

CASE PRESENTATION

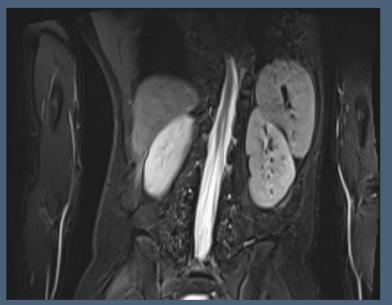
MENTOR: DR R V MALI KAHER UNIVERSITY J.N.MEDICAL COLLEGE ,BELAGAVI PRESENTOR: DR HARSHITA

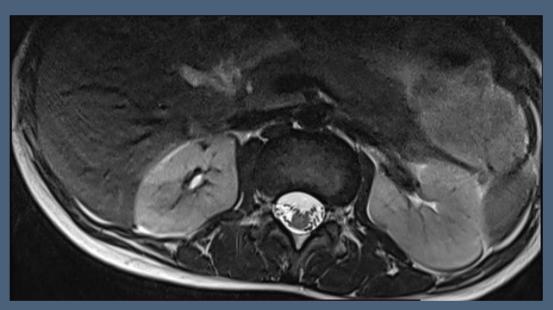
HISTORY

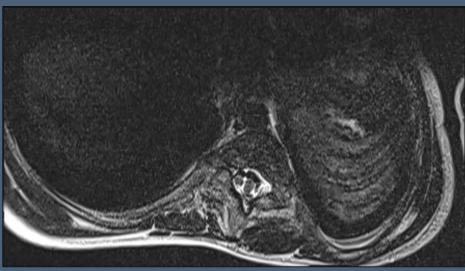
- 14 year old female presented with complains of backache since 1 year for which she was advised MRI spine
- No history of trauma

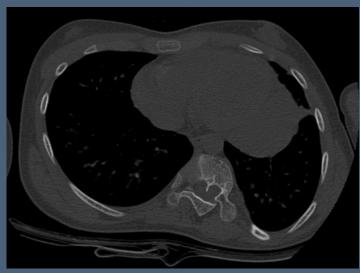
INVESTIGATIONS

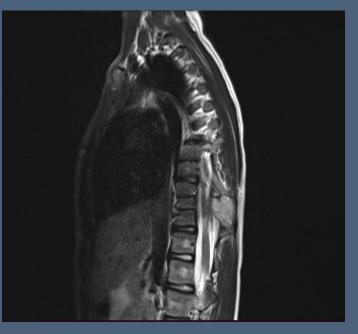
- Low Haemoglobin LDH borderline raised
- AFP within normal limits



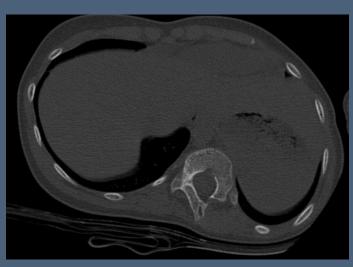


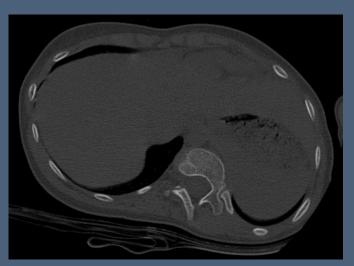












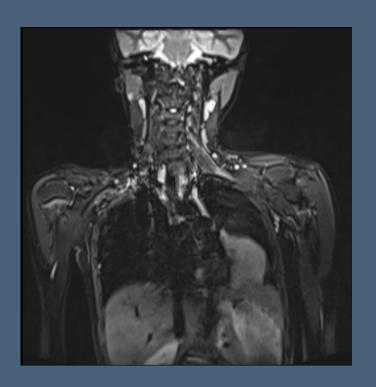
MRI FINDINGS:

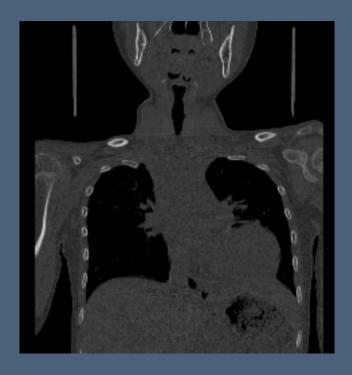
- Splitting of the spinal cord with duplication of dural sac noted from the level of D8 to D11 vertebral body level, for an approximately length of 4.0 cm suggestive of diastematomyelia
- There is partial fusion of the D9, D10 and D11 vertebral bodies and their posterior elements with rudimentary intervertebral disc resulting in focal scoliotic deformity at that level with convexity towards left side.
- The cord is seen to end at the lower border of L2 vertebral body with filum terminale running close to the posterior aspect of the dural sac ...? Tethered.

On CT correlation,

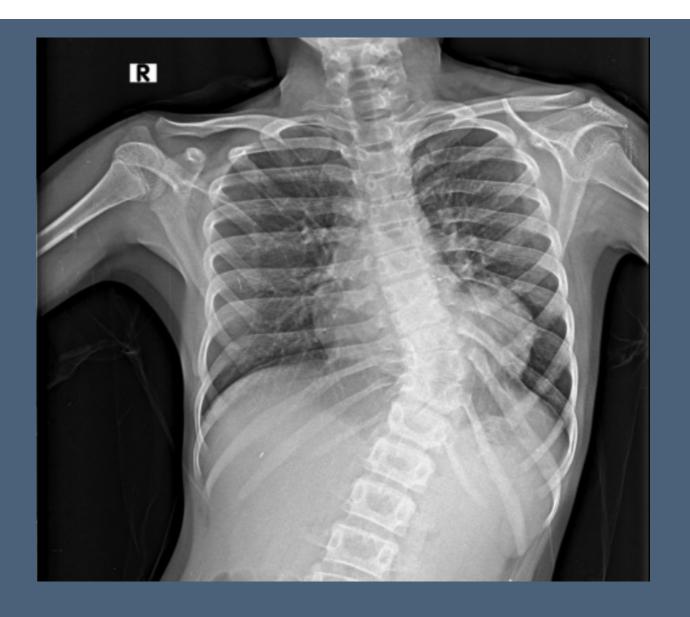
- There is seen a midline osseous septum at the level D9 vertebral body arising from the posterior aspect of the vertebral body dividing the canal dividing into two parts.
- There is seen non fusion of the posterior element of D3, D11 and S3 vertebral bodies suggestive of spina bifida.

INCIENTALLY,





Mass lesion noted with broad base towards the mediastinum was noted on left side for which further evaluation was suggested.



FINDINGS ON CHEST XRAY Levoscoliosis with fused lower thoracic vertebrae There is seen a homogenous opacity in the left lower zone with broad base towards the mediastinum forming an obtuse angle & silhouetting the left border- likely to be anterior mediastinal mass.

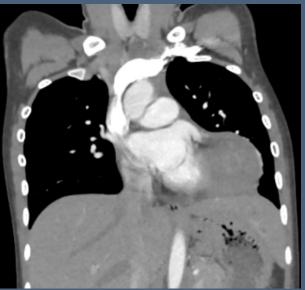






THIN PLAIN







THIN ARTERIAL

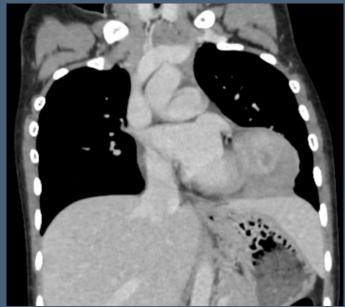






THIN VENOUS







THIN DELAYED

FINDINGS ON CECT THORAX

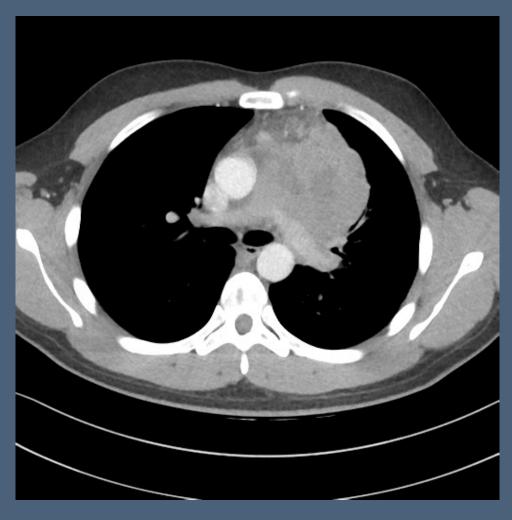
- Well-defined isodense heterogeneously enhancing soft tissue density mass lesion seen in the vascular compartment along the basal segment of left lower lobe (at the level of D6-D10 vertebral bodies) with broad base towards the mediastinum
- Medially, the lesion shows loss of fat planes with adjacent pericardium & focal loss of fat planes with left ventricular myocardium.
- Inferiorly , the lesion is seen to closely abut the left dome of diaphragm with few areas of indistinct fat planes

DIFFRENTIAL DIAGNOSIS

- LYMPHOMA
- INVASIVE THYMOMA
- INFLAMMATORY MYOFIBROBALSTIC TUMOUR

LYMPHOMA

	POINTS IN FAVOUR	POINTS IN AGAINST
•	Most common mediastinal tumours in adolescents	Isolated to the mediastinum- absence of other lymph nodes
•	Asymptomatic mediastinal mass discovered incidentally	
•	Predominantly solid and invasive- Pericardial involvement (infiltration)	Absence of cystic low-density areas



LARGE HETEROGENEOUS MEDIASTINAL MASS WITH INVOLVEMENT OF MEDIASTINAL STRUCTURES: IN PARTICULAR, THERE IS ENCASEMENT AND MASS EFFECT ON THE MAIN AND LEFT PULMONARY ARTERY.

INVASIVE THYMOMA

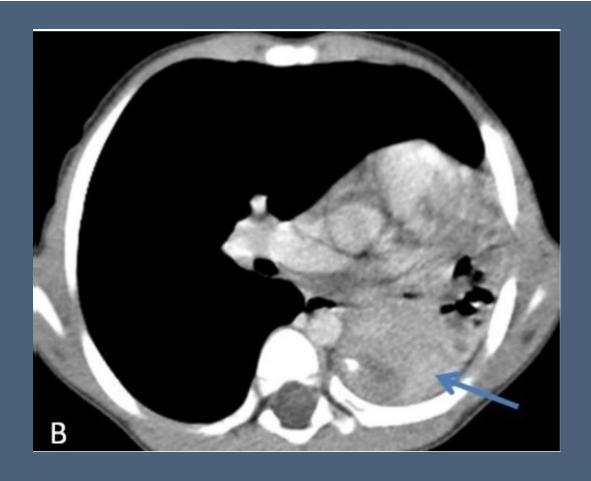
POINTS IN FAVOUR	POINTS IN AGAINST
Solid and heterogeneously enhancing nature with invasion-Loss of fat planes with the pericardium and left ventricular myocardium	Age group
	Often associated with myasthenia gravis
	Visceral compartment



THERE IS A LARGE LOBULATED SOFT TISSUE DENSITY MASS IN THE ANTERIOR MEDIASTINUM WITH SMALL FOCI OF CALCIFICATION.

INFLAMMATORY MYOFIBROBALSTIC TUMOUR

POINTS IN FAVOUR	POINTS IN AGAINST
Age group	
Soft tissue density mass with heterogenous enhancement	Broad base towards mediastinum in the vascular compartment
Absence of lymphadenopathy	Lack of calcification



THERE IS A SOFT TISSUE DENSITY HETEROGENOUSLY ENHANCING MASS IN THE LEFT LOWER LOBE CONTAINING A SMALL CALCIFICATION.

FOLLOW UP:

• Complete excision of the lesion was done on 22.04.25

HISTOPATHOLOGICAL REPORT

Surgical Pathology Report with Immunohistochemistry SD 1061/2025

Name: Chandana Sadashiv Hurakadi

Age: 14 years

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Sex: Female

Referring surgeon: Dr Santosh Kurbet

Hospital: KLE Prabhakar Kore

Hospital, Belgaum

Received: 30/04/2025

IHC Reported on: 16/05/2025

Origin of Tissue: Mediastinal tumour

Clinical Diagnosis: Middle mediastinal tumour adherent to lung and mediastinum close to

pericardium and no hilar lymph nodes. ? Benign ? Malignant

Immunohistochemistry:

Block : SD 1061A/25

Markers: LCA(PD7/26+2B11,MM), Pan CK(AE1/AE3,MM), S 100(15E2E2,RM), SMA(1A4, MM),

ALK 1(5A4,MM) from Biocare

Findings: Immunohistochemically, the spindle-shaped and large cells are positive for AE 1/AE 3 (Pan CK, cytoplasmic), focally positive for SMA (cytoplasmic). The dense round cell infiltrate is diffuse and is positive for LCA. The spindle shaped cells and large squamoid cells are distinctly positive for anaplastic lymphoma kinase (ALK 1) and negative for S-100.

Impression:

Excised mediastinal tumour (middle) whose histology with immunohistochemistry show features of an inflammatory myofibroblastic tumour-ALK positive, of borderline malignancy and is limited by the capsule.

INFLAMMATORY MYOFIBROBALSTIC TUMOUR

EPIDEMIOLOGY:

- Rare in children and young adults (mean age ~3-4 years)
- Rare with their incidence reported at approximately 0.04-1% of all the <u>pulmonary neoplasms</u>
- Can affect any age group, around 25% of cases occur in those under 18 years of age.
- It is the most common primary mass of the lung in children.

> TYPICAL LOCATION:

- Often arises in lung parenchyma (especially peripheral lower lobes) or mediastinum
- Pulmonary IMTs are usually solitary and peripherally placed, most commonly in the lower lobes
- Extrapulmonary IMTs can occur in mediastinum, pleura, chest wall, etc.

> RADIOGRAPHIC FEATURES:

- CT: Typically seen as a single , well defined (can be multiple in \sim 5% of cases) peripheral, lobulated mass with lower lobe predominant occurrence.
 - Calcification can occur (commoner in tumors occurring in children).
 - Usually shows heterogeneous enhancement with contrast.
 - In children, coarse calcifications are common (seen in ≈29% of cases) and may appear as dense flecks on CT (bright areas).
 - Cystic change or cavitation can occur but is less frequent.
- MRI shows IMTs as iso- to hypointense on T1 and hyperintense on T2, with mild contrast enhancement.

> INVASION PATTERN:

- IMTs are generally slow-growing but can be locally aggressive.
- They may infiltrate adjacent lung, pleura or chest wall.
- Very rarely, they invade pericardium or myocardium.
- Unlike malignancies, true lymph node spread or distant metastases are uncommon.
- Local extension across anatomical planes is possible.

> PATHOLOGY

- It is generally considered to fall within the benign spectrum of tumors.
- Microscopically they are characterized by abundant inflammatory infiltrate comprising of predominantly plasma cells, lymphocytes, histiocytes, admixed with a variable proportion of fibroblasts and myofibroblasts.
- On immunohistochemistry, the tumor cells can exhibit
 - strong diffuse positivity with smooth muscle actin and vimentin
 - negativity for cytokeratin, CD34 and \$100

> PROGNOSIS AND TREATMENT:

- Complete surgical excision is the mainstay and is usually curative; 5-year survival is excellent with R0 resection.
- Recurrence occurs in a minority (≈20%) and is more likely if margins are positive or tumor is invasive.
- Because many IMTs are ALK-positive, targeted therapy with ALK inhibitors (e.g. crizotinib) is effective in unresectable or metastatic cases.
- Chemotherapy or steroids have been used in unresectable IMTs.

CLINICAL AND RADIOLOGICAL PRESENTATION IS VARIABLE AND NONSPECIFIC AND THE DIAGNOSIS IS RARELY MADE BEFORE SURGICAL MANAGEMENT.

THANK YOU