WELCOME TO THE GRAND ROUND PRESENTATION

A 38-year-old woman with recurrent leg ulcer

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Salient Feature

Mrs.X, 38 years old, homemaker, normotensive, non diabetic, hailing from Brahmanbaria admitted into the hospital on 06.02.24 with the complaints of

- Leg ulcers for 3 months
- Fatigability for same duration

- Three months back she noticed multiple painful pigmented elevated lesions on her lower limbs. The lesions showed progressive color change from red to dark. She also mentioned that initially there were some blisters over her legs. There was no itching, burning sensation, discharge or exfoliation from the lesions.

- Few days later she developed ulcers on the outer aspect of her right lower leg and on the left foot. The ulcers were progressively increasing in size, painful with oozing of serous fluid, but there was no bleeding. These type of lesions appeared recurrently on the legs for the last 3 years during winter.

- She also complained of fatigue for the last 3 months which was gradually increasing and she couldn't do her regular household activities properly due to this problem. There was no diurnal variation of her fatigability.

- On query she gave history of recurrent pain and swelling of the small joints of hands involving the proximal interphalangeal joints and both knee joints with morning stiffness which lasted for around 2-3 hours and improved with activity for last 3 years.

- On further query she stated that she had recurrent history of nasal crusting and epistaxis for 3 years which was more during winter season.
- She also mentioned sequential color change of her fingers and toes (pale>blue>red) on exposure to cold.

- There was no history of fever, shortness of breath, cough, hemoptysis, pain in abdomen, jaundice, oral ulcer, photo sensitivity, dryness of eyes or mouth, pain in the eyes. Her bowel and bladder habits were normal.

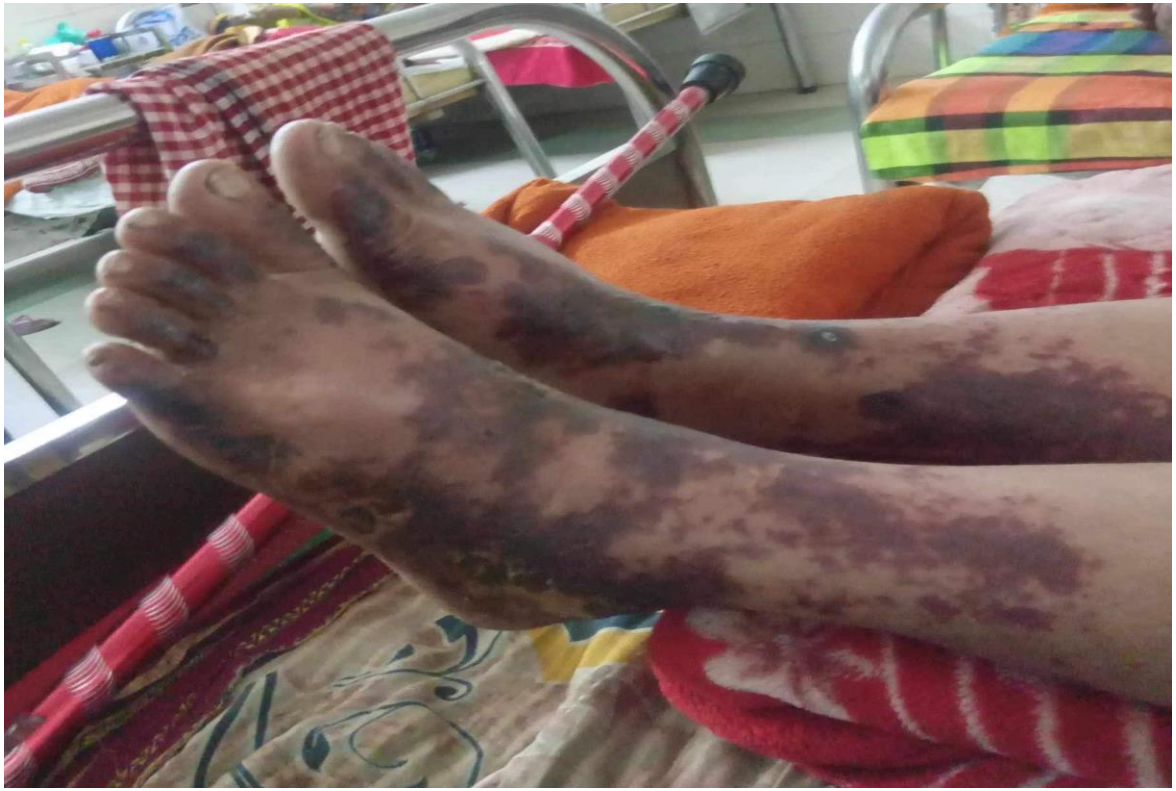
- For her problems she consulted with many physicians and was admitted into a tertiary care hospital 2 months prior to this admission.
- She was taking methotrexate 15 mg once weekly for last 2 years and prednisolone 10 mg once daily for last 3 months.

- She had no history of allergic conditions

- Her menstrual cycles were regular with normal flow. She had three children, her one child passed away at the age of 5 years due to leukemia and another one at the age of 17 days due to pneumonia. She had no history of abortion and her pregnancy period was uneventful.

- On general examination she had malar rash. Her pulse was 80 beats/min, arteria dorsalis pedis and anterior tibial artery pulsations were absent bilaterally. Her blood pressure was 110/65 mmHg and respiratory rate was 16 breaths/min. Her temperature was 98 degree F.

- During admission skin examination revealed there were multiple painful palpable purpura which were mostly hyper pigmented and some were erythematous, involving lower extremities.

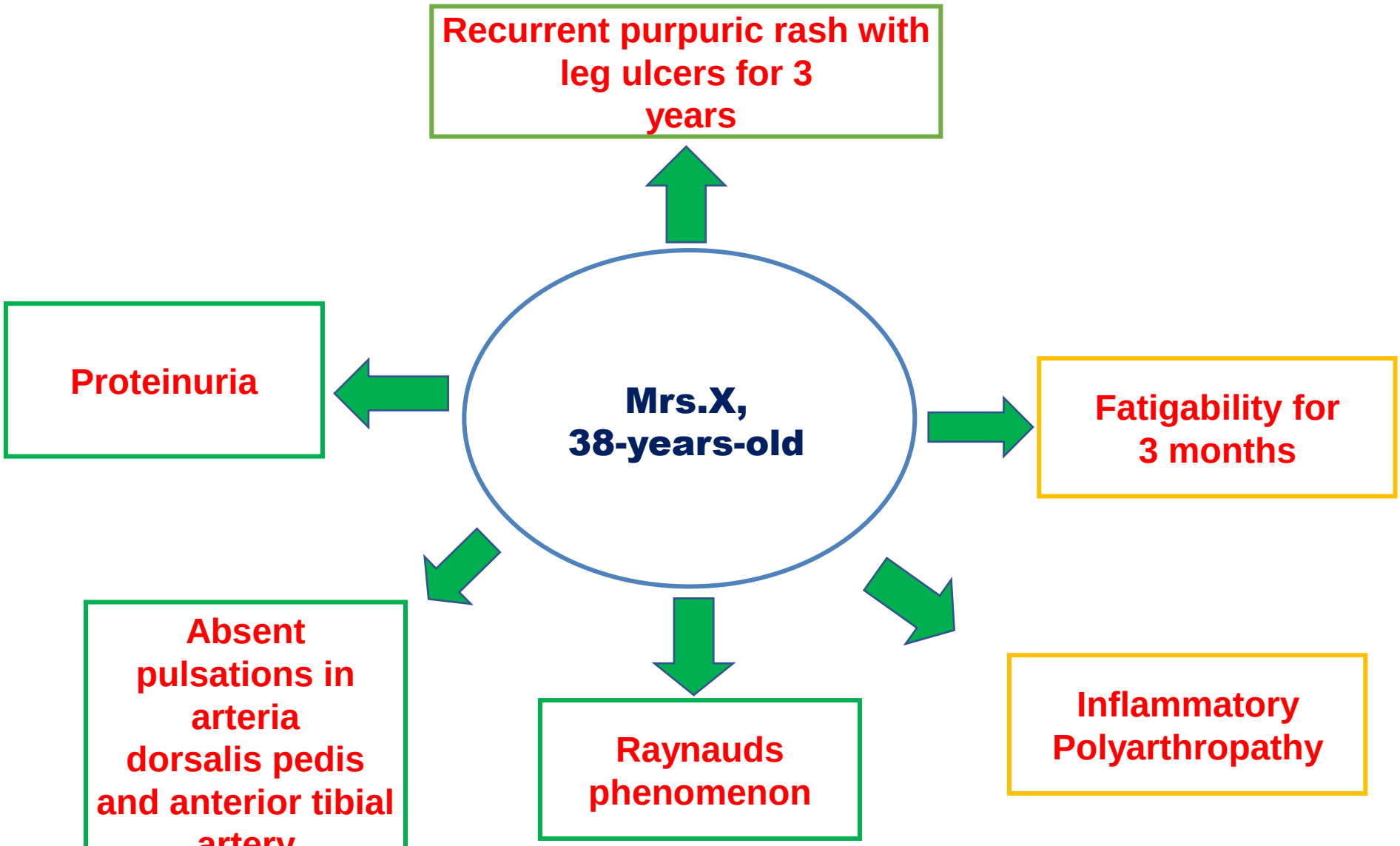


- On 22.2.24- There was a solitary ulcer present over the right lateral malleolus, 4 cm × 2 cm in diameter with irregular margin, necrotic floor, tender base and oozing. Some small ulcers were present over the left foot. Ulcers were surrounded by hypo and hyper pigmented skin.

- Bedside urine dipstick test revealed protein : ++, RBC : NIL.

- Other systemic examination revealed no abnormality.

Problem List:



Provisional Diagnosis ???

Provisional Diagnosis

Systemic lupus erythematosus with Vasculitis

Differential diagnoses

1. Granulomatosis with polyangitis
2. Rheumatoid arthritis with vasculitis
3. Polyarteritis Nodosa

INVESTIGATIONS

CBC

Date	05.06.22	21.8.22	Unit
Haemoglobin	13	10.7	g/dL
ESR	26	95	mm in 1st hour
RBC	$4.4X\ 10^6$	$3.75X\ 10^6$	/cumm
White blood cell	7500	7500	/cumm
Platelets	$250X10^3$	$350X10^3$	/cumm
Hct	39.5	32.4	%
MCV	89.1	86.4	fl
MCH	29.3	28.5	pg

CBC

Date	09.12.2023	29.1.24	Unit
Haemoglobin	9.4	11.9	g/dL
ESR	86	20	mm in 1st hour
RBC	3.3X 10^6	4X 10^6	/cumm
White blood cell	8000	9830	/cumm
Platelets	390 X10^3	360X10^3	/cumm
HCT	27.3	35.9	%
MCV	82.7	87.1	fl
MCH	28.5	28.9	pg

Peripheral blood film

9/12/2023

- **RBC** : Normocytic & normochromic with rouleaux
- **WBC** : Mature with normal count and distribution
- **Platelet** : Normal

- **COMMENT**: Normocytic normochromic anaemia,moderate.

Urine R/E

	05/06/22	11/01/23	18/01/23	9/12/23	14/12/23	29/1/24
Appearance		Light Turbid				
Specific gravity		1.032				
Protein	Trace	+++	Trace	+	Trace	+
Bilirubin		1				
Leukocyte		+				
Blood					Present	Trace
Epithelial cells	0-2/HPF	12-15/HPF	1-2/HPF	1-3/HPF	1-3/HPF	0-2/HPF
Pus cells	0-2/HPF	8-12/HPF	3-4/HPF	1-2/HPF	1-2/HPF	1-2/HPF
RBC		1-2/HPF	12-15/HPF			0-2/HPF

Urine C/S

- **18.01.2023** : No growth

S. electrolytes

Parameters	13/02/24 (mmol/L)
Sodium	137
Potassium	3.86
Chloride	95
Carbon dioxide	28
Bi-carbonate	26

Test	09.06.22	21.08.22
ALT(SGPT)	10 U/L	9 U/L

Test	8/6/22 (mg/dl)	21/8/22 (mg/dl)	04/09/23 (mg/dl)	9/12/23 (mg/dl)	30/1/24 (mg/dl)
Serum Creatinine	0.86	1.12	0.94	1.05	0.95

Test	Value	Reference Value
Urinary Total Protein(UTP)24 hrs	0.76 gm/day	<0.2 gm/day
24hrs Urine Volume	1600 ml/day	1200-1800 ml/day
Urinary Protein Creatinine Ratio	1.29	<0.2 Normal

Investigations

Test	11.01.23	11.12.23	17.12.23
ANA	Negative	Negative	Negative

Test	11.1.23	29.1.24
Anti-ds-DNA	Negative	Negative

Test	8.6.22	14.2.24	Reference Value
Anti-CCP	4.52	6.1	<4.00 U/ml

Test	05.06.22	29.01.24
RF	Negative	Negative

Investigations

- **ENA profile** : Negative

Test	Result
ENA Profile	
Nucleosome	Negative
Histone	Negative
SmD1	Negative
U1-SM/RNP	Negative
SS-A/Ro60KD	Negative
SS-A/Ro52KD	Negative
SS-B/La	Negative
Scl70	Negative
CENP-B	Negative
Jo-1	Negative
Po (RPP)60	Negative
PCNA	Negative
PM-Scl	Negative
Mi-2	Negative
Ku	Negative
AMA-M2	Negative
DFS70	Negative
ds-DNA	Negative
RNP-A	Negative
RNP-C	Negative

Method : LIA

Investigations

- **Anti-Phospholipid IgG and IgM** : Negative

Test	11.1.23	11.12.23
C-ANCA	Negative	Negative
P-ANCA	Negative	Negative

Investigations

Test	20.12.23	17.2.24	Bio. Ref. Interval
Serum C3	1.51		0.90-1.80 g/L
Serum C4	O.245	0.254	0.10-0.40 g/L

Investigations

Test	05.06.22 (mg/L)	22.08.22 (mg/L)	09.12.23 (mg/L)
CRP	6.37	4.44	3

Test	06.06.22 (mmol/L)	14.12.23 (mmol/L)
RBS	6.5	6.1

Investigations

❖ **Viral markers:**

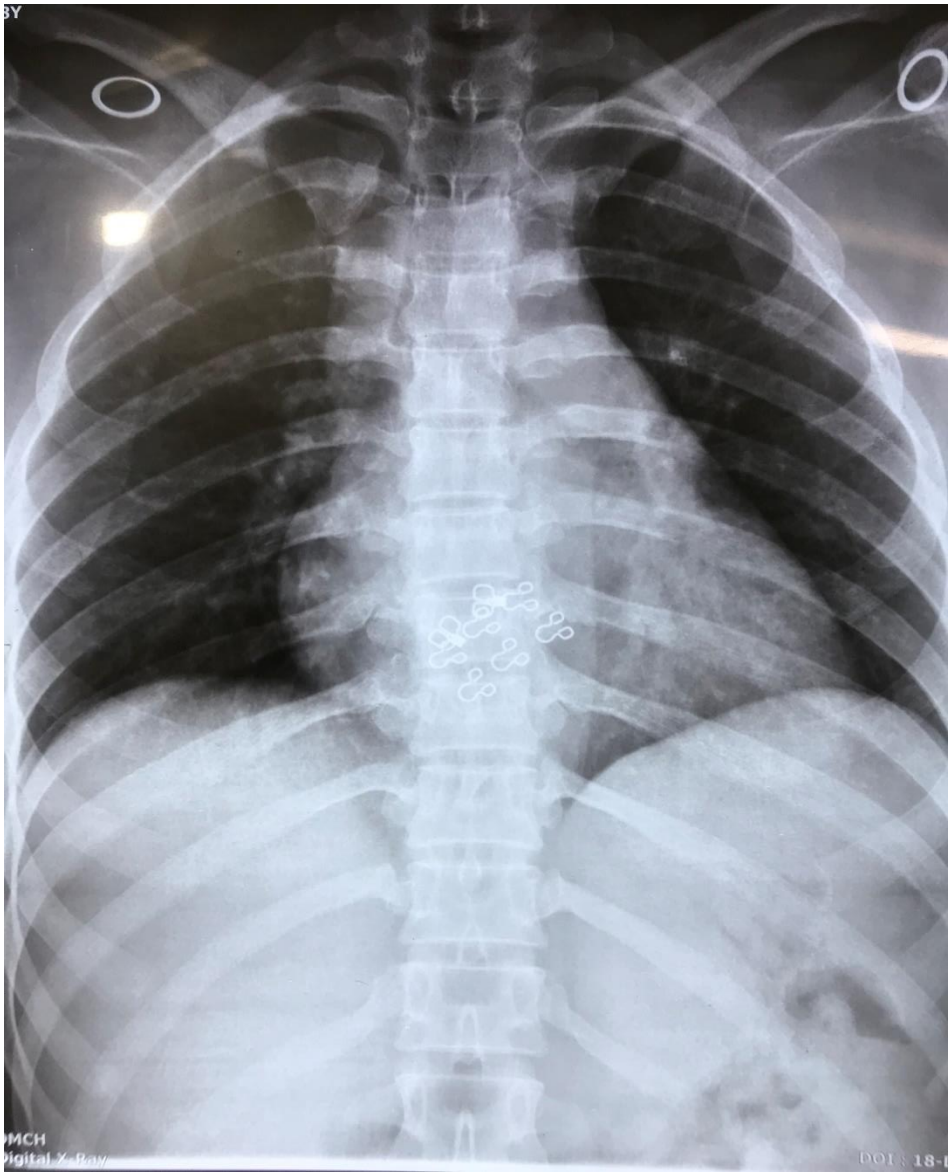
	4.9.23	17.12.23	10.2.24	20.2.24
HBsAg	Negative	Negative		
AntiHCV	Negative		Negative	
AntiHBc(Total)				Positive
Anti HIV(1+2)				Negative

Hormone analysis

Hormone	4/9/23	13/08/22	Normal value
TSH	5.73		0.55-4.78 mIU /L
FT3		3.54	2.89-4.88 pmol/L
FT4		14.34	9-19.05 pmol/L

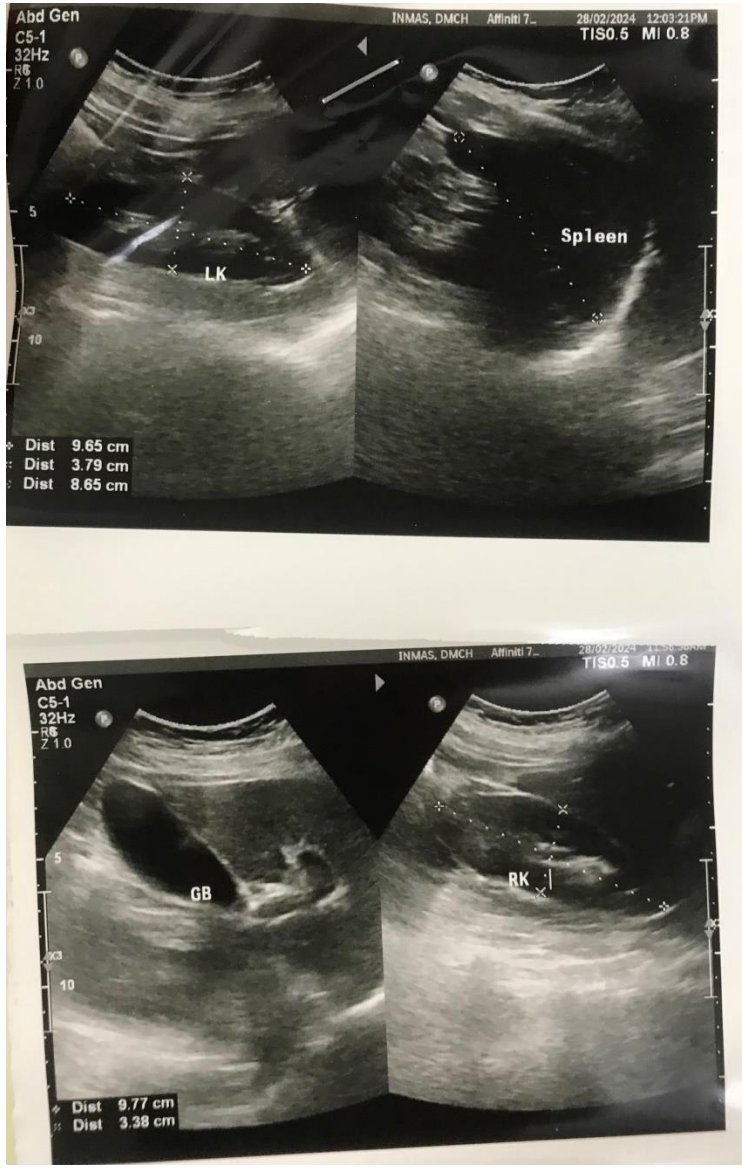
Chest xray P/A view

11/01/2023 : Normal



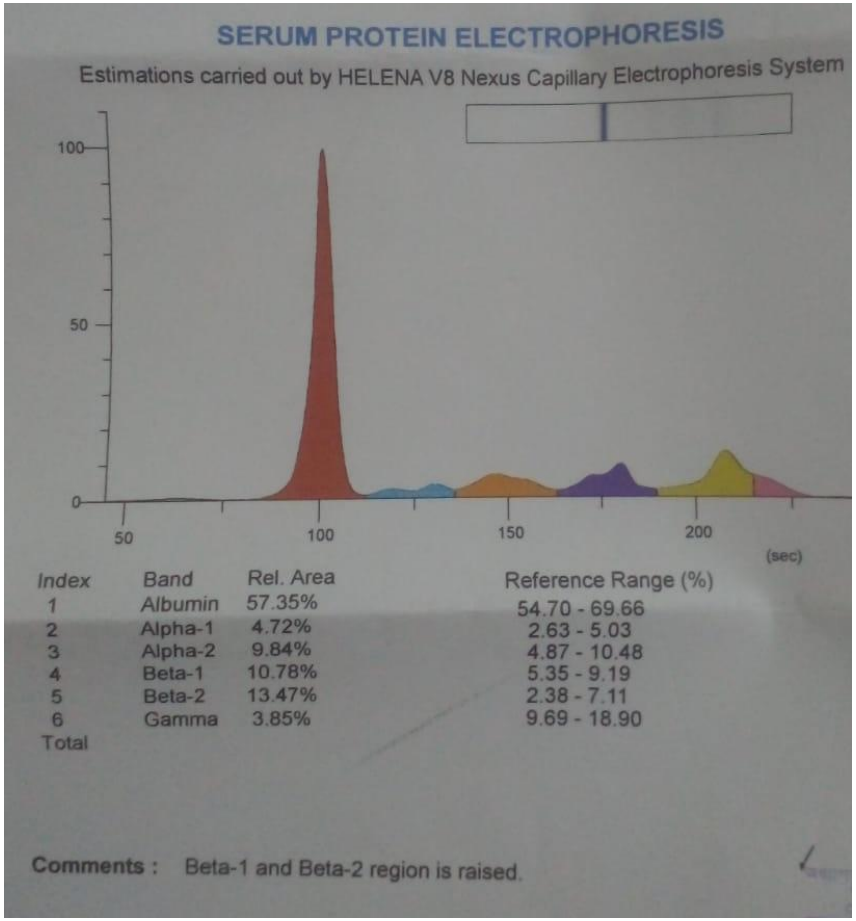
USG of whole abdomen

- No abnormality is seen



Serum Protein Electrophoresis

Comments: Beta-1 and Beta-2 region is raised



Duplex Study of Both lower limb vessels

20/8/23 :

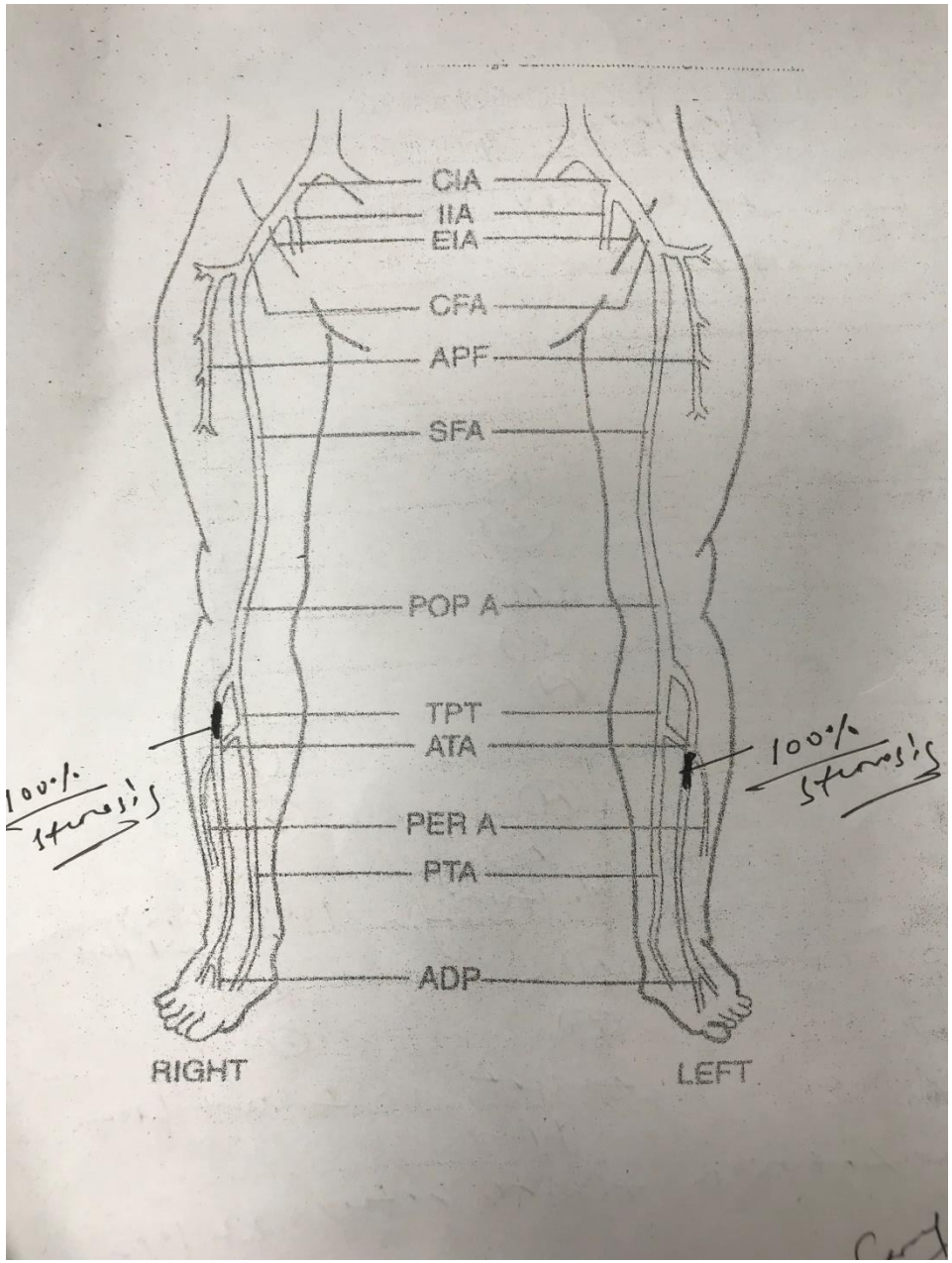
- Mild Atherosclerotic changes were seen at all the arteries of both lower limbs without any significant flow reduction.
- Apparently normal arterial and venous system at both lower limbs.

25/02/24

- Normal arterial and venous system at both lower limbs.
- Hemodynamically non-significant mild athersclerotic changes noticed in major arteries of both lower limbs.

Peripheral (Lower Limb) Angiography Report

- **Impression :** Anterior tibial artery-
- Right leg : origin 100% stenosis
- Left leg : mid 100% stenosis


CIA
IIA
EIA
CFA
APF
SFA
POP A
TPT
ATA
PER A
PTA
ADP
RIGHT
LEFT
100% stenosis
100% stenosis

Skin tissue for histopathology and DIF

- Sections from skin show moderate hyperkeratosis with moderate acanthosis with elongation of rete ridges. There is a large subcorneal bulla is present containing fibrinous material without any acantholytic cells. Dermis shows dense perivascular infiltration of lymphocytes and neutrophils. No cytoplast is present.
- DIF : No deposit of IgG, IgA, IgM, C3 or C1q is present.

- **Diagnosis** : Consistent with pemphigus foliaceus.

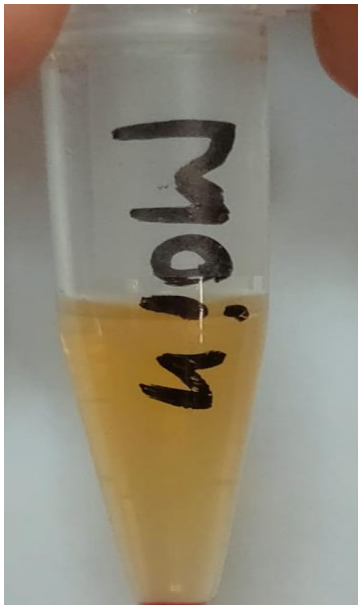
WHAT NEXT ??

Cryoprecipitate test

- Patients blood Collected at >37^{0}C→centrifuged →serum separated Cooled at 4^{0}C → cryo-precipitate appeared → on rewarming dissolves again

Refrigerating 4^{0}C

Rewarming



Control

Confirmed diagnosis

Cryoglobulinaemic vasculitis

ANY SUGGESTIONS??


THANK YOU!