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GRANULOMATOSIS WITH POLYANGITIS – RARE INITIAL RADIOLOGICAL PRESENTATION

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ABSTRACT

Granulomatosis with polyangiitis is a small vessel vasculitis and ANCA related.30-year female presented with complaints of rhinorrhea for 4 months. Xray chest showed doubtful Right midzone opacity. Coagulation profile and hemogram was unremarkable with raised ESR and Proteinuria on urine analysis. Sputum studies for PTB was negative. HRCT thorax showed *solitary soft tissue mass lesion in Right lower lobe with spiculated margins suggestive of neoplasm*. Biopsy of Lung mass was suggestive of tissue was consistent with granulomatosis with polyangiitis and C – ANCA was positive. She developed ear, eye symptoms with skin rashes later. Diagnosis of Granulomatosis with polyangiitis with sinusitis, mastoiditis, scleritis, vasculitic skin rash, kidney proteinuria was made and treated with Prednisolone, Azathioprine, Mycophenolate Mofetil following which patient improved and is on follow up.

Key-words: Granulomatosis with polyangiitis, vasculitis, soft tissue mass with spiculated margins, anti-neutrophil cytoplasmic antibodies.

INTRODUCTION

Pulmonary vasculitis is a group of disorders which involve inflammation of the blood vessels of the lungs. Granulomatosis with polyangiitis, previously known as Wegener's granulomatosis is a rare disorder with prevalence of 24 to 157 per million population.[1] It is ANCA associated vasculitis involving small vessels and classified under primary idiopathic small vessel vasculitis according to 2012 Chapel hill consensus statement of nomenclature. [2] Most patients have radiological involvement during presentation with lung and kidney involvement. There are no criteria to diagnose this diagnose and it requires clinical suspicion in a scenario of multi organ involvement. The patient had clinical and radiological findings suggestive of lung malignancy with no other organ involvement during initial presentation. Biopsy followed by histopathological examination was definitive and conclusive of the disease along with serum ANCA levels.

CASE REPORT:

A 30 years female presented complaints of rhinorrhoea for 4 months in duration. Physical examination was unremarkable other than deviated nasal septum and sinusitis. Hemogram and coagulation profile were unremarkable other than raised ESR and proteinuria on initial testing. Chest Xray was done in view of preoperatively evaluation of septoplasty which revealed doubtful retrocardiac right lower zone homogenous opacity (figure 1).

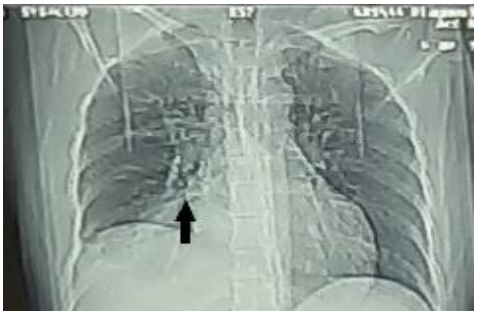

Figure 1:Chest Xray PA view showing doubtful opacities on right lower lobe lung.

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Sputum studies for pulmonary tuberculosis was negative. To further investigate the doubtful opacities on Chest Xray, Computed Tomography of the thorax was done and it revealed a large single soft tissue mass lesion in the Right lower lobe of size 6cm* 5.6cm with spiculated margins suggestive of malignancy (figure2). The demographics of the patient and initial clinical presentation made clinical suspicion of vasculitis as one of the differential diagnoses as patient did not respond to empirical medical treatment for sinusitis.

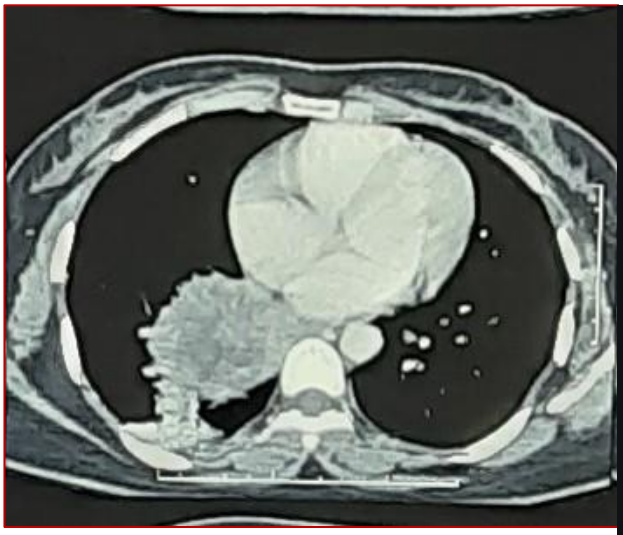

Figure 2: Computed Tomography showing Solitary soft tissue mass with spiculated margins on Right lower lobe lung.

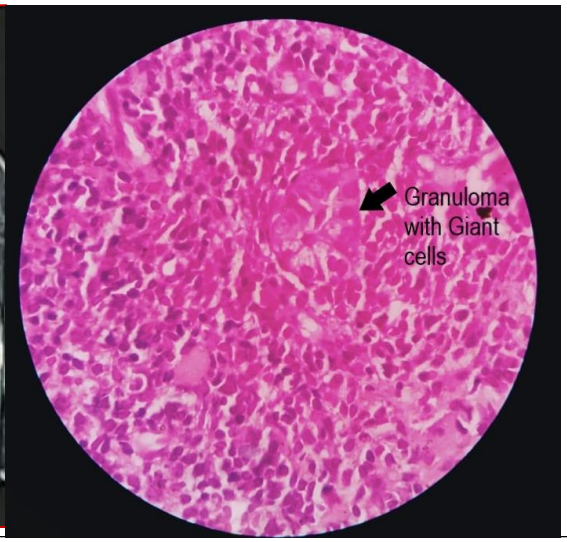

Figure3:HPE H&E stain(40x) of CT guided biopsy of Right lower lobe lung mass showing ill formed granuloma with Giant cell

Image guided biopsy and Haematoxylin and Eosin staining of the mass revealed ill formed granuloma and necrosis with vessel involvement suggestive of Granulomatosis with polyangiitis (figure 3 and 4) which was confirmed with positive C ANCA test.

The patient developed ear, eye, skin, urinary symptoms later over the next few months. Final diagnosis of Granulomatosis with polyangitis with sinusitis, mastoiditis, scleritis, vasculitic skin rash and proteinuria was diagnosed in the next 6 months.

Initial steroid therapy consisting of 1mg per kg of body weight per day of methyl prednisolone followed by maintenance therapy of Prednisolone along with Azathioprine, Mycophenolate mofetil was started. The patient showed clinical and radiological improvement following treatment and on regular follow-up.

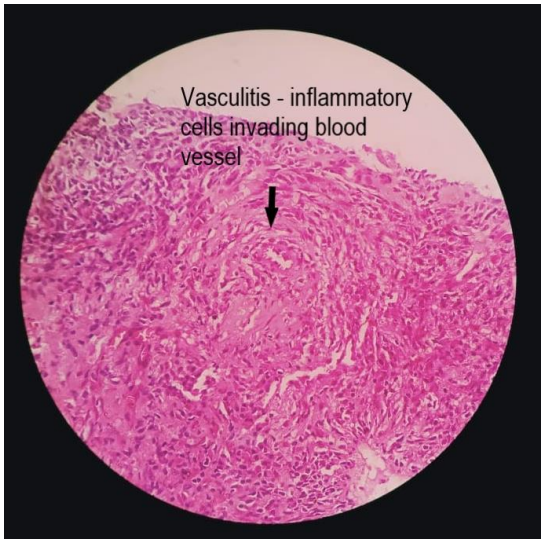

Figure 4:HPE H&E stain (40x) of CT guided biopsy of Right lower lobe lung mass showing vasculitis

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DISCUSSION:

Granulomatosis with polyangiitis is small vessel ANCA related vasculitis. Vasculitis is not classified according to etiology but rather similar clinic pathological features of inflammation according to 2012 Revised international Chapel hill consensus conference nomenclature.[2] Histopathological examination shows small vessel involvement distal to arterioles. Etiology is unknown but possible postulated mechanisms include autoantibody formation and neutrophil activation and ANCA production which causes chemokine production and repeated cycle of inflammation causing three major histopathological changes which includes lung parenchymal necrosis, vasculitis and granulomatous inflammation. ANCA related vasculitis are of two types namely P ANCA (perinuclear) which targets Myeloperoxidase enzyme and C ANCA (cytoplasmic) which targets Proteinase 3 enzyme on Indirect Immuno-fluoroscopy.

Clinical presentation of GPA is highly variable multiple organ systems involving upper airway disease(90 to 95%), pulmonary parenchymal disease(54 to 85%), alveolar haemorrhage (5 to 15%), glomerulonephritis(51 to 80%), gastrointestinal tract(less than 5%), eyes(35 to 52%), nervous system(20 to 50%), heart(8 to 16%), skin(33 to 46%). The initial presentation of renal involvement is around 40%.[3]

Radiological abnormalities of granulomatosis with polyangiitis (more than 80% of the cases) can be multiple nodular or multiple masses bilaterally (55 to 70%) or cavitary (35 to 50%) opacities involving bilateral lungs and multiple lobar involvement. Other presentation involve multiple consolidations bilaterally. Ground glass opacities can also be present in case of Diffuse Alveolar haemorrhage. CT sinus abnormalities were present in 70 to 90% of cases.[4]

Granulomatosis with polyangiitis is a systemic vasculitis with involvement of Upper and lower respiratory system, kidneys, skin, ears, eyes. The initial clinical and radiological presentation is highly variable. C ANCA serum test has a sensitivity of 90 to 95% when tested during active disease with multiorgan involvement which drops to 65 to 85% when single organ involvement is there and further reduces during remission. Specificity of C ANCA is 90% whereas p ANCA is positive is vasculitis and other autoimmune disorders.[5]

Hoffman and colleagues found that lung biopsies have diagnostic yield in 90% of cases.[3]

Acute glomerulonephritis with RPGN diagnosed with urinary sediments on urinalysis, haematuria, red cell casts, proteinuria, increased BUN and increased creatinine will be eventually present in most of the cases but need not be present during initial presentation.[6]

The uniqueness of this case lies in the atypical radiological presentation of granulomatosis with polyangiitis which presented as solitary soft tissue mass with speculated margin which is highly suggestive of malignancy. We are one of the first to report such a radiological presentation in granulomatosis and polyangiitis and rare in literature till date. Patient had only upper respiratory tract infection on initial presentation which was unresponsive to empirical medical management hence High degree of clinical suspicion along with biopsy and C ANCA level aided in diagnosis

LEARNING POINTS;

- Granulomatosis with polyangiitis can have atypical presentations and should be considered as one of the differential diagnoses in a case of solitary soft tissue mass on CT scan
- Temporality will play a major role when it comes to initial clinical presentation which may or may not include kidney and multiorgan involvement
- Radiological patterns of pulmonary diseases are only suggestive of a particular disease but histopathology is the good standard for diagnosis of vasculitis

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