GRANULOMATOSIS WITH POLYANGIITIS

WITH INITIAL PRESENTATION AS ILD.

Presentor- DR SHIVAM ARORA (PG3 MEDICINE MAMC),

Moderators- DR SURESH KUMAR(DIRECTOR PROFESSOR), DR HARPREET SINGH(ASSOCIATE PROFESSOR), DR NIDHI ANAND(ASSISTANT PROFESSOR), DR MAYANK(SR MEDICINE)

HISTORY

A 36 year female patient presented to us with complaint of

shortness of breath for 3-4 weeks which was initially on exertion but progressed to dyspnea at rest over a duration of 3 weeks, associated with dry cough.

- No history suggestive of orthopnea/PND
- No history of pedal edema, palpitations
- No complaint of fever
- No complaint of cough
- No complaint of hemoptysis
- No complaints of increased sweating, bleeding from any site
- No weight loss
- No decreased urine output
- No joint pains, rashes
- No history suggestive of raynauds phenomenon, intermittent claudication

PAST HISTORY

- Patient is known case of hypothyroidism and type 2 diabetes mellitus (on treatment)
- No history of any prior admissions or previous medical interventions

PERSONAL HISTORY

- Mixed diet pattern
- Normal sleep awake cycle
- Chullah user for 20-25 years
- Non alcoholic
- Normal bladder/bowel habits

FAMILY HISTORY

- No similar complaints in family

EXAMINATION

- Conscious, oriented to time place and person
- PR 108 bpm, regular, good volume, no radioradial, no radiofemoral delay
- BP 126/72 mm Hg in right arm In sitting position
- RR 22/min, with use of accessory muscles
- temp- 98.1 F
- Sp02 81% on Room air

- No pallor
- No icterus
- No cyanosis
- No lymphadenopathy
- No pedal edema/facial puffiness
- JVP raised
- No clubbing
- No thyroid swelling

- CVS : s1 s2 were heard with NORMAL apex with loud p2
- Resp: bilateral fine crepts in bilateral(left>right) infrascapular, and infra-axillary area an axillary area
- P/A soft non tender

PROVISIONAL DIAGNOSIS

Hypothyroidism with type 2 diabetes mellitus with

1. LRTI
2. Pulmonary embolism
3. ILD with pulmonary hypertension

Blood investigation	
Hb	10.1
Hct	31.6
TLC	5800
DLC	82/13/4/1
PLT	1.57
TBil	0.6
urea	35

CXR PA findings:
Bilateral lower zone opacity
with central trachea
Ecg- right axis deviation with sinus tachycardia

Urm- normal
D dimer-2400

RAT negative

ABG analysis
PH 7.32
PO2 56
PCO2 38
HCO3 23.2

- 2D ECHO – dilated RA/RV/ moderate PAH/ mild RV dysfunction
  ? Pulmonary embolism
- CECT chest plus CTPA planned
- ANA profile sent

Measurements

LA/Ao 2.8/1.8	LVIDd/LVIDs 4.0/2.6	FS	EF (m-mode) ~60%
RVID	IVSd/IVSs 0.9	PWTd/PWTs 0.8	RA
SVC	IVCi/IVCx	MPA	LPA
RPA	As Ao	Des Ao	MVA
ASD/VSD/PDA		LV Mass	2D-LV Volumes
EF			

2D Echo Description

- Dilated RA/RV
- mod TR
- (RVSP = 50 + RAP)
- Dilated IVC (2.5 cm) ↓ 50% collapsibility

Doppler Data

MV
AV MPA = 3.0 cm
TV TAPSE = 1.4 cm
PV IVC =
HR
PA pressure

Other:

Final impression

Dilated RA/RV / mod PAH / mild RV dysfunction

Rule out CTPA ? Pulmonary Embolism

Signature

- CTPA – feature suggestive of pulmonary artery hypertension with n o evidence of thromboembolism.

Cardiomegaly with mild dilatation of RA, RV

visualised lung fields show mosaic attenuation pattern diffusely with interspersed ground glass densities predominantly in both lower lobes with fibrotic bands

PROVISIONAL DIAGNOSIS

- T2DM/ Hypothroidism/ILD/NSIP pattern/Pulmonary hypertension

TREATMENT GIVEN

- Oxygen therapy
- Injection monocef 1 g IV BD
- T Azee 500 mg OD
- Injection MPS 60 mg OD
- Injection Lasix 20 mg BD

- RA factor- negative
- Ana was negative by elisa and if was still awaited
- crp was raised
- C anca and p anca were awaited

Patient was symptomatically improved and discharged on personal request

Home based oxygen therapy @2-4 L/min

T Wysolone 40 mg OD (to be tapered)

T Pantop 40 mg OD

T Lasix 20 mg BD

READMISSION (2ND)

Patient was lost to follow up on OPD basis and presented after 8-10 weeks with complaints of

Fever x 15 days on and off

Acute exacerbation of shortness of breath for 4-5 Days

Bilateral upper limb skin rashes for 4 days

Cough with blood stained sputum for 4 days

ON EXAMINATION

- PR 110 bpm
- BP 70/42 mm Hg in right arm on sitting position
- RR 26/min, abdominothoracic
- Sp02 79% on Room air

  95% on oxygen@ 10l/min

Bilateral Pedal edema present

- CVS- S1 S2 loud p2
- Respiratory system-fine crepts in b/l interscapular infrascapular areas decreased air entry in rt axillary area
- Bilateral upper limb has palpable purpuric rashes

Department of Pathology

GB PANT INSTITUTE OF POST GRADUATE MEDICAL EDUCATION & RESEARCH, NEW DELHI (GIPMER)

AUTOIMMUNITY REPORT FORM

Name Seema Devi Age/ Sex 36/F LAB NO 928/12

CR No 944954 Ward /Bed 27

Referring Physician Dr S. Kumar Contact telephone No

1. ANA by indirect Immunofluorescence on Hep 2 cells (1:40/1:80/1:100) shows Speckled (1+) nucleolar (1+)

2. ds DNA by ELISA.....

Normal Value: < 25 IU/ml (negative), > 25 IU/ml (positive)

3. Nuclear Antigen Line Assay.....

4. Liver Line Assay.....

5.P ANCA 4.4 U/ml (Negative) Normal Value : ~~<4~~ 9.6 U/ml (Negative)> 9 U/ml (Positive)

6. C ANCA 70 U/ml (Positive) Normal Value : ~~<3.5~~ 6.7 U/ml (Negative) >3.5U/ml (Positive)

7. AntiLKM.....Normal Value : <20 units (Negative), 20 – 24.9 units (Border Line Positive), > 25 units (Positive)

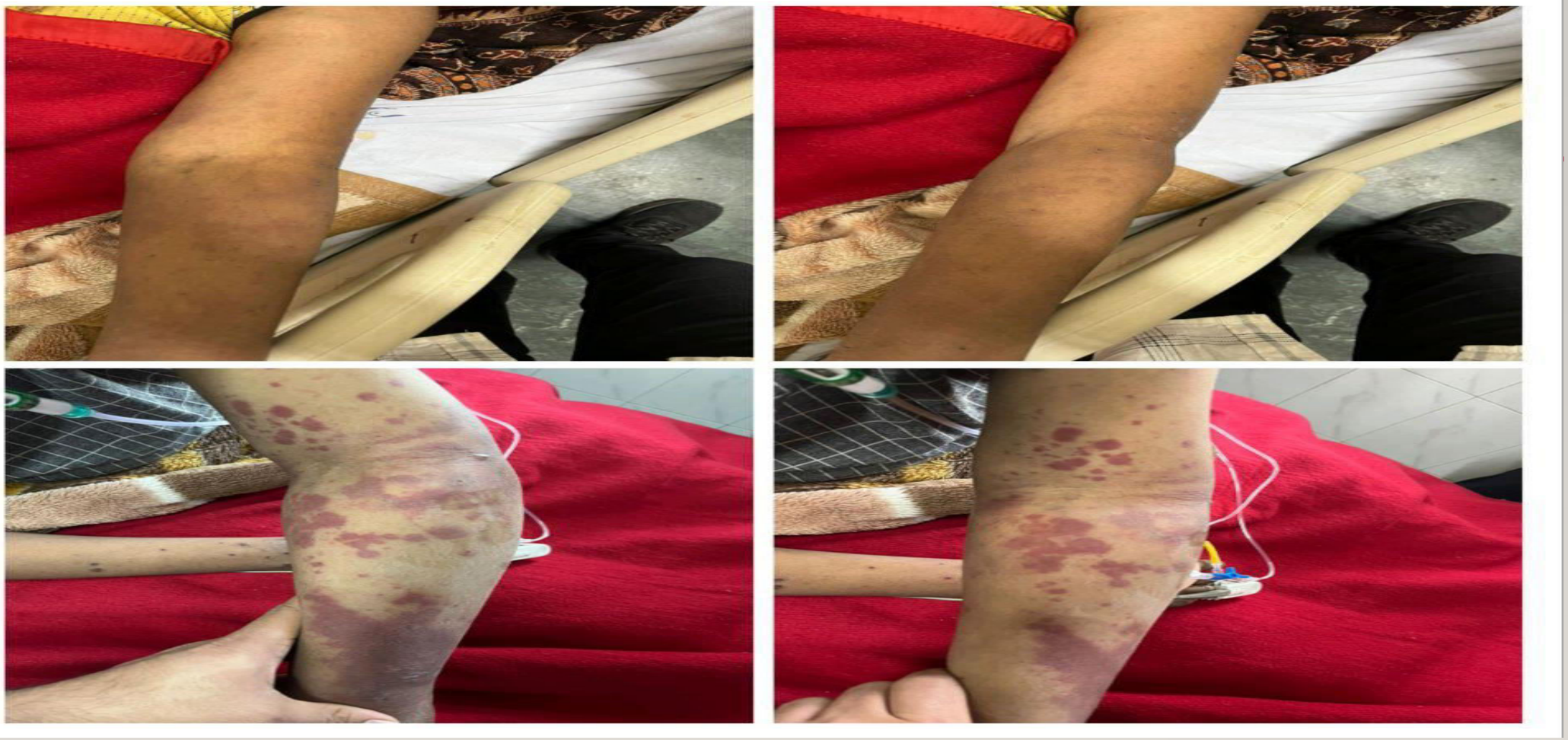
8. AntiGBM.....

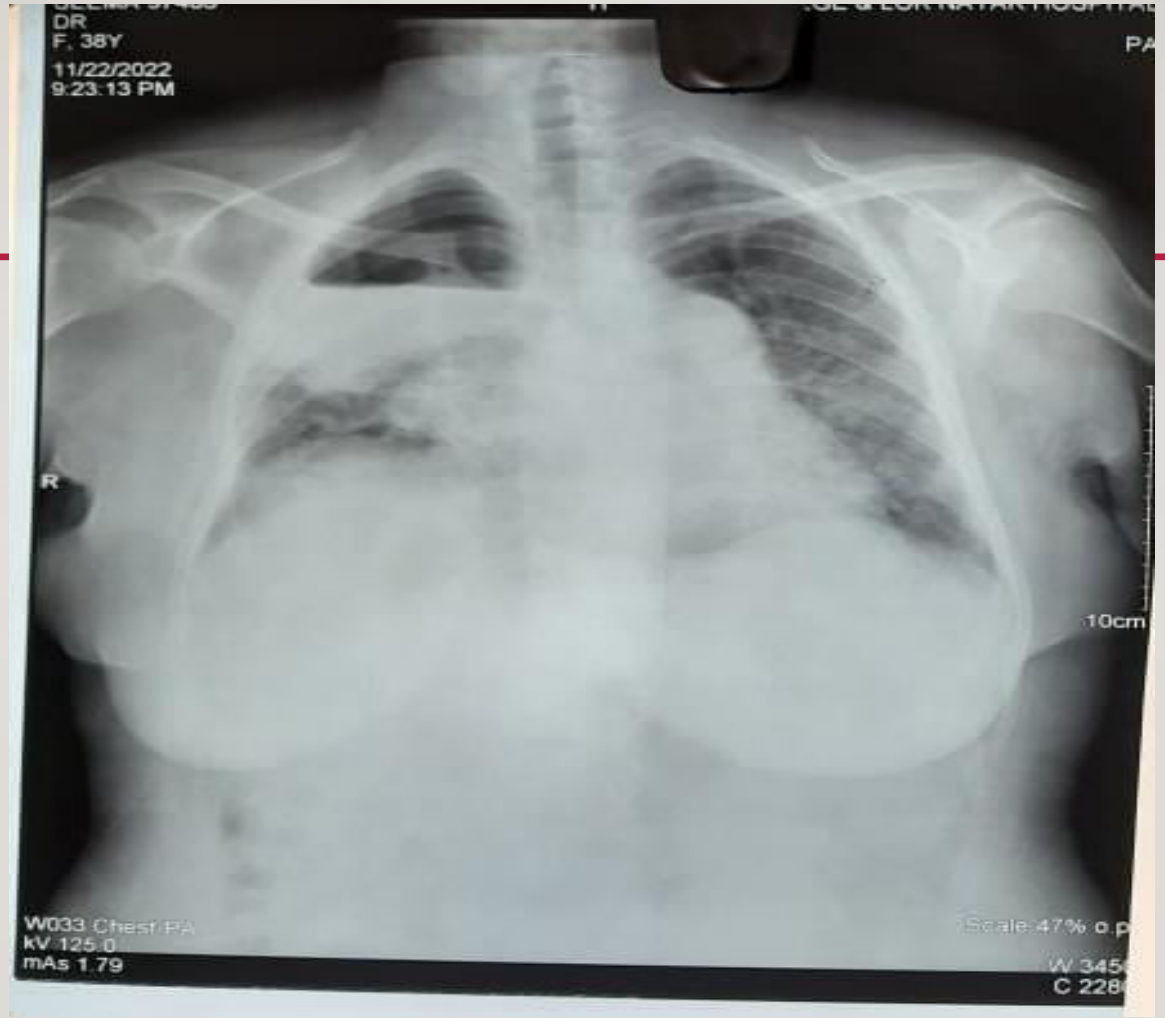
Impression:

LLE

Reported By: Dr. V. V. Babra Date 20/12/12

MGIPRRNO—1625 GB Pant/2017—20 Pads.




DR
F, 38Y
11/22/2022
9:23:13 PM
R
10cm
W033 Chest PA
kV 125.0
mAs 1.79

BLOOD INVESTIGATION:

Investigation	value
Hb	10.1
Hct	33.8
TLC	5700
DLC	82/14/2
PLT	2.51
Tbil/Dbil	0.8/0.5
ALT	16
ALP	178
AST	26
TP/SA	6.8/2.4

investigation	Value
Sputum KOH	Only oral flora seen
Sputum c/s	No growth
Blood c/s	No growth
Repeat blood c/s	No growth

TREATMENT

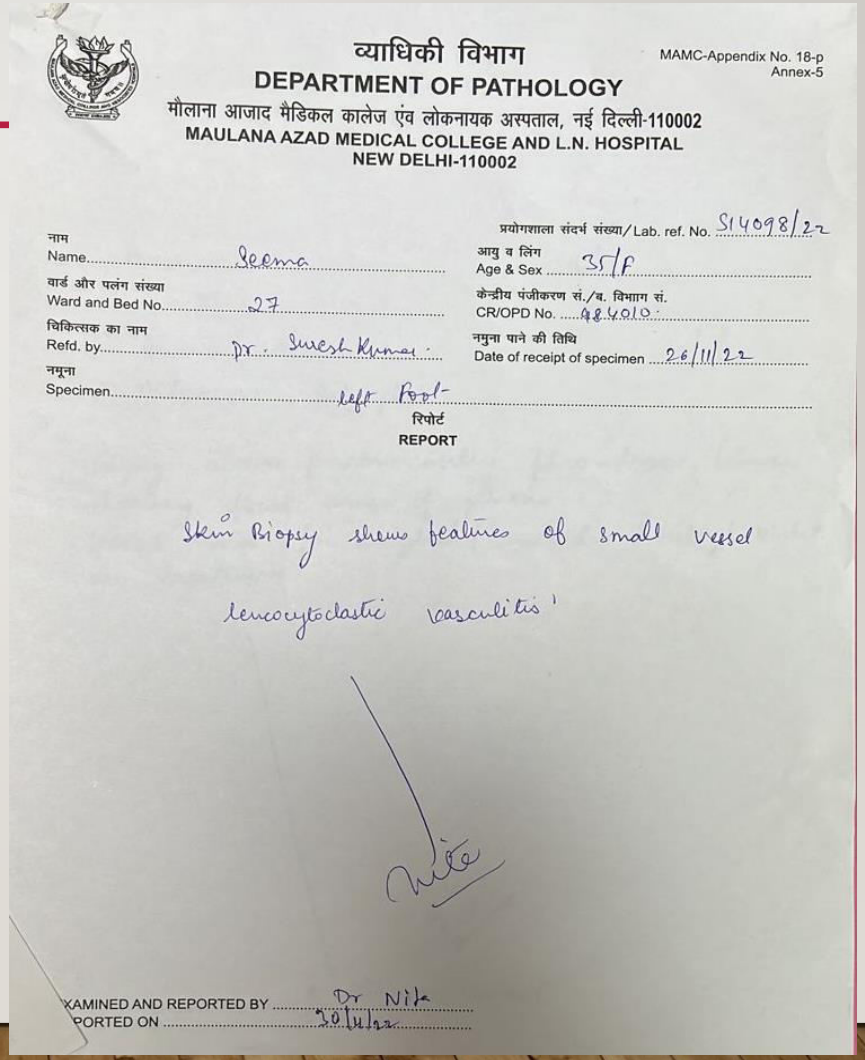
- Inj noradr 80ug/min
- inj clindamycin 600 mg tds
- Inj. tazact

PROVISIONAL DIAGNOSIS

- Hypothyroidism/Type 2 dm with ild with pulmonary hypertension with cor pulmonale with rt upper zone cavity with air fluid level
- ?lung abscess with sepsis with purpura fulminans
- ?wegners with active disease

SKIN BIOPSY REPORT

- Small vessel leukocytoclastic vasculitis



व्याधिकी विभाग
DEPARTMENT OF PATHOLOGY
मौलाना आजाद मैडिकल कालेज एंव लोकनायक अस्पताल, नई दिल्ली-110002
MAULANA AZAD MEDICAL COLLEGE AND L.N. HOSPITAL
NEW DELHI-110002

MAMC-Appendix No. 18-p
Annex-5

नाम
Name Seema

प्रयोगशाला संदर्भ संख्या/Lab. ref. No. S14098/22

वार्ड और पलंग संख्या
Ward and Bed No. 27

आयु व लिंग
Age & Sex 35/F

चिकित्सक का नाम
Refd. by Dr. Suresh Kumar

केन्द्रीय पंजीकरण सं./ब. विभाग सं.
CR/OPD No. 484010

नमूना
Specimen Left Foot

नमूना पाने की तिथि
Date of receipt of specimen 26/11/22

रिपोर्ट
REPORT

Skin Biopsy shows features of small vessel leucocytoclastic vasculitis.

EXAMINED AND REPORTED BY Dr Nita
REPORTED ON 30/11/22

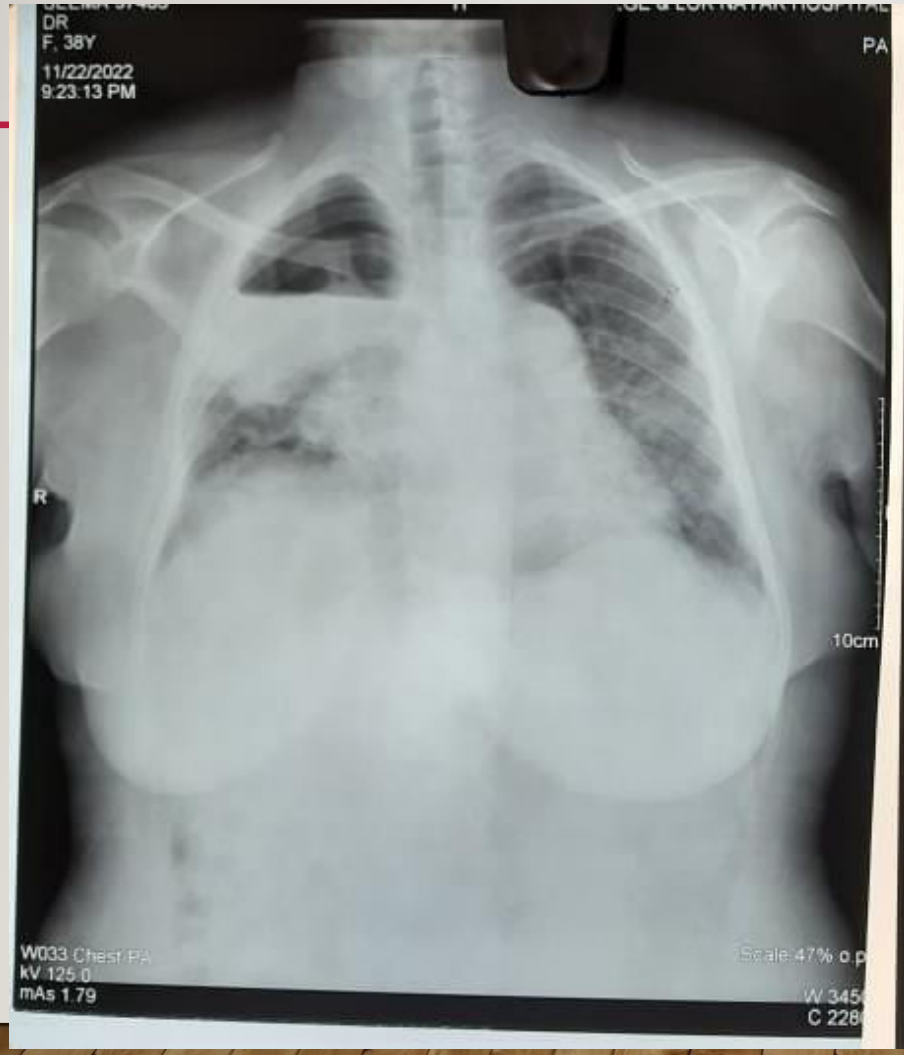
FINAL DIAGNOSIS

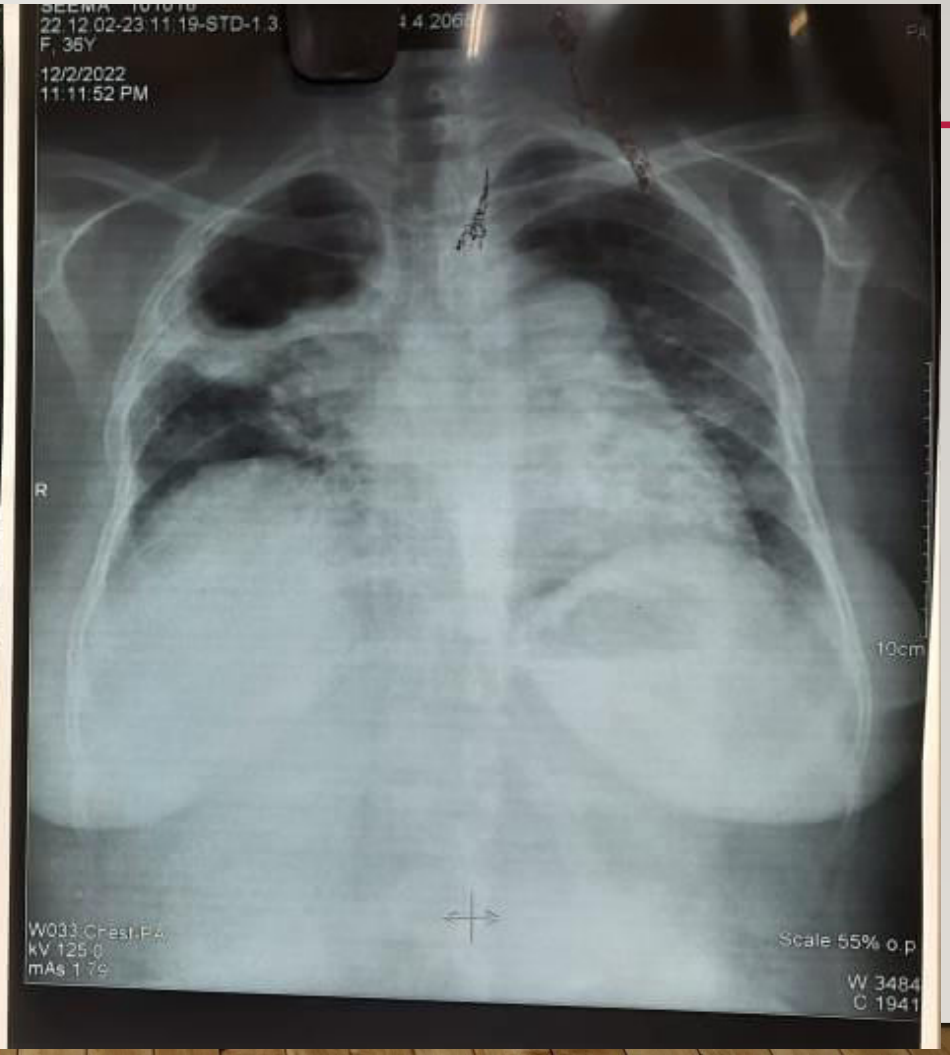
- Hypothyroidism/ Type-2 DM/ILD/Pulmonary hypertension /ILD/ Right upper zone cavity /c-ANCA positive small vessel vasculitis likely Wegner's

TREATMENT GIVEN

- Inj methylprednisolone 1 gm i/v OD for 3 days followed by-
- Tab prednisolone 60mg OD
- Tab cyclophosphamide 150mg OD
- Inj Tranexa 500 mg i/v TDS
- Tab eltroxin 150 mcg OD BBF
- Tab metformin 500 mg BD

CHEST X-RAY FILMS





FOLLOW UP

- Patients oxygen requirement came down
- Nor adr was tapered
- skin lessions started improving
- cough and hemoptysis resolved

WHY THIS CASE?

- ILD with Wegners is relatively uncommon .
- There was no renal involvement and ENT involvement

The lung is affected in several distinct ways. Pulmonary nodules, which are necrotizing and frequently cavitate (Fig. 94.5), often occur before or in the absence of systemic vasculitis and thus are a common feature of "localized"[78,79] or "limited"[80] GPA. Nodular disease is often asymptomatic or merely produces cough. Alveolar hemorrhage, characterized by mild to life-threatening dyspnea and hemoptysis, is also common but is clinically and radiographically distinct from nodular disease.[82,83] Other well-described pulmonary manifestations include pleuritis[56] and endobronchial lesions distal to the subglottis[84]; the frequency of the latter may be underestimated because they can only be definitively diagnosed by bronchoscopy. Clinically significant pulmonary fibrosis and bronchiectasis is relatively uncommon in GPA because parenchymal lung disease generally heals without scarring.

Thank you