CLINICAL MEETING

DEPARTMENT OF RESPIRATORY MEDICINE

DEPARTMENT OF RADIODIAGNOSIS

SOLITARY FIBROUS TUMOUR OF PLEURA WITH DOEGE-POTTER SYNDROME

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Chief complaints

63 year male, Housekeeper, Tobacco chewer
Never-smoker and No-comorbidities

Presented with complaints of :

Gradual onset , Progressive dyspnea since 1 month duration (mMRC grade II)

Dry cough since 1 month (Increased on Left recumbent posture)

Fatigue+ , weight loss+

No h/o of fever, wheeze, chest pain, palpitations, hemoptysis or syncope

Past surgical history : suprapubic cystostomy done 2 years back

Clinical Examination

Patient was thin built and underweight

General physical examination :

- Recurrent episodic Diaphoresis +
- CLUBBING +

VITALS:

Temp: Afebrile
PR: 110/min. All peripheral pulses were felt equally.
BP: 110/70mmHg
RR: 24 breaths/min
Spo2: 93% on Room air

Clinical Examination

RESPIRATORY SYSTEM:

- TRIALS sign + to Right
- Reduced chest wall movements of entire Left Hemithorax
- Dull note percussion on all areas on Left hemithorax
- Decreased Breath sounds on Left hemithorax
- Decreased Vocal Resonance on Left hemithorax

- No Shifting dullness

Other System examinations were WNL

DIFFERENTIAL DIAGNOSIS:

1. Massive Left pleural effusion
2. Left lung mass lesion

Investigations

Routine Laboratory investigations were WNL

Thyroid Function test was normal

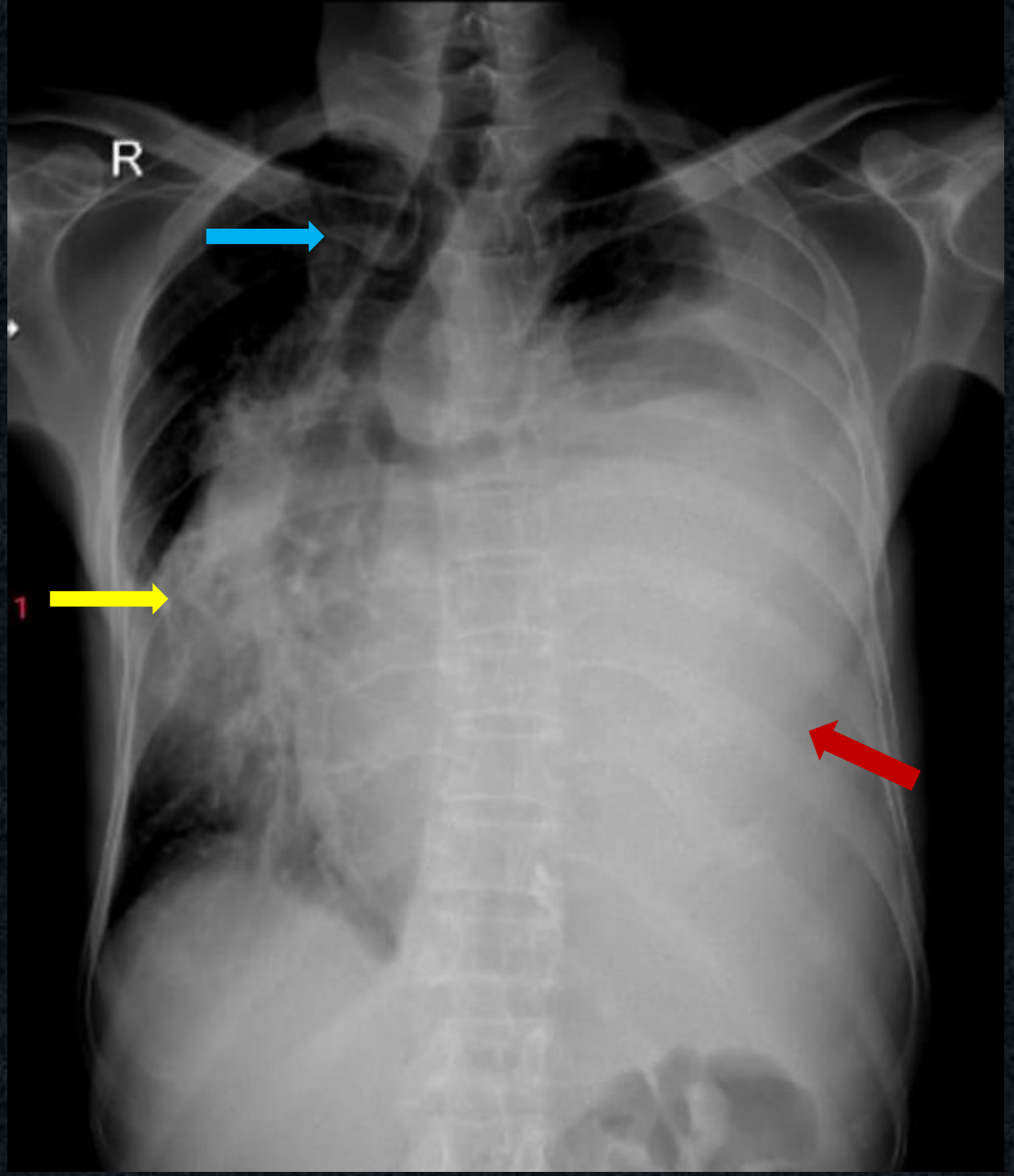
Fasting BSL <60mg/dl
(Recurrent hypoglycemic episodes in morning)

Reason for Episodic Diaphoresis

Sputum examination was inconclusive

ABG showed mild hypoxia (Po2 : 70.2mm HG)

Chest Radiograph
(on admission)

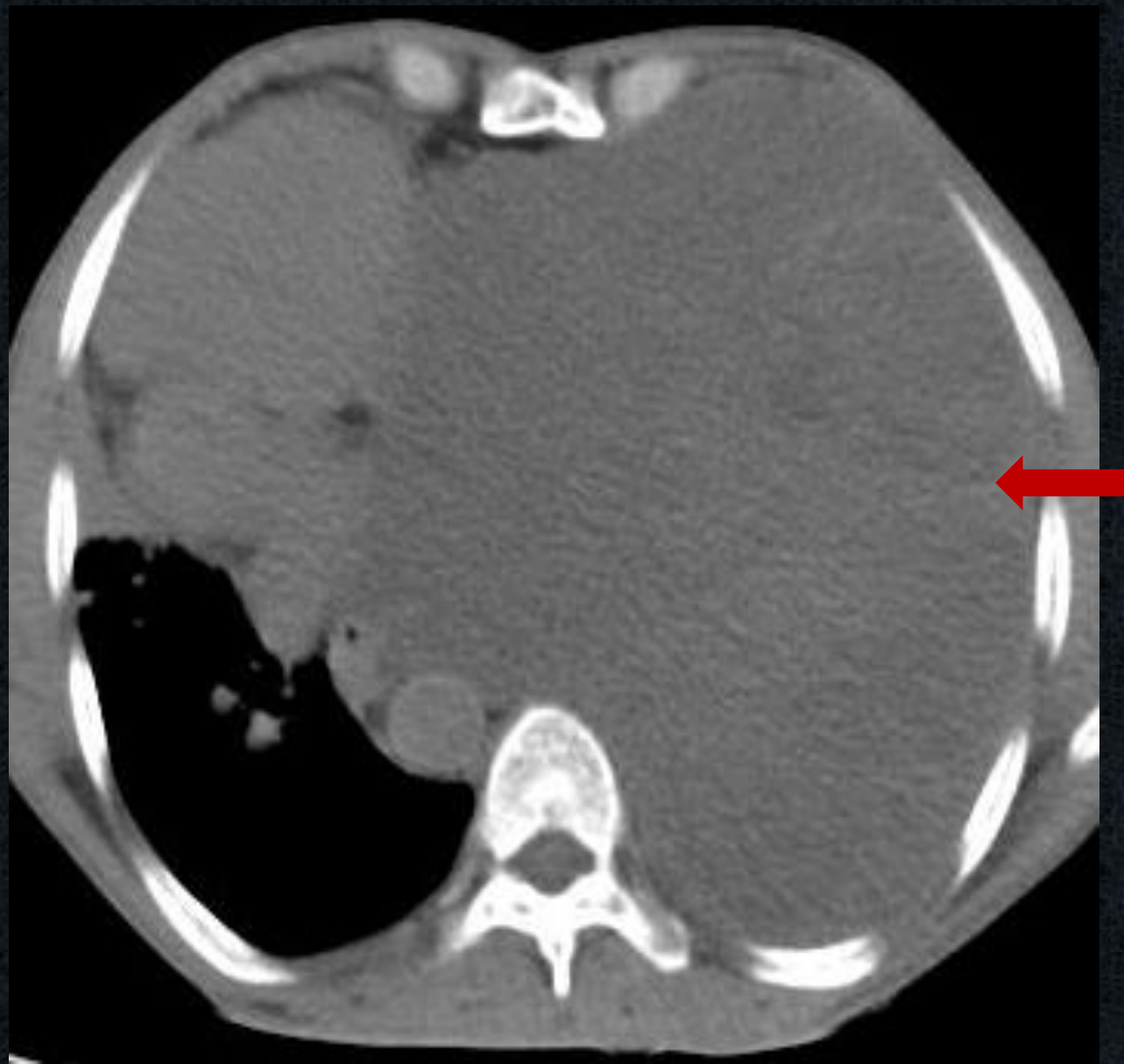


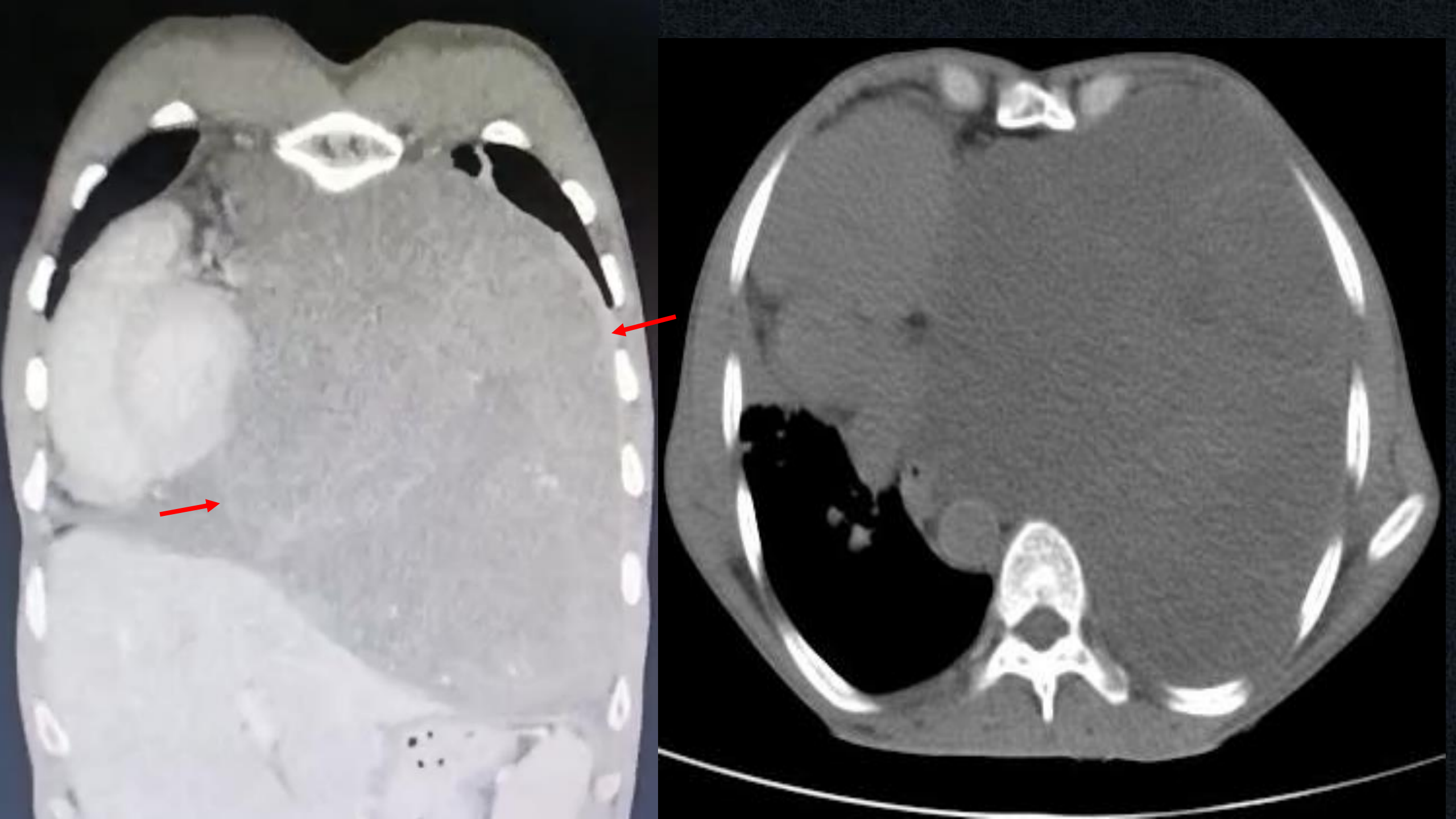
USG THORAX:
Suggestive of Left sided *soft tissue mass* with Left mild pleural effusion

Investigations

CECT THORAX

Suggestive of large Left sided ***pleural based*** soft tissue lesion measuring 150mm x 171mm x 206mm. Lesion showed heterogenous enhancing areas depicting necrosis within tumor mass.
Complete collapse of left middle & lower lobe noted.





DIFFERENTIAL DIAGNOSIS

SOLITARY FIBROUS TUMOR
(Benign localized mesothelioma)

MALIGNANT MESOTHELIOMA

METASTATIC PLEURAL DISEASE

PLEURAL LYMPHOMA

FIBROSARCOMA

DPU
Dr. D.Y. PATIL VIDYAPEETH, PUNE
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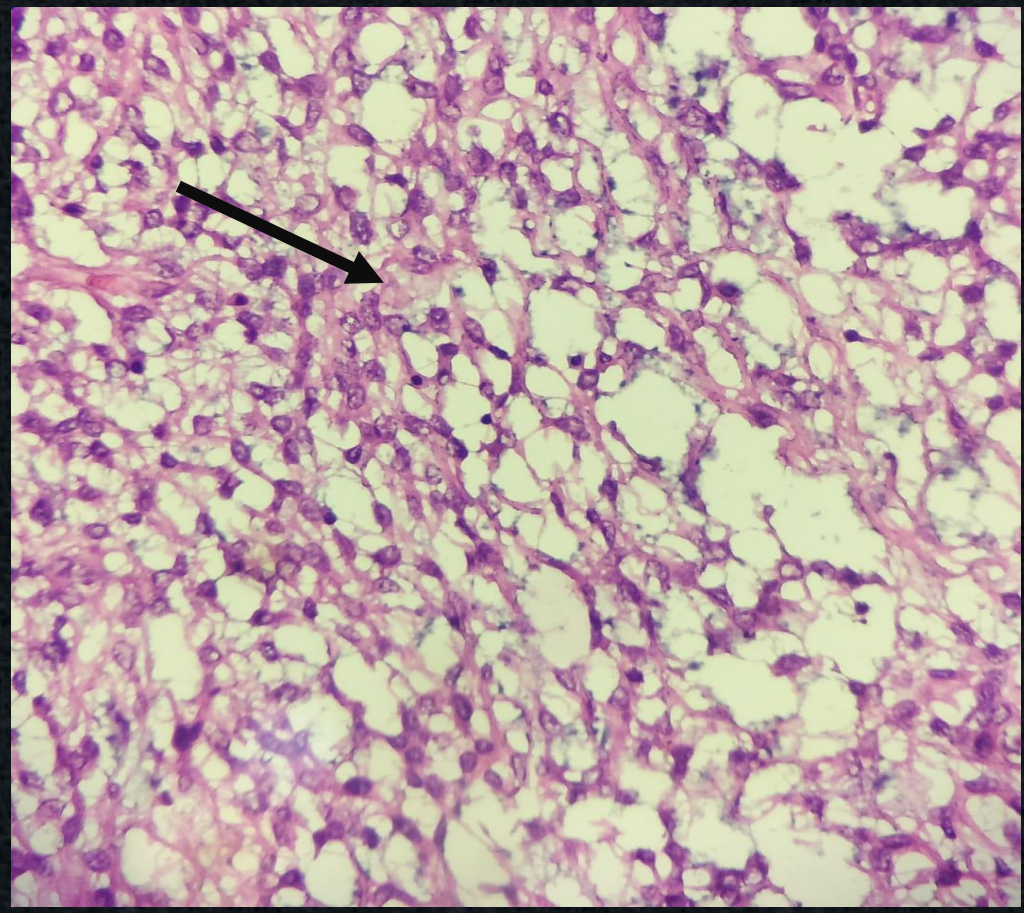
Image guided Trucut Biopsy



Irregularly arranged *Spindle cells* in background of collagenous stroma with low mitotic activity and absence of necrosis.

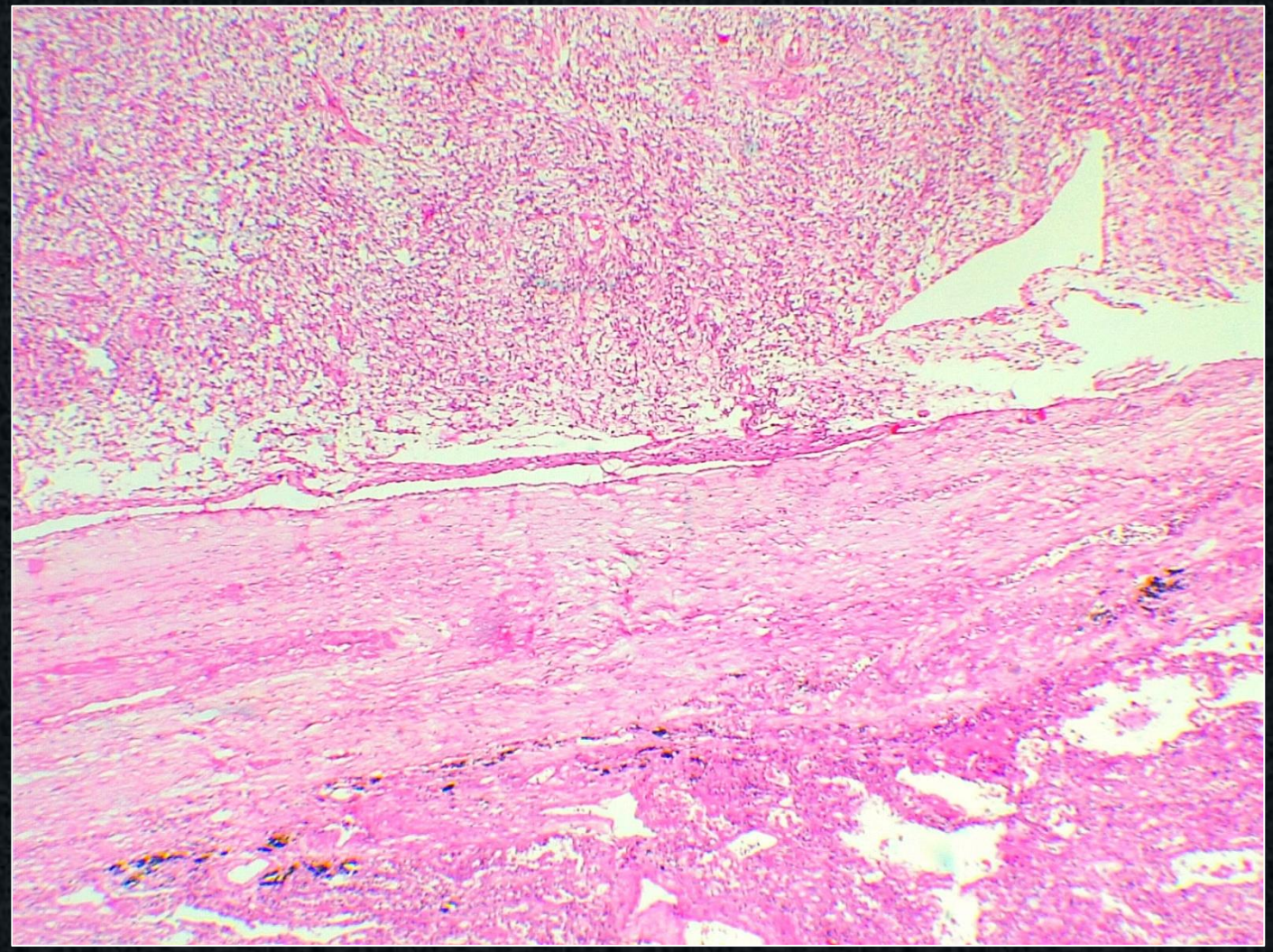
Immunohistochemistry

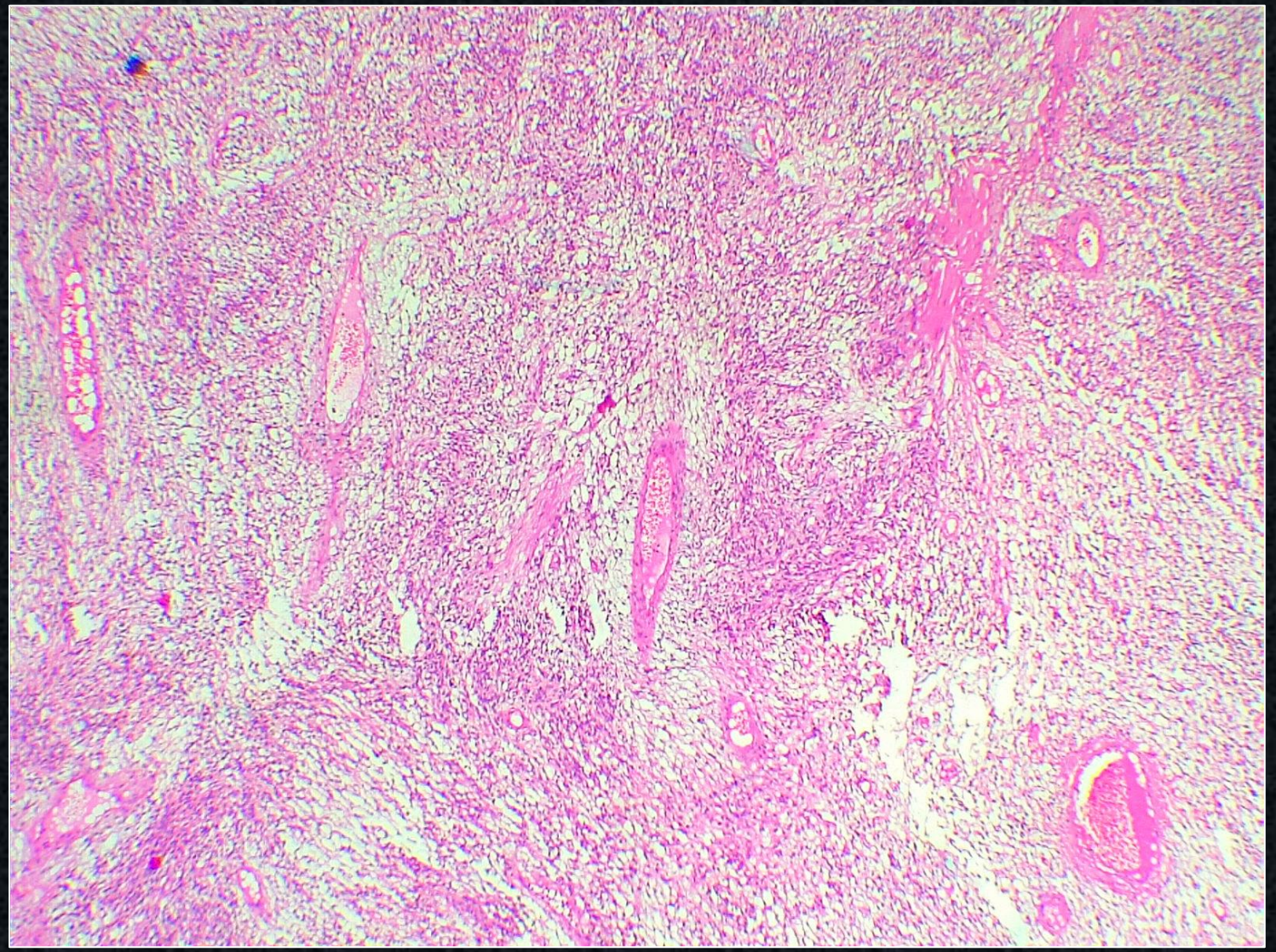
Positive : STAT6, CD34, Bcl2

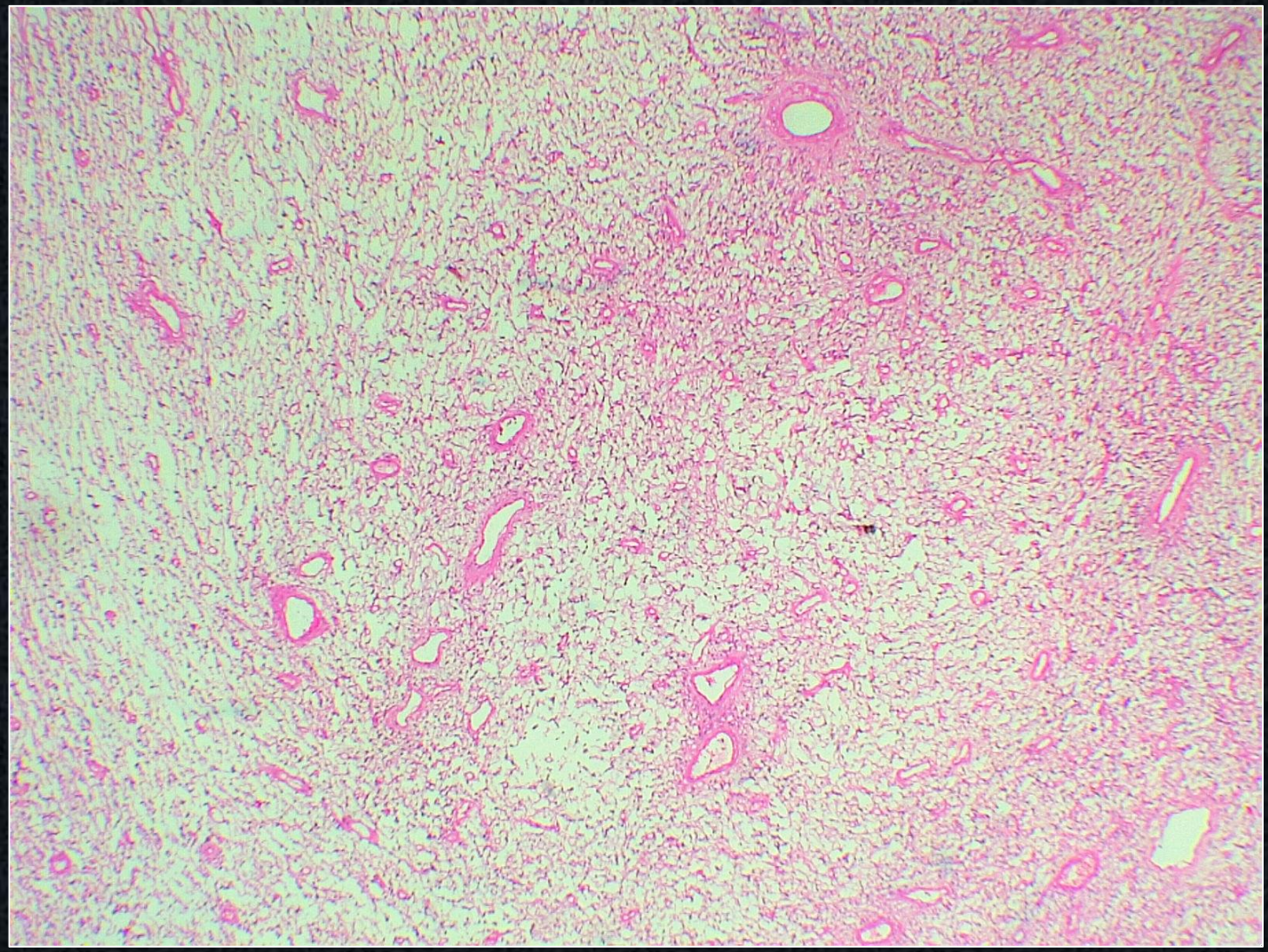
Negative : CK, TLE1, S100

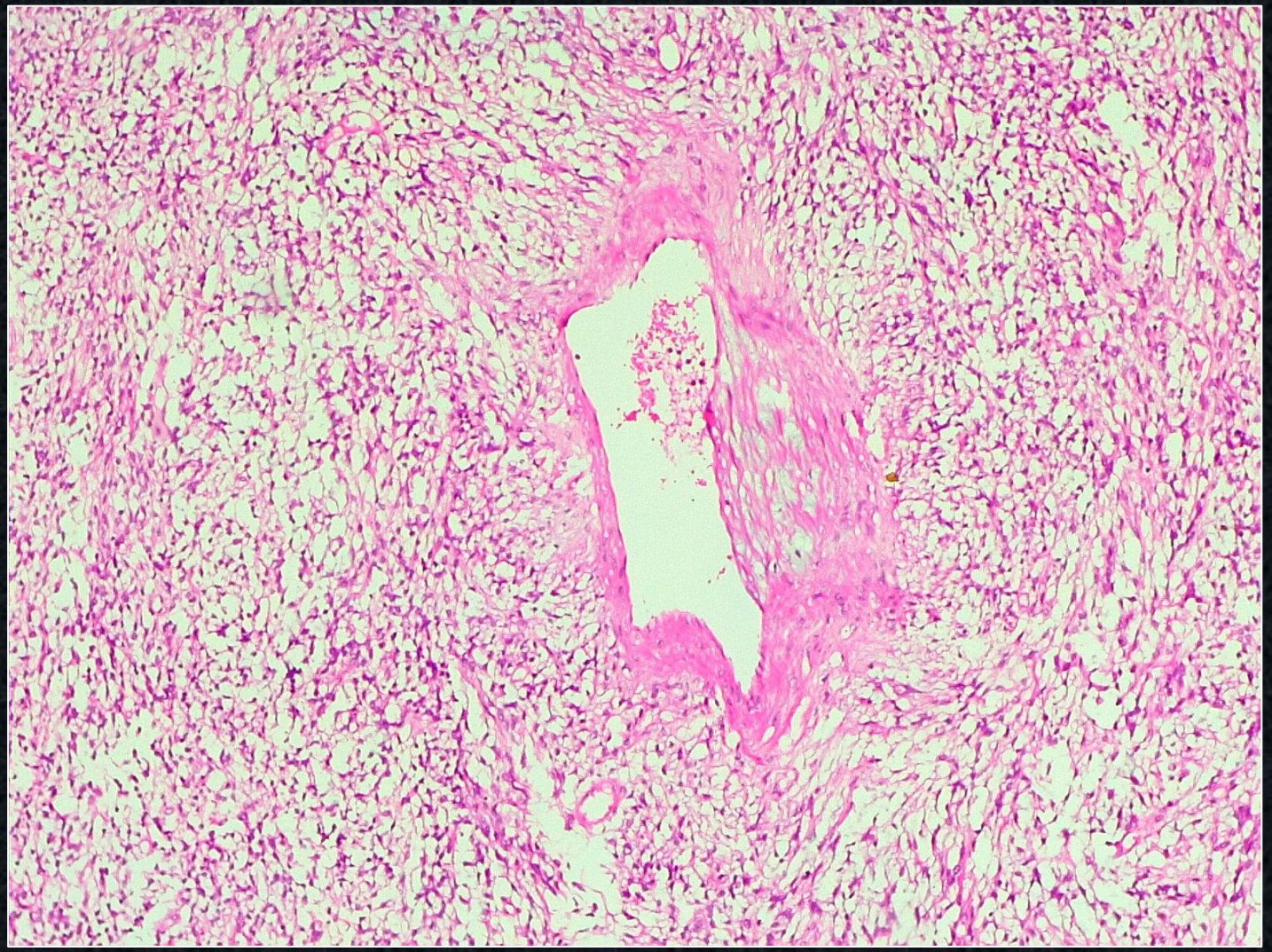
Diagnosis of :

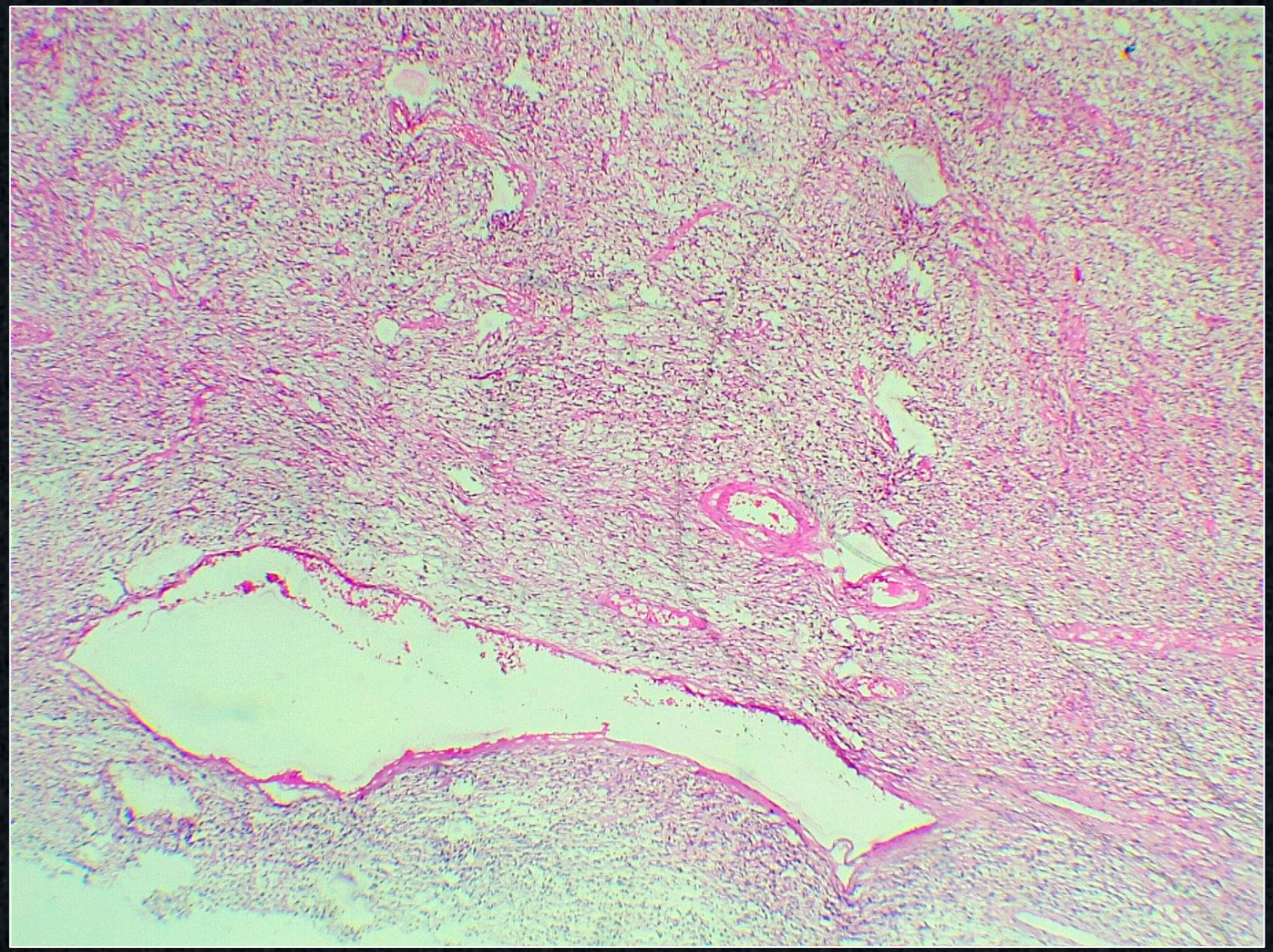
SOLITARY FIBROUS TUMOUR of pleura

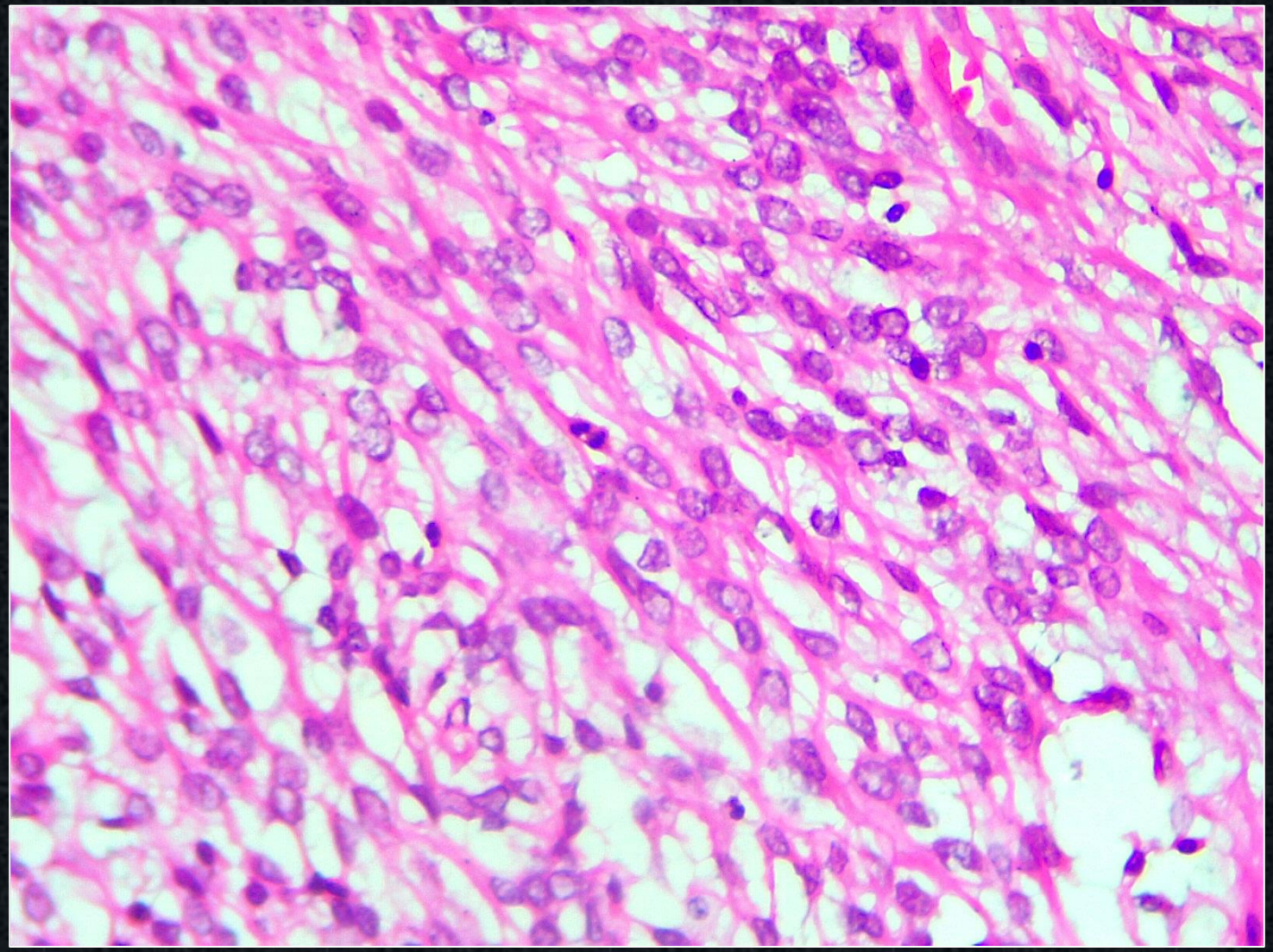


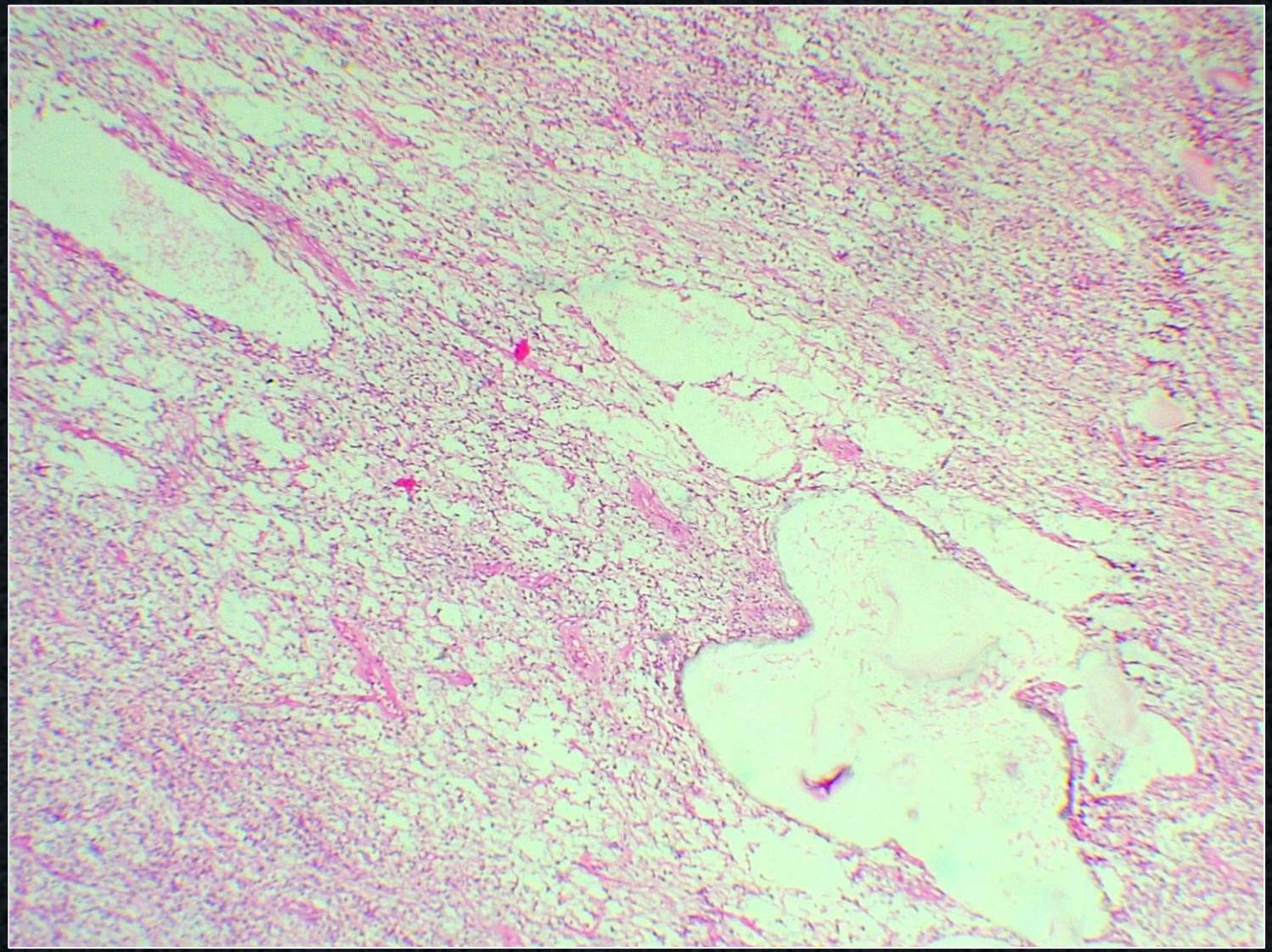


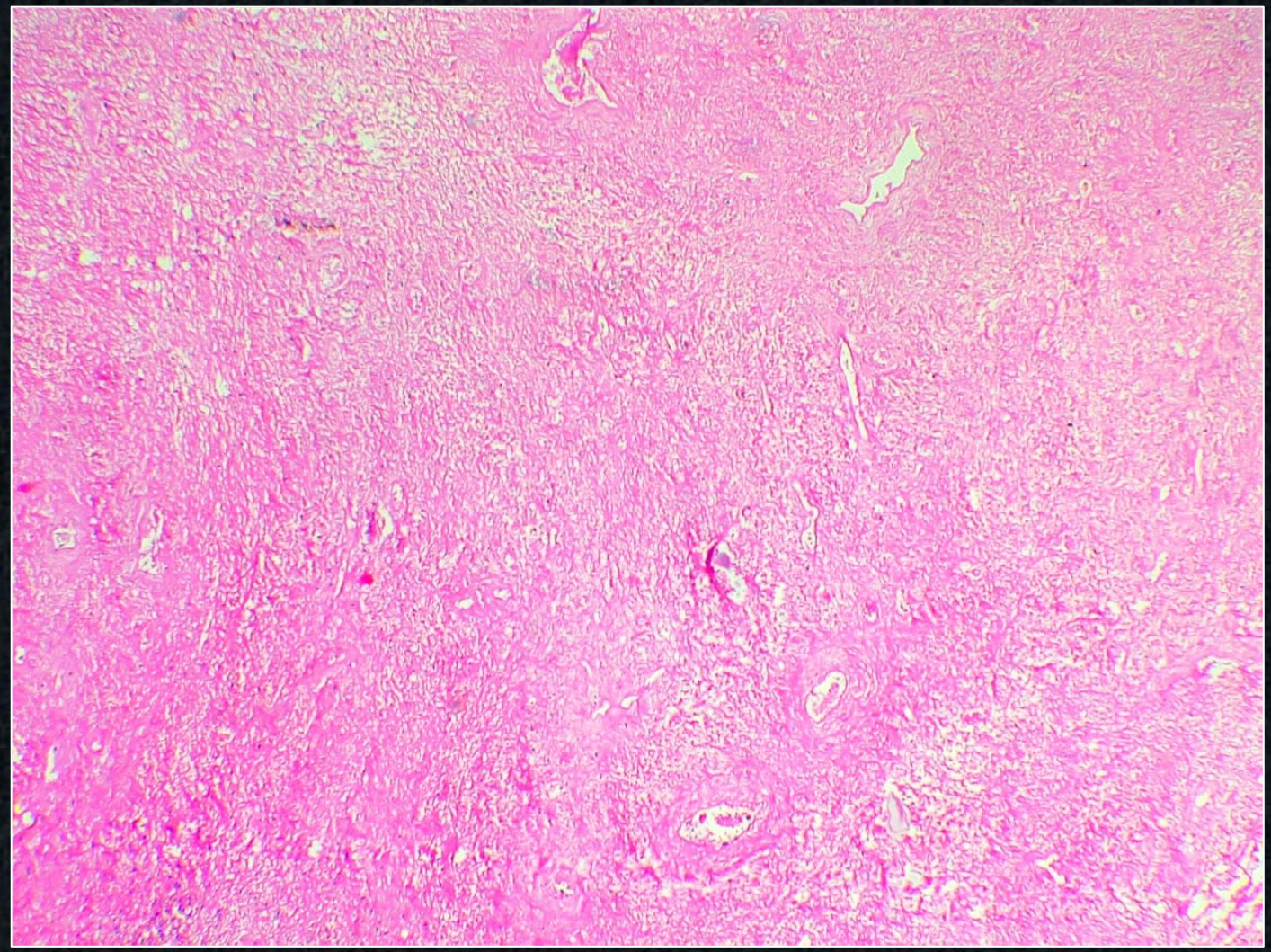












Management

MEDICAL Management :

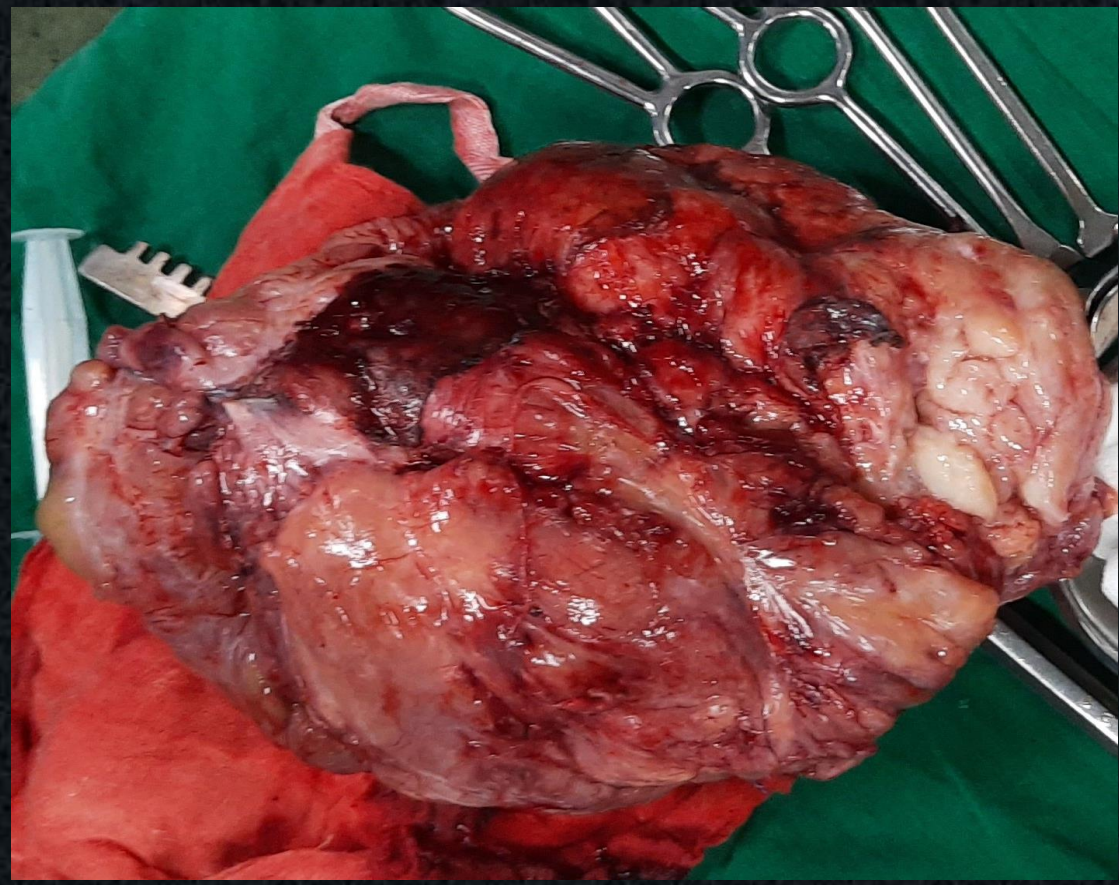
- Low Flow O_2 therapy was given
- Empirical I.V antibiotics for 5 days
- I.V Dextrose for recurrent hypoglycemic episodes

In view of SFTP, opinion of Surgical Oncology was sought and Excision of tumor was advised.

Management

SURGICAL Management :

- A left postero-lateral thoracotomy was performed
- Tumor occupied most of left hemithorax and was adherent to left upper lobe, left hemi-diaphragm and parietal pleura.
- Adhesions were separated and air leak repaired. Partial Left upper lobectomy done.
  Tumor was resected in toto.
- Left middle & lower lobe expanded completely



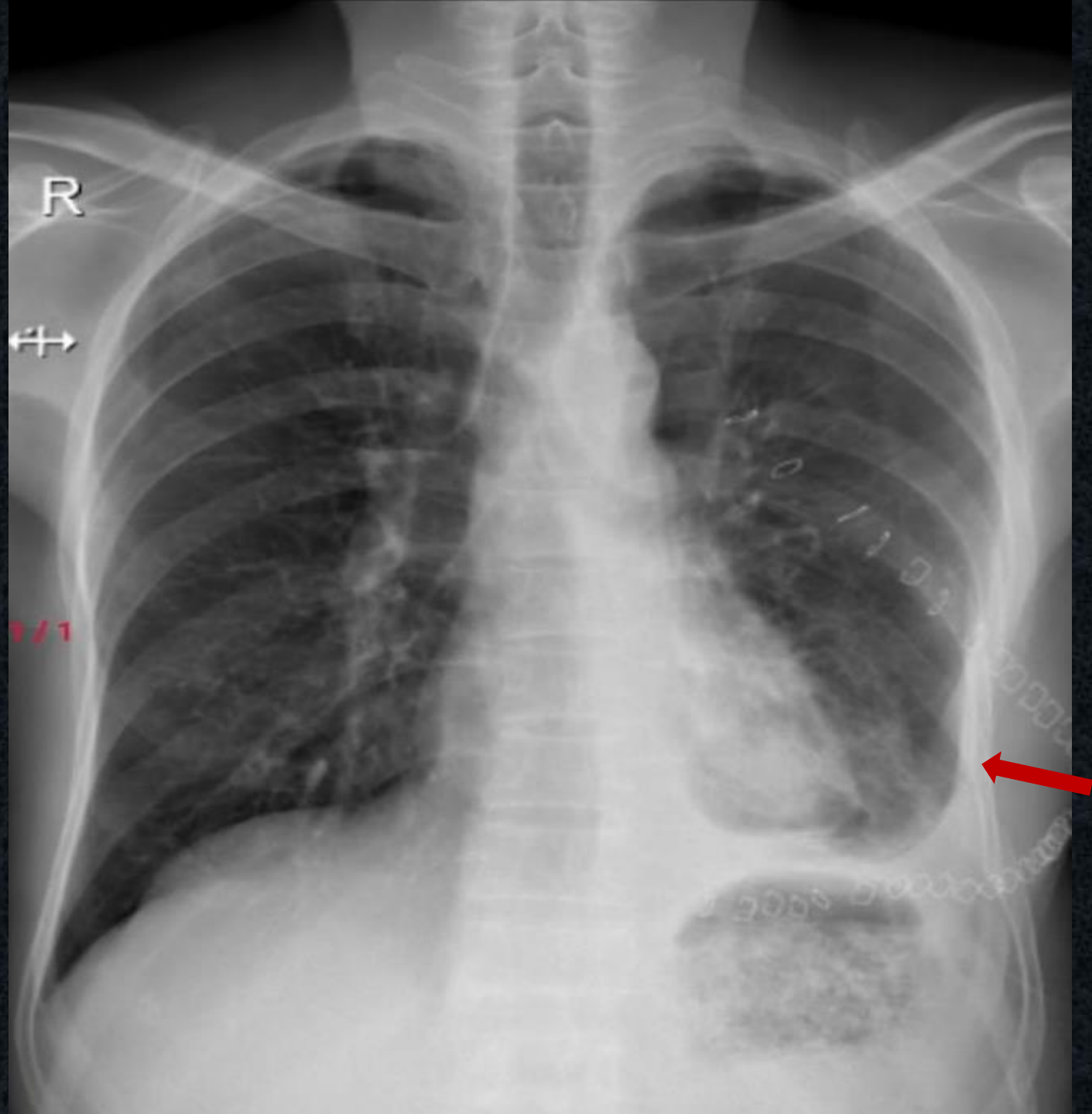
Tumor size : 27 x 25 x 11 cm

weight 3.5Kg

Patient had a smooth post operative recovery.

Post-op Chest Radiograph

Patient was discharged after 10 days and is on regular follow-up.
No signs of recurrence



Discussion

Solitary Fibrous Tumor of Pleura are rare neoplasms derived from mesenchymal cells of pleura

SFTP account for 2% of all soft tissue tumors and 5% of pleural tumors

Asymptomatic in 50% of patients. Can be Malignant (7-60%)

No relation between asbestos exposure and SFTP

Less than few hundred cases have been reported in literature since 1930s

Cough, dyspnea and chest pain are most common symptoms

Hypertrophic Osteoarthropathy seen in 20% of cases

'Doege-Potter syndrome' is a paraneoplastic syndrome associated with the tumor characterized by recurrent hypoglycemia (4%)

Rare & unique presentation of SFTP

First described in 1930 by Doege and Potter

Only 5 such cases have been reported in India

Diagnosis is established by biopsy of tumor.

Histology: uniform elongated *spindle cells* with varied amounts of collagen

IHC: Tumor cells express STAT6, CD34, Vimentin, Bcl2 and Negative for CK

Treatment of choice is surgical removal of tumor by Thoracotomy or VATS

Clinical Pearls

SFTP carries good prognosis
5 year survival rate were 97% for benign and 89% for malignant

Doege-Potter syndrome should be suspected in any non-diabetic patients presenting with recurrent hypoglycemic attacks

Complete resection is associated with best prognosis.
Recurrence risk <10%

Yearly radiological follow up is adviced

Acknowledgements

Department of Onco-Surgery

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Department of CVTS Anesthesia

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Dr. D.Y. PATIL VIDYAPEETH, PUNE
(DEEMED UNIVERSITY)

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