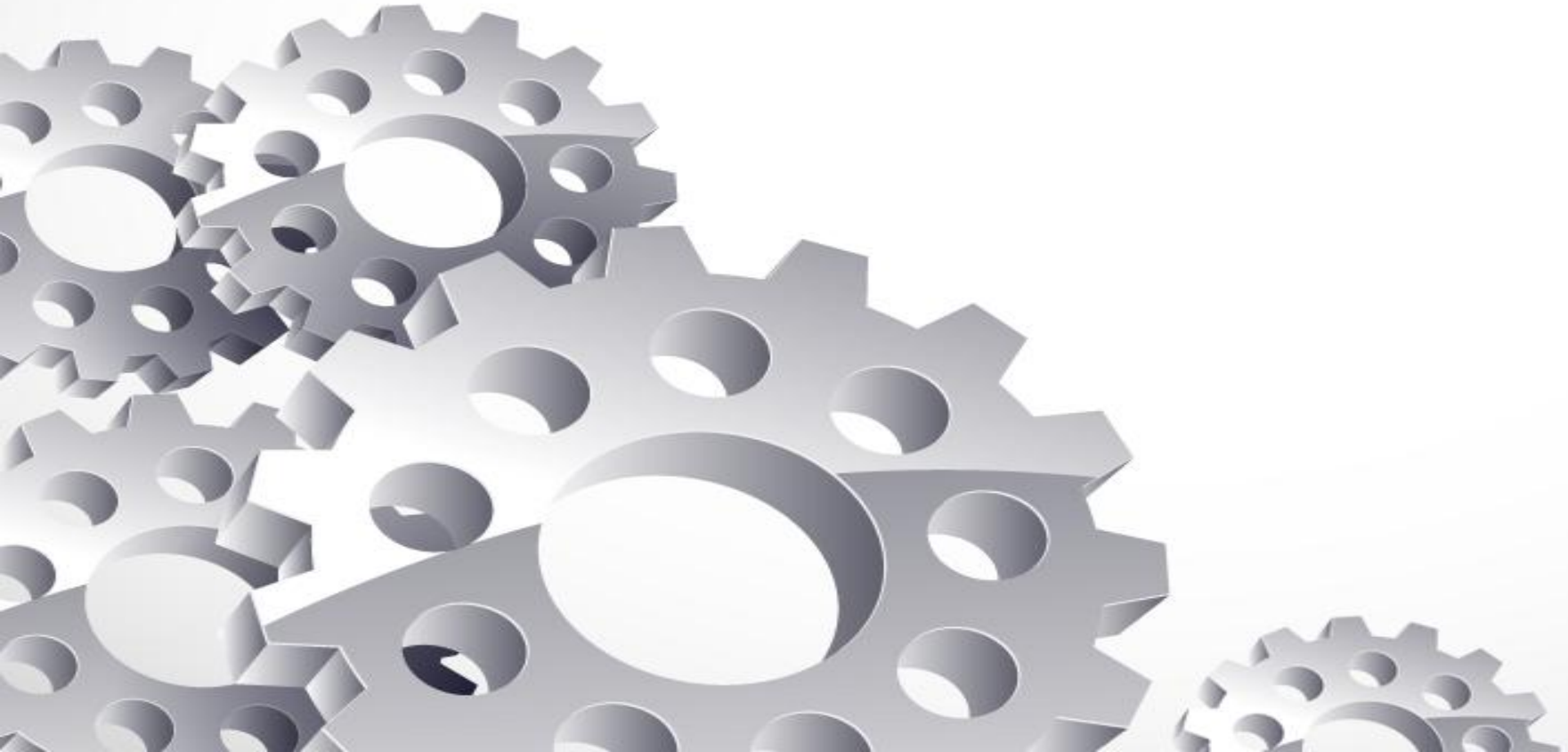
MEDIASTINAL LYMPHADENOPATHY



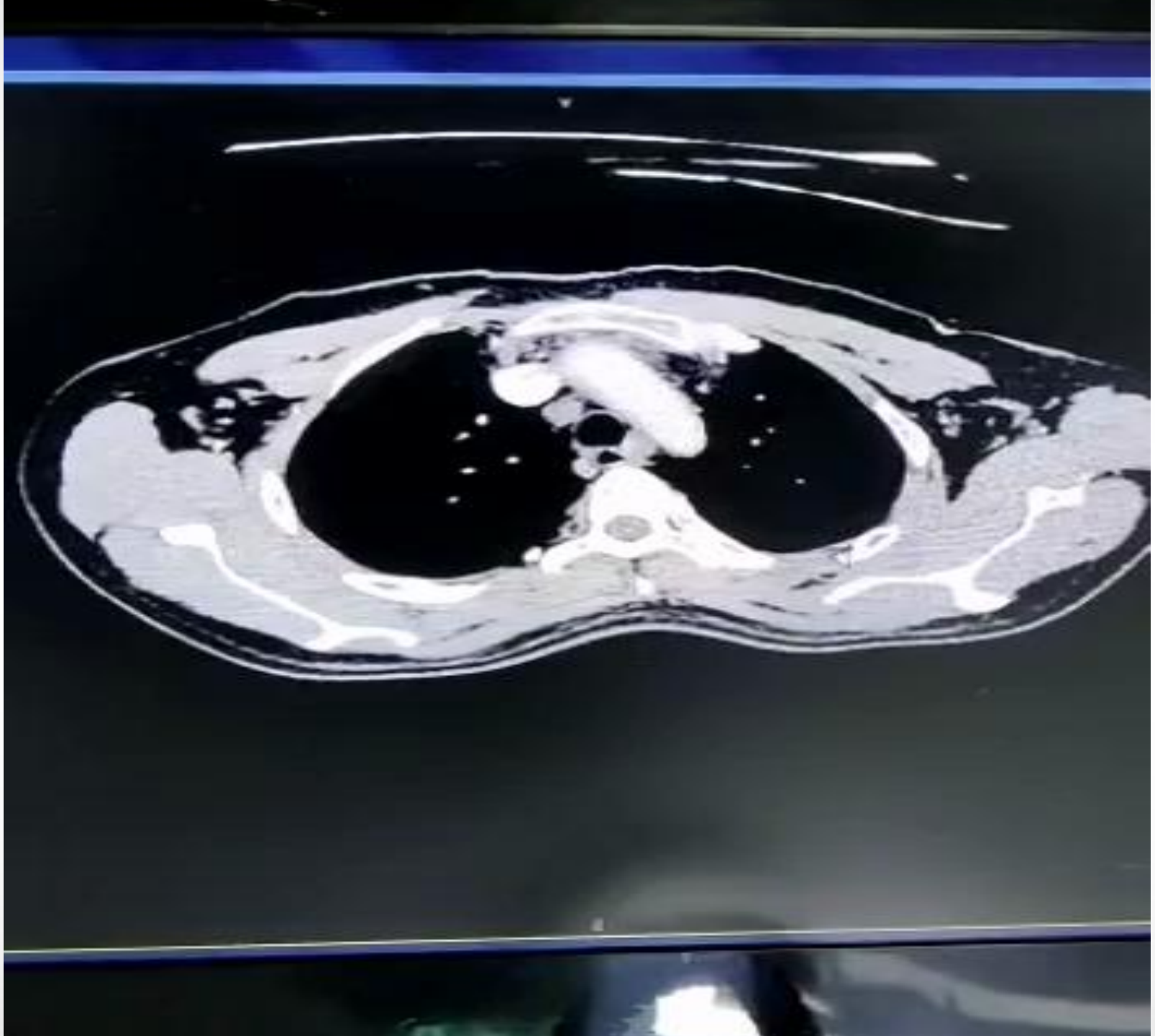
- 45 year old male, farmer by occupation, non - smoker
- H/O **fever for 10 months**, high grade , intermittent nature, with no diurnal variation.
- H/O **weight loss 4 kg over 8 months**
- There is no H/O cough, hemoptysis, wheeze, chest pain.
- Patient was initially evaluated outside.

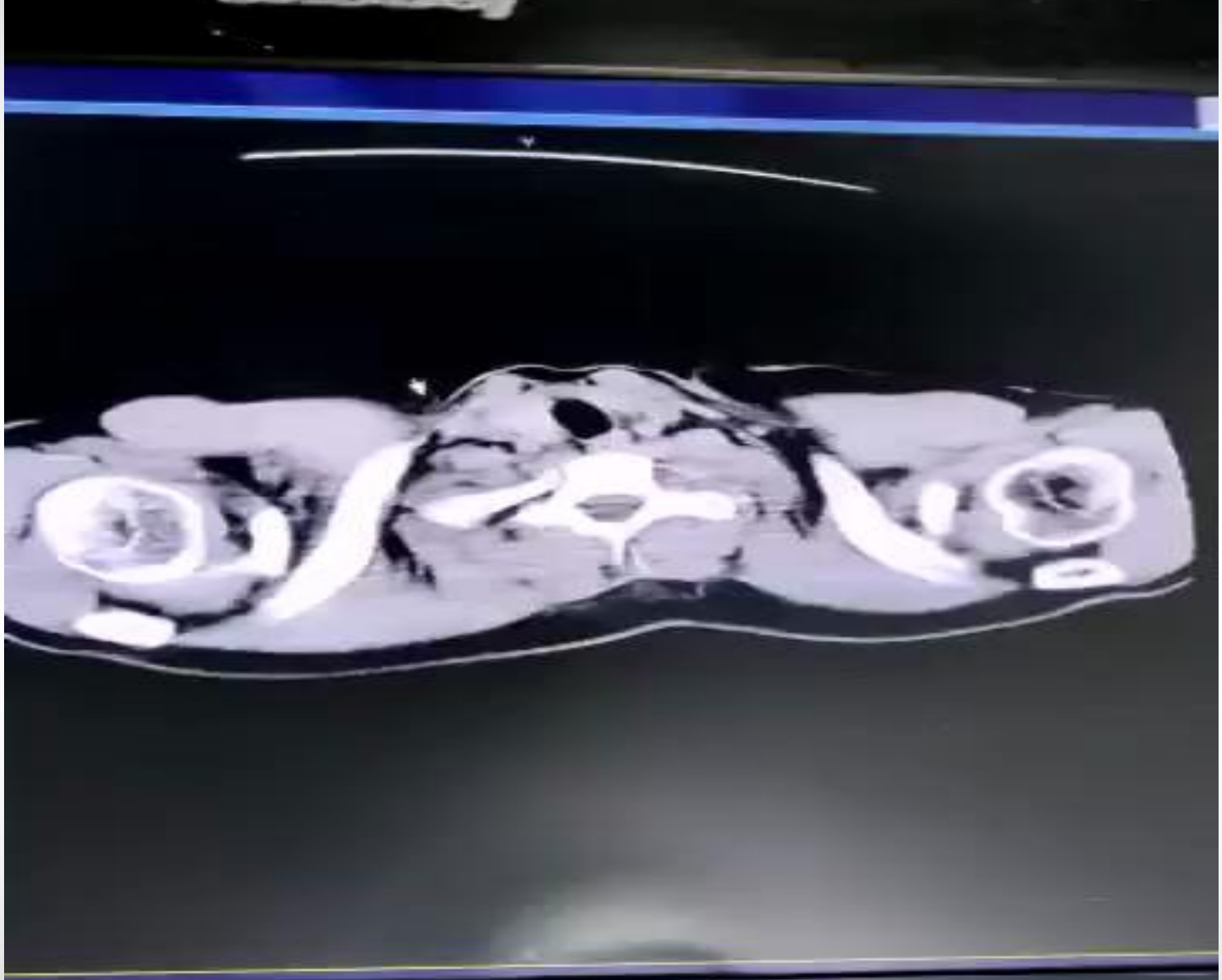
- **CT scan showed multiple enlarged homogenous discrete lymph nodes in pre tracheal,bilateral para tracheal , aorto pulmonary and subcarinal region.**

DIFFERENTIAL DIAGNOSIS

- TUBERCULOSIS
- SARCOIDOSIS
- LYMPHOMA
- METASTASIS FROM A PRIMARY TUMOUR.

- He had been given empirical anti tubercular treatment for 3 months, but fever was not subsiding.
- Hence he was advised to undergo biopsy of the lymph node.





ACE - 54.3	SERUM CALCIUM 9.27
PT - 17.3 INR 1.45	FERRITIN 1322
TOTAL PROTEIN - 7.96 ALBUMIN - 3.34 GLOBULIN - 4.62	Hb - 9
bil - 1.04 SGOT/SGPT = 16/26	iron 16 TIBC - 181

Patient underwent biopsy under FFP cover.

Specimen No:
D20/4185

Clinical History:
Fever with lymphadenopathy

Specimen:
CT-guided biopsy from subcarinal lymph node

Gross:
A, B. Received in formalin and normal saline respectively is a specimen labelled as "CT-guided biopsy from subcarinal lymph node" consisting of multiple greyish white linear cores and fragmented bits longest cores measuring 0.7 cm in length. A1, B1- All.

Microscopy:
Sections demonstrate portion of a lymph node with a few follicles and interfollicular areas, the latter composed of a mixture of small lymphocytes and plasma cells and occasional immunoblasts. A few Russell bodies are also seen. No evidence of RS type cells, necrosis and granulomas.
Immunohistochemical stains are performed using appropriate positive and negative controls. Cd3 and CD5 positive T-cells are noted in the paracortical areas, whereas CD20 positive cells are seen in the follicles where the follicular dendritic networks are highlighted with CD23. CD30 stains scattered few immunoblasts. CD138 highlights plasma cells that show a mixture of kappa and lambda light chain stained plasma cells. CD10 is negative. Ki67 shows increased proliferative activity in germinal centers and is low in the remaining tissue.

Final Diagnosis:
A, B. Subcarinal lymph node, CT-guided biopsy:
Portion of lymph node with lymphoplasmacytic infiltrate (see note).

Note:
No definite evidence of lymphoma is seen on histology and immunohistochemistry in the submitted biopsy. No e/o granulomas and metastatic malignancy. Plasma cells present appear polyclonal. Correlation with clinical findings as well as laboratory data including serum protein electrophoresis is suggested.
[illegible] is in agreement (MM, VG).

Nature of material :

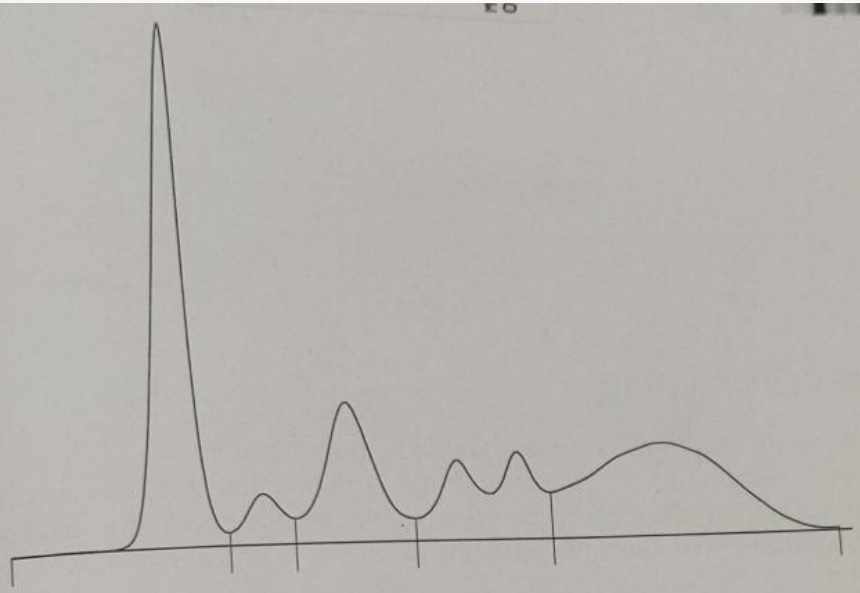
2 paraffin blocks labelled D20/4185 A1, B1 of CT guided biopsy from a subcarinal lymph node.

Microscopic Description :

Benign reactive lymphoid tissue.

There are no granulomas present in the serially sectioned biopsy.

The biopsy was also screened for expression of CD20, CD3, CD5, CD10, CD23 & CD30 with results as expected in a reactive process.



Serum protein electrophoresis

Fractions	%	Ref. %		Conc.	Ref.Conc(g/dl)	
Albumin	34.5	53.8 - 65.2	<	3.2	3.5 - 4.9	A/G Ratio: 0.53
Alpha 1	4.1	1.1 - 3.7	>	0.4	0.1 - 0.4	T. P.: 9.14 g/dl
Alpha 2	15.7	8.5 - 14.5	>	1.4	0.4 - 0.8	
Beta	15.0	8.6 - 14.8	>	1.4	0.6 - 1.0	
Gamma	30.7	9.2 - 18.2	>	2.8	0.8 - 1.4	

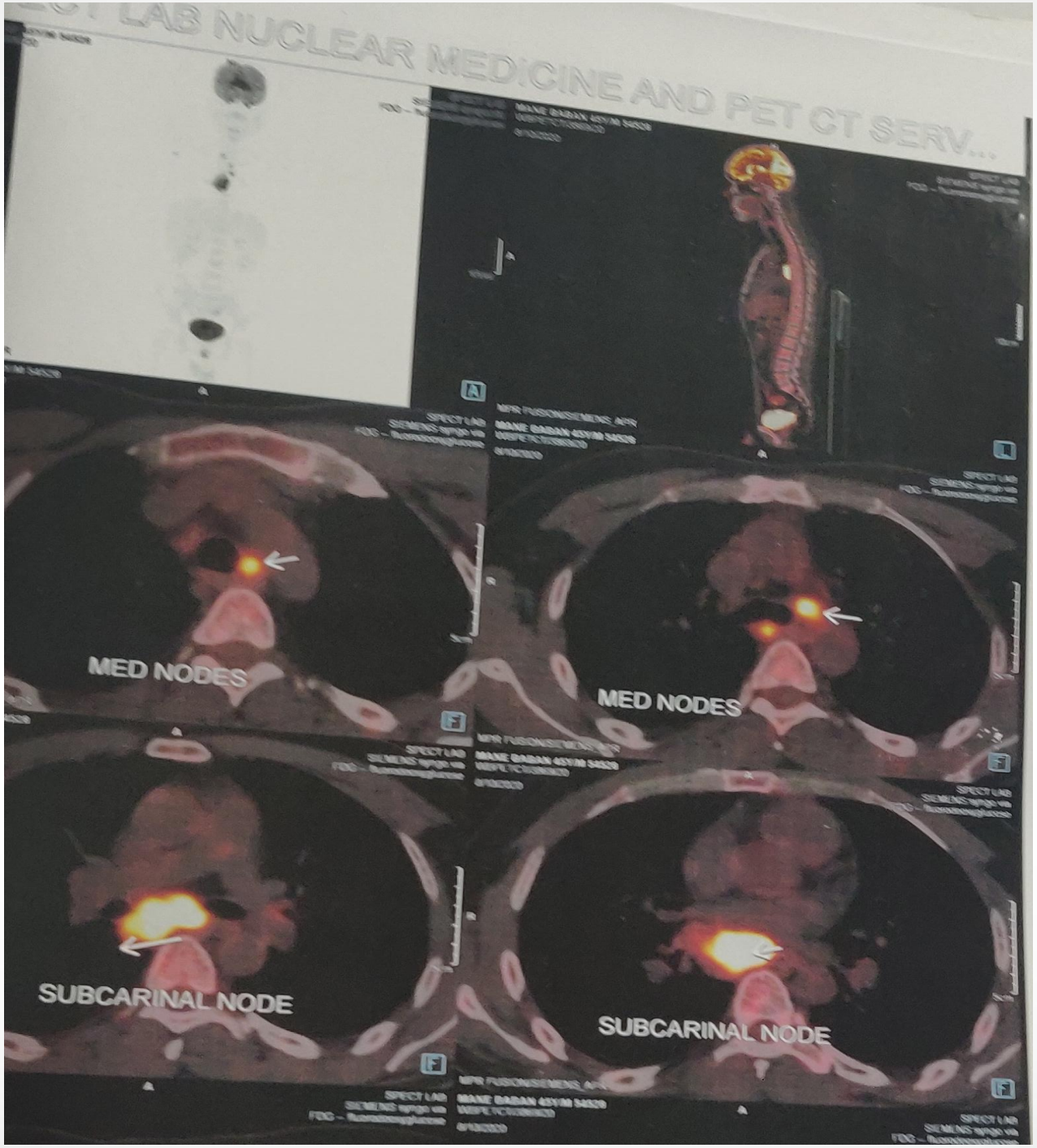
Comments : **Polyclonal Gammopathy**

- Patient came to us for further opinion.
- O/E patient was hemodynamically stable.
- No constitutional symptoms.

- Repeat cbc shows hb 8.6 , total count 6500, platelet 478.
- morphology - few tear drop cells, few fragmented rbc, anisocytosis, microcytosis, hypochromia.

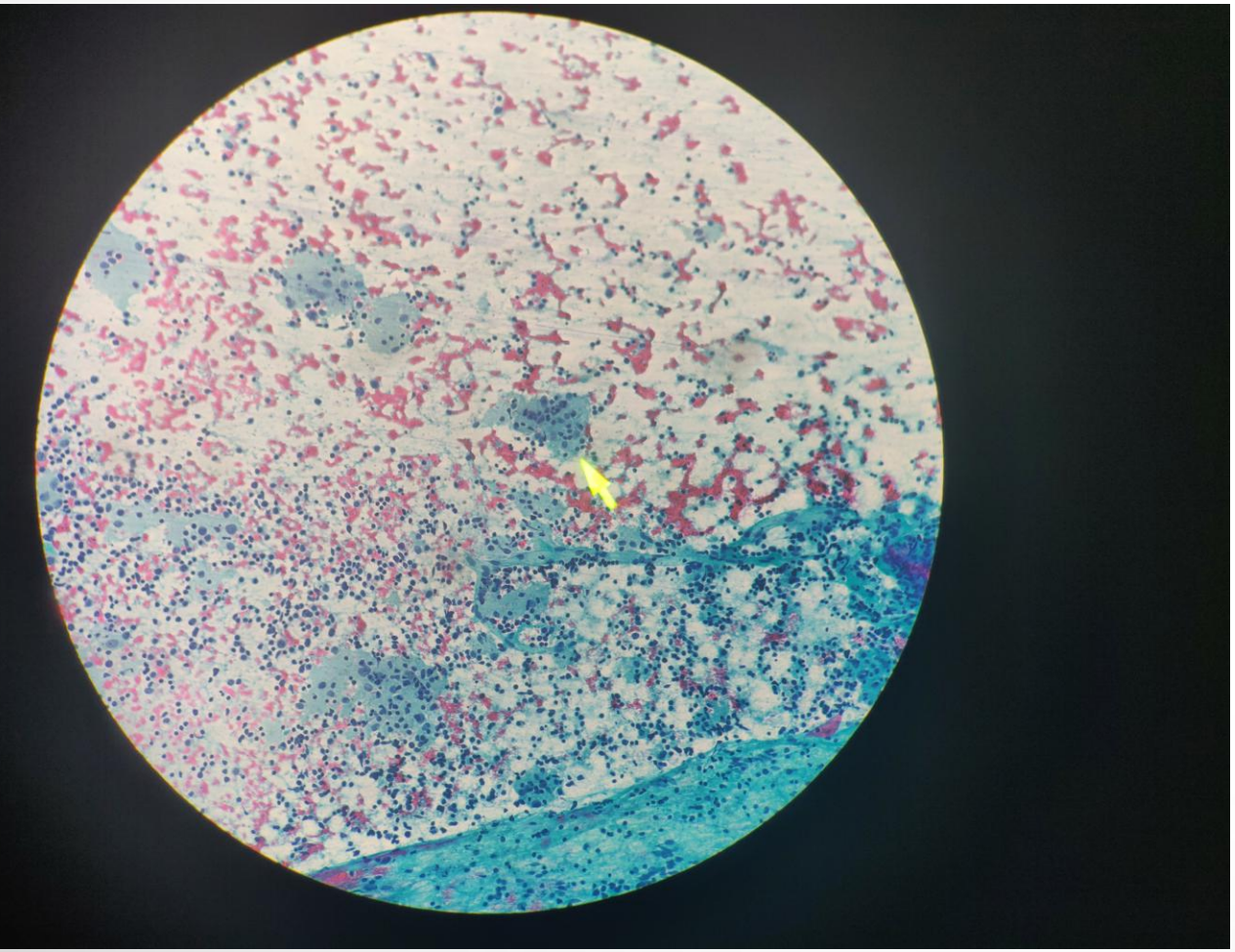
HPLC-D10

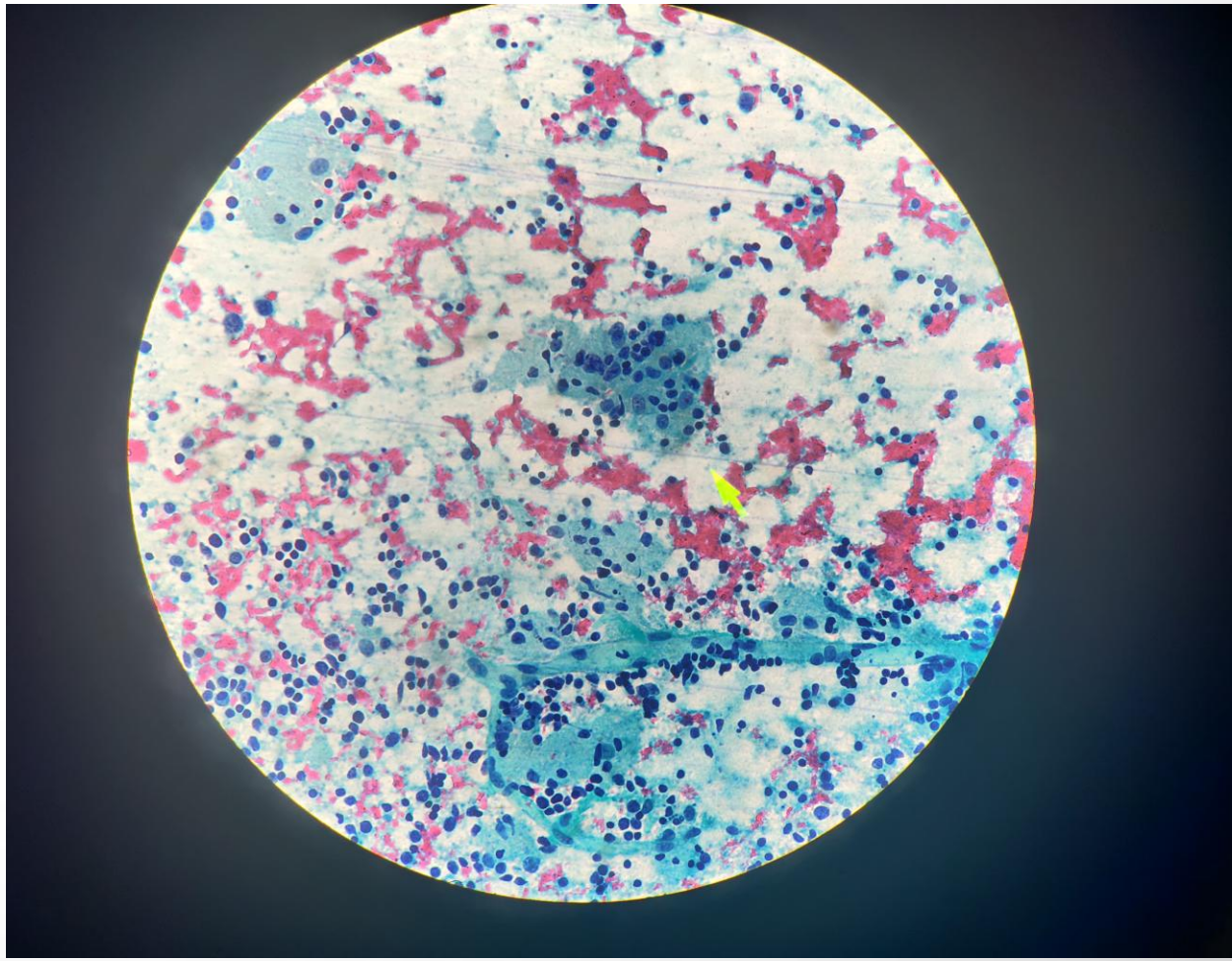
FOETAL HEMOGLOBIN :	Less than 0.8	HPLC	%	Upto 4.0 %
HEMOGLOBIN A	79.0	HPLC	%	
HEMOGLOBIN A2	4.4	HPLC	%	2.0 - 3.5
MPRESSION ::-	Suggestive of Beta thalassemia heterozygous.			
dvice	1) Family studies. 2) Molecular studies for confirmation.			

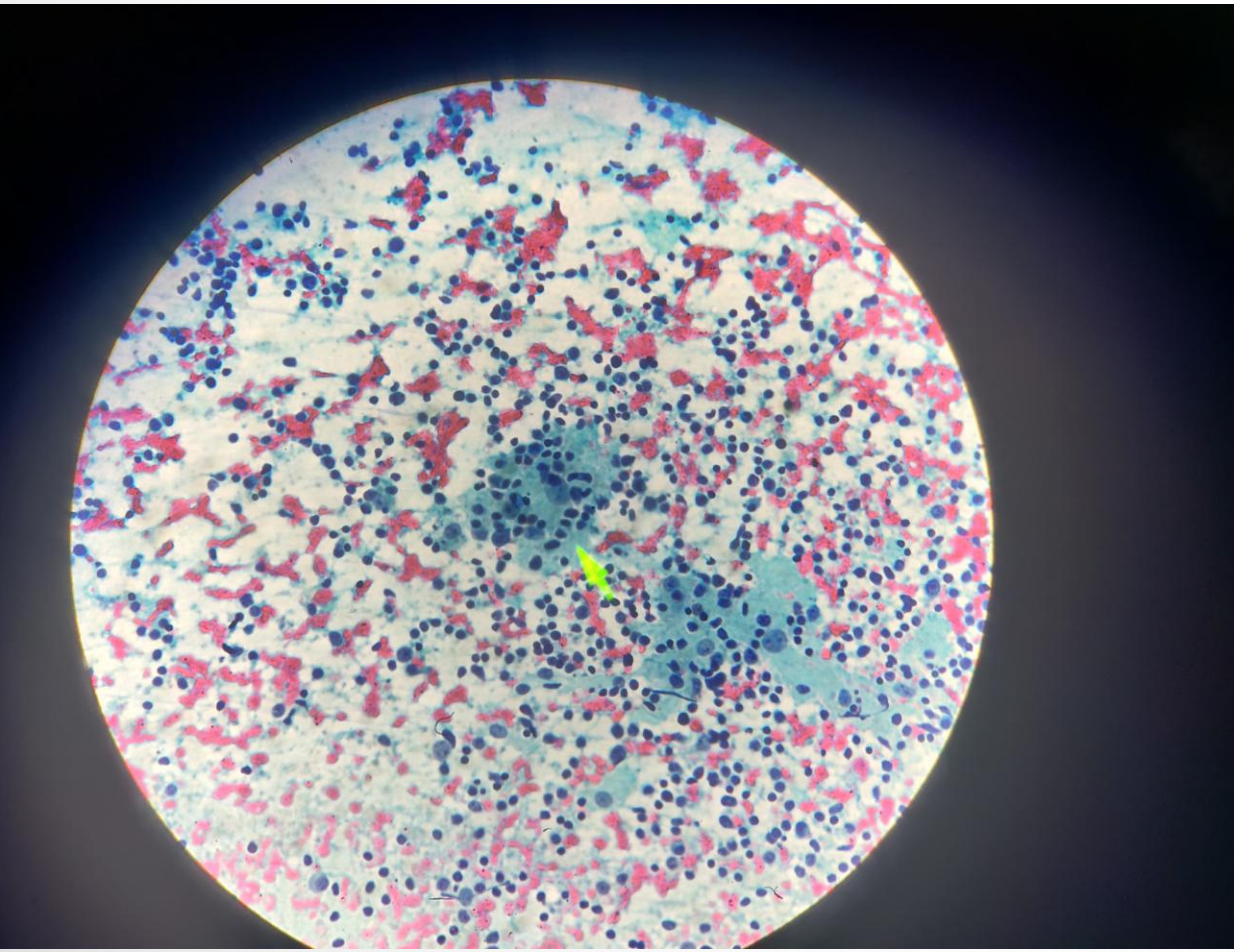

LAB NUCLEAR MEDICINE AND PET CT SERV...
MED NODES
MED NODES
SUBCARINAL NODE
SUBCARINAL NODE

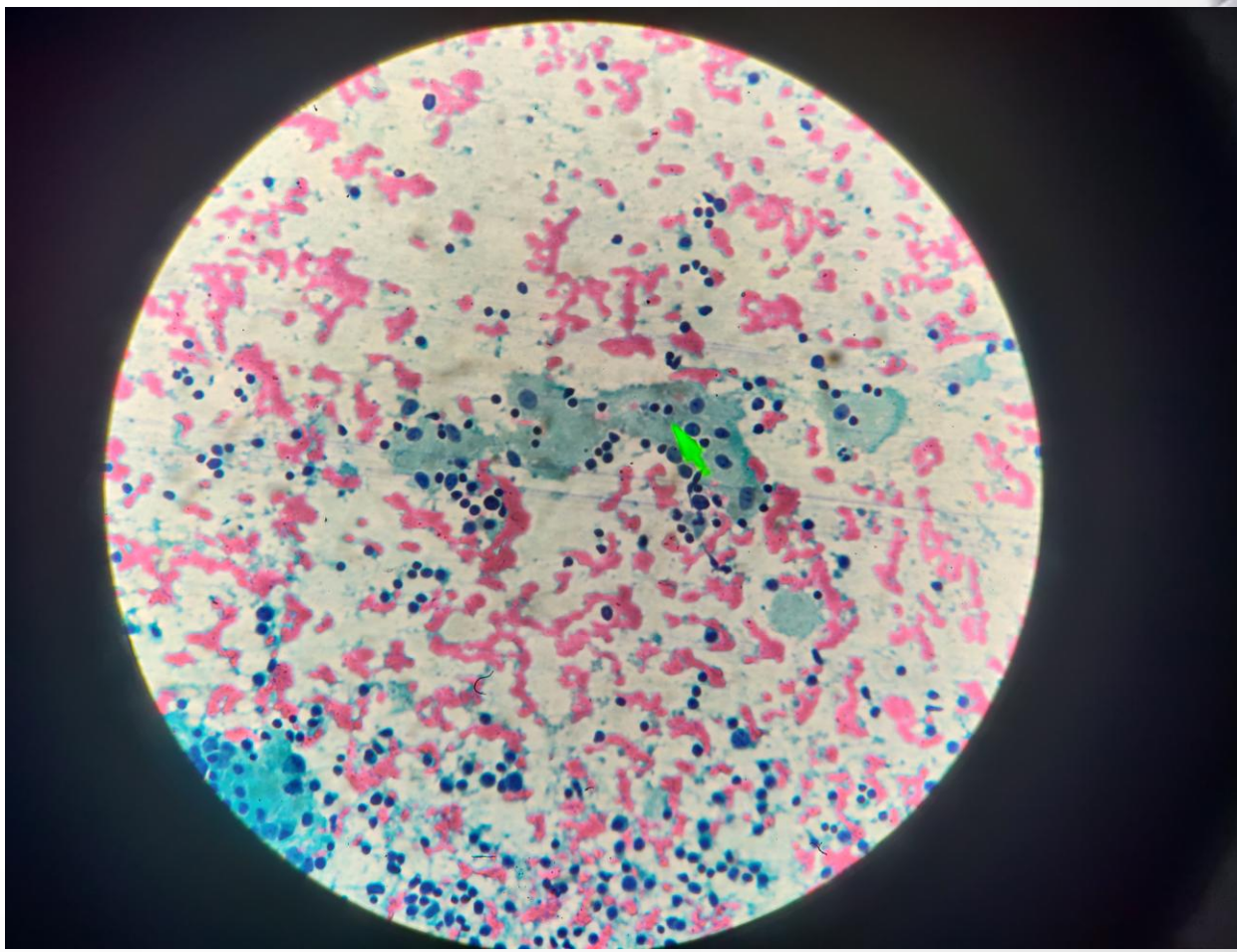
- We planned for EBUS-TBNA.
- His PT- INR was 29 and 2.3.
- Hence after correction with vitamin K, ebus-tbna from subcarinal lymph node taken under FFP cover.

EMPERIPOLESIS









- Emperipolesis is an uncommon biological process, in which a cell penetrates another living cell. Unlike in phagocytosis where the engulfed cell is killed by lysosomal enzymes of the macrophage, the cell exists as viable cell within another in emperipolesis and can exit at any time without any structural or functional abnormalities for either of them.

TB-GENEXPERT ULTRA

* SPECIMEN	EBUS NODAL ASPIRATE
* M TUBERCULOSIS DNA	NOT DETECTED

Principle

The GeneXpert Dx system integrates and automates sample processing, nucleic acid amplification, and detection of the target sequences in simple or complex samples using real-time PCR and reverse transcriptase PCR.
A portion of the rpoB gene containing the 81 base pair "core" region is amplified. The probes are able to differentiate between the conserved wild-type sequence and mutations in the core region that are associated with Rifampicin resistance

Limitations

* **REPORT**

CLINICAL DETAILS:

C/o fever. HRCT- S/o subcarinal lymphadenopathy. To rule out Koch's/ malignancy.

SPECIMEN:

EBUS-TBNA from subcarinal lymph node

GROSS:

Received 5 dry and 5 fixed slides

MICROSCOPY:

Smears are cellular and show mixed population of small and large lymphocytes with few plasma cells and neutrophils. Numerous histiocytes with round vesicular nuclei are seen some of which show evidence of emperipolesis. No granuloma/ necrosis or malignant cells are seen.

IMPRESSION:

EBUS-TBNA from subcarinal lymph node :-

Features are those of Reactive lymphadenitis with histiocytosis

Above features suggest possibility of Rosai Dorfman disease (Sinus histiocytosis with massive lymphadenopathy)

Note: Suggest clinical follow up.

*** End Of Report ***

ROSAI - DORFMAN DISEASE

- Patient was given Prednisolone 40 mg , Thalidomide 100 mg .
- There is significant symptom improvement, with no fever after steroids.

- Rosai-Dorfman disease is a rare disorder characterized by overproduction (proliferation) and accumulation of a specific type of white blood cell (histiocyte) in the lymph nodes of the body (lymphadenopathy), most often those of the neck (cervical lymphadenopathy).
- In some cases, abnormal accumulation of histiocytes may occur in other areas of the body besides the lymph nodes (extranodal).
- These areas include the skin, central nervous system, kidney, and digestive tract.

- The symptoms and physical findings associated with Rosai-Dorfman disease vary depending upon the specific areas of the body that are affected.
- Symptoms of Rosai-Dorfman disease develop due to the overproduction and accumulation of histiocytes in the channels (sinuses) that allow for the passage of lymph (sinus histiocytosis).
- The disorder predominantly affects children, adolescents or young adults. The exact cause of Rosai-Dorfman disease is unknown.

- Isolated mediastinal lymphnodes also should be evaluated and whenever possible , get tissue biopsy done as there can be rare possibility like Rosai Dorfman disease, Castleman's disease.

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Intrathoracic manifestations of Rosai–Dorfman disease

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KEYWORDS
Rosai–Dorfman disease;
Interstitial lung disease;
Histiocyte;
Lymphadenopathy;
Macrophage;
Chest CT

Summary
Introduction: Rosai–Dorfman Disease (RDD), also known as Sinus Histiocytosis with Massive Lymphadenopathy (SHML), is a rare monocyte/macrophage proliferative disorder of varied biological behavior. Although cutaneous and lymph node involvement are relatively well-described, intrathoracic manifestations of RDD have only occasionally been reported.
Methods: We conducted a retrospective computer-assisted search of the Mayo Clinic record from 1976 to 2005 for patients with histopathologic evidence of RDD on organ biopsy. Clinical characteristics were abstracted from charts and thoracic manifestations recorded. Survival was estimated using the national social security database.
Results: A total of 21 patients were diagnosed with RDD over a period of 30 years; 9 had intrathoracic manifestations (43%). Main pulmonary symptoms included dyspnea and cough. Age at the time of diagnosis, gender, race, smoking history, mortality and time of survival after diagnosis were no different between RDD patients with and without intrathoracic manifestations. The most common radiographic thoracic manifestation was mediastinal lymphadenopathy (6 patients). Cystic change, interstitial lung disease, and airway disease were radiographically evident in 4 patients. Seven patients were treated at some point in the course of their disease, most commonly with oral corticosteroids. At the time of last follow-up 87% were alive, with a median (IQR) time interval since diagnosis of 8 years (4–9.7).
Conclusions: Intrathoracic manifestations of RDD are relatively common and include mediastinal lymphadenopathy, airway disease, pleural effusion, cystic and interstitial lung disease. Although limited in size, this series suggests the prognosis of patients with RDD and intrathoracic manifestations is relatively good.

The most common presenting symptom in the vast majority of patients is painless cervical lymphadenopathy.[1—3,12] Cutaneous manifestations are reported to occur in about 10% of patients, and generally consist of asymptomatic xanthoma-like, yellowish or reddish-brown papules, nodules and plaques which may ulcerate.[3,10] Respiratory tract involvement has been described in less than 3% of the cases in a large review of 423 registry patients and carries a worse prognosis.[3] Pulmonary parenchymal involvement has also been reported in a case report.[13] In the current series, we describe intrathoracic manifestations in 9 out of 21 RDD patients; in 5 patients the intrathoracic involvement was determined based on the presence of mediastinal lymph-adenopathy which could not be explained by alternative etiologies, and was associated with confirmatory biopsies of other sites, predominantly enlarged cervical nodes and skin.

Rosai-Dorfman Disease: Rare Presentation as Isolated Mediastinal and Hilar Lymphadenopathy

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Disclosures can be found in Additional Information at the end of the article

THANK YOU