GRANULOMATOSIS WITH POLYANGIITIS

Dr.Thilak N *, Professor Dr. S.Sujatha**
*Post graduate student, Department of General Medicine, Karpaga Vinayaga Institute of Medical Sciences and Research Centre

INTRODUCTION

- Wegener's granulomatosis is a systemic vasculitis that usually involves the lungs, upper respiratory tract and kidneys.
- Common presentations include recurrent sinusitis with purulent and bloody nasal discharge.
- Classic triad of upper and lower respiratory tract disease and glomerulonephritis.
- Approximately 85 – 90 % of patients with wegener's granulomatosis have pulmonary involvement during the course of their disease.
- ANCAs (90% of patients), usually directed against proteinase-3 (less commonly against myeloperoxidase present in severe, active disease).
- Pulmonary symptoms include cough, hemoptysis, dyspnea, and sometimes chest pain
- The most common radiographic findings are pulmonary infiltrates and nodules.
- Nodules are usually multiple and bilateral and often cavitary.
- The renal disease dominated the clinical picture with glomerulonephritis in about 77% of patients

CASE REPORT

- 53yr old female
- k/c/o Hypothyroidism (on thyroxine replacement)
- Presented with h/o - low grade fever- chills, small joints pain,dry cough
- Got treated outside as viral fever But symptoms persisted
- Patient developed - ear congestion
- cough with expectoration and blood tinged sputum
- Patient got admitted outside for the same
- in the course of hospital stay she developed B/L pedal edema

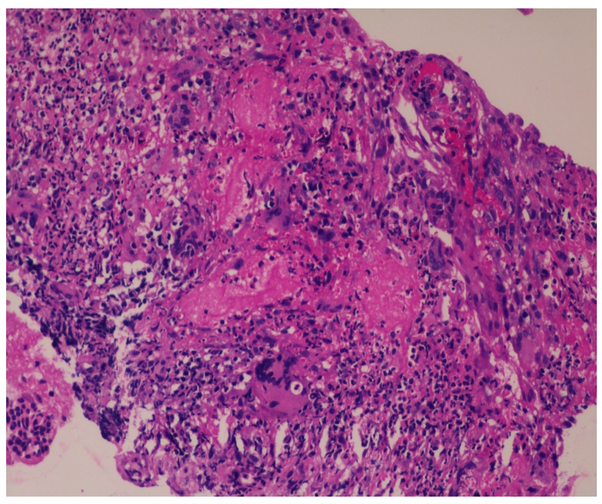
she presented with loss of weight(5kgs in a month) & appetite

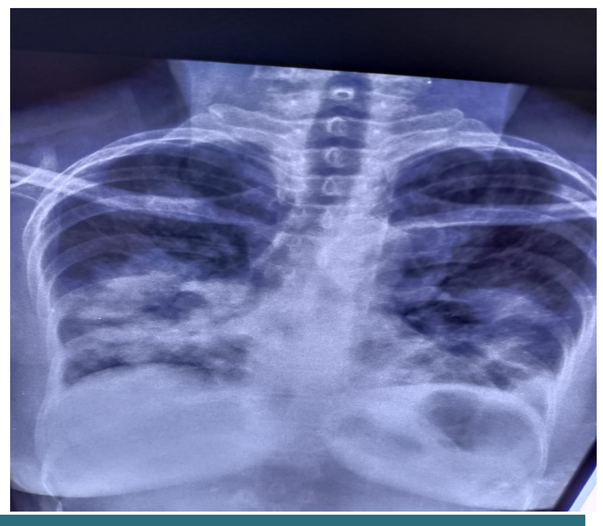
EXAMINATION

General examination:
- Both lower limb swelling present
- Afebrile
- Pallor present

Presenting vitals :
- HR- 82beats /min , normal volume, regular rhythm
- BP-120/70 mmHg
- Spo_2-98% with room air
- RR-16cycles /min

Systemic examination :
- CVS- S1,S2 heard ,no murmurs
- RS-B/L air entry present ,
- CNS- no focal neurological deficit
- P/A- soft ,not distended ,No organomegaly





DIFFERENTIAL DIAGNOSIS

- Eosinophilic Granulomatosis with Polyangiitis
- Cryoglobulinemia
- Goodpasture Syndrome
- Hemolytic-Uremic Syndrome
- Membranous glomerulonephritis
- Lymphomatoid granulomatosis
- Microscopic polyangiitis
- Langerhans Cell Histiocytosis

INVESTIGATIONS

INV	RESULTS	INV	RESULTS	INV	RESULTS
RBS	182mg/dl	HB	7.9	T.CHOLESTROL	108
UREA	27mg/dl	TC	22,100	TGL	141
CREATININE	1.8mg/dl	DC	88/7/2/3.1	HDL	42
T.BILIRUBIN	1	ESR	20/55	VLDL	18.9
D.BILIRUBIN	0.4	plt	9.14 lac	LDL	43.4
SGOT	57				
SGPT	36	Urine		SODIUM	136
T.PROTEIN	5.2	albumin	+	POTASSIUM	4.8
T.ALBUMIN	2.9	Sugar	+	CHLORIDE	95
TSH	2.39uIU/ml	Pus cells	2-3 hpf	BICARBONATE	38.6
HBA1C	6.1%	Epi cells	1-3 hpf	PT/INR	15.8/1.0
CRP	45.6ug/ml	HIV	Non reactive	Pro calcitonin	0.30ng/ml

CT-Chest: Multiple patchy lung parenchymal air space opacities as detailed could represent lung parenchymal changes associated with vasculitis with possible secondary infection and consolidation pattern.

Echo :
No RWMA
Normal LVEF,EF- 71%
No diastolic dysfunction
C-ANCA –Positive
PR3 ANCA +
Ecg : Normal sinus rhythm , no obvious ST-T abnormality
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CT guided lung biopsy from Rt lower lobe & bronchoscopic biopsy from Rt middle lobe
Nectrotizing granulomatous inflammation with neutrophilic microabscessess and vasculitis
.

Kidney biopsy - Diffuse proliferative glomerulo nephritis with focal segmental necrotising glomerulo nephritis, cellular crescents - pauci-immuno proliferative

DIAGNOSIS

A diagnosis of Granulomatosis with polyangitis is done by Antibody study,radiological and histopathological study.

TREATMENT

- Early treatment is crucial in preventing the devastating end-organ complications of this disease, and often in preserving life.
- While granulomatosis with polyangiitis may involve the sinuses or lung for months, once proteinuria or hematuria develops, progression to advanced chronic kidney disease can be rapid.
- Current practice divides treatment into 2 phases:
  - induction of remission - **Rituximab plus prednisone,Cyclophosphamide and prednisone**
  - maintenance of remission. - **azathioprine,methotrexate,rituximab**

DISCUSSION

- Wegener's granulomatosis is one of the most common forms of systemic vasculitis
- and typically involves medium and small sized blood vessels.
- The clinical syndrome typically begins with a severe, often destructive sinusitis or rhinitis, or with a persistent pneumonitis.
- Signs of renal damage and generalized vasculitis accompany or follow the respiratory disease

CONCLUSION

- As a syndrome, Wegener's granulomatosis can be distinguished from other related categories of arteritis and granulomatous disease.
- No specific etiologic agent has been identified. A mechanism of hypersensitivity apparently operates in the pathogenesis of the illness.
- Pathologic findings include necrotizing granulomatous lesions in the respiratory tract, generalized focal necrotizing vasculitis and glomerulitis.

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References

1. Zeek PM. Periarteritis nodosa and other forms of necrotizing angiitis. N Engl J Med. 1953; 248:764-772.
2. Fauci AS, Haynes BF, Katz P. The spectrum of vasculitis: Clinical, pathologic, immunologic, and therapeutic considerations. Ann Intern Med. 1978; 89:660-676.
3. Hoffman GS, Kerr GS, Leavitt RY, Hallahan CW, Lebovics RS, Travis WD, Rottem M, Fauci AS. Wegener granulomatosis: An analysis of 158 patients. Ann Intern Med. 1992; 116:488-498
4. Harrison's 20th edition
5. Wegener F. Über eine eigenartige rhinogene Granulomatose mit besonderer Beteilgung des Arteriensystems und der Nieren. Beitr Path Anat. 1939; 102:36-38
6.Harrison's Rheumatology