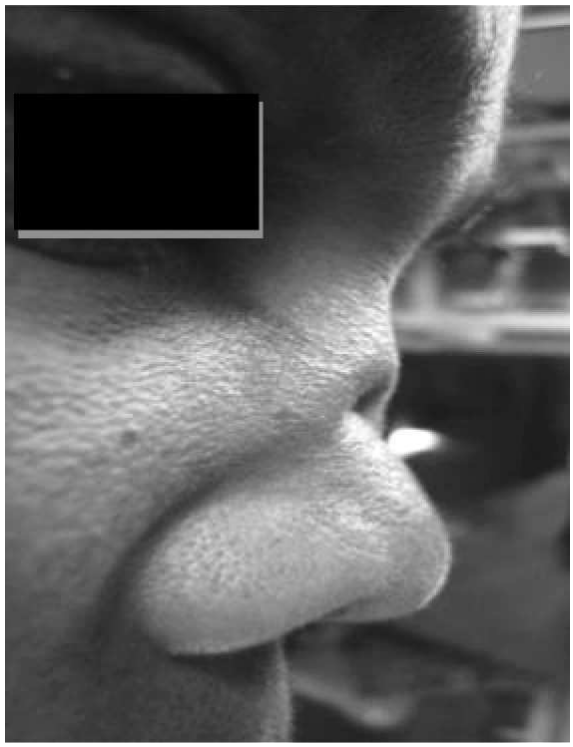

Figure 5 : Saddle nose deformity

Discussion

Wegener's granulomatosis is a distinct clinicopathological entity characterised by granulomatous vasculitis of upper and lower respiratory tract together with glomerulonephritis. It is the most common pulmonary vasculitidis with an estimated prevalence of 3 in 1,00,000 and with a male to female ratio of 1:1. Mean age of onset is 40 years.

Lung involvement typically appear as multiple bilateral nodular cavitating infiltrates which on biopsy reveal typical necrotising granulomatous vasculitis[1]. Immunopathogenesis is not clear; however it is proposed to be due to an abnormal cell mediated immune response to an exogenous or even endogenous antigen that enters through or resides in the upper airway.

Clinical manifestations

Involvement of upper airway occurs in 95% of patients. They often present with severe upper respiratory tract findings such as paranasal sinus pain, tenderness and purulent or bloody nasal discharge, with or without nasal mucosal ulceration. Nasal septal perforation may follow leading to saddle nose deformity.

Pulmonary involvement may be manifested as asymptomatic infiltrates or as cough, hemoptysis, dyspnoea and chest discomfort. Endobronchial disease may lead to obstruction with atelectasis.

Eye involvement may range from conjunctivitis to dacryocystitis, episcleritis, scleritis and retroorbital mass lesions.

Renal disease may occur as either mild glomerulitis with proteinuria, hematuria and red blood cell casts or as rapidly progressive renal failure. Skin and cardiac manifestations are also seen.

Lab findings include

Elevated ESR

Mild anaemia and leukocytosis

Mild hypergammaglobulinemia

Slightly elevated rheumatoid factor

Elevated c ANCA

ACR Criteria for classification[2]

1. Nasal or oral inflammation

2. Abnormal chest Xray

3. Active urinary sediment

4. Granulomatous inflammation on biopsy

Two or more of these if present is suggestive of Wegener's granulomtosis.

cANCA have been detected in more than 90% patients with active generalised WG and in 40 to 70 % with active regional WG[3].

Treatment

Oral cyclophosphamide (2 mg/kg/day) and prednisolone is the initial treatment of choice for Wegener's granulomatosis. By 3 to 6 months, assuming complete remission is achieved, azathioprine or methotrexate can be substituted for cyclophosphamide. Treatment should be continued for a minimum of 12 to 18 months. Relapse can be treated with cyclophosphamide and prednisolone.

References

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