

# An Uncommon Presentation of a Rare Disease- Anterior Mediastinal Leiomyosarcoma Presenting as Anterior Mediastinal Mass

Dr. Vikas Marwah<sup>MD</sup><sup>1</sup>, Dr. Robin Choudhary<sup>MD</sup><sup>2\*</sup>, Dr. Arpita Pemmaraju<sup>MD</sup><sup>3</sup>, Dr. Virender Malik<sup>MD, DNB</sup><sup>4</sup>

<sup>1</sup>Pulmonary Medicine Prof and HOD, Dept of Pulmonary, Critical Care and Sleep Medicine, AICTS, Pune, India

<sup>2</sup>Pulmonary Medicine, Senior Resident, Dept of Pulmonary, Critical Care and sleep Medicine, AICTS, Pune, India

<sup>3</sup>Pathology, Dept of Pathology, AICTS, Pune, India

<sup>4</sup>Radiology, DM (Interventional Radiology and Neuroradiology), Assoc Prof, Dept of Radiology, AICTS, Pune India

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\*Corresponding author: Dr. Robin Choudhary

Pulmonary Medicine, Senior Resident, Dept of Pulmonary, Critical Care and sleep Medicine, AICTS, Pune, India

## Abstract

Soft tissue sarcomas are a rare group of malignancies which arise from smooth muscle cells. Leiomyosarcomas arising from anterior mediastinum is rarely reported in the literature. The current COVID-19 pandemic has caused unprecedented havoc worldwide. It is also causing delay in diagnosis of other diseases. Herein we report a case of an elderly male who was managed as a case of COVID pneumonia and on follow-up review in post COVID clinic was diagnosed to have anterior mediastinal leiomyosarcoma and was managed with chemotherapy.

**Keywords:** SARS-CoV2, Leiomyosarcoma, Soft tissue sarcoma.

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## INTRODUCTION

Soft tissue sarcoma (STS) are rare group of malignancies which arise mainly from embryonic mesoderm and accounts for 1% of all malignancies. Leiomyosarcoma (LMS) is one of the most common type of STS. It generally arises from smooth muscle cells or its precursor cells and it shows a predilection for soft tissues and abdomino-pelvic organs. Mediastinal leiomyosarcoma is a very rare tumour and typically arises from posterior mediastinum. Mediastinal leiomyosarcoma arising from anterior mediastinum is more rarely reported. COVID-19 has created havoc worldwide and now the aftermath is more dreadful. It has masked the clinical features of many underlying diseases due to which patients are reporting late. We report a case of an elderly male who reported to our post- COVID clinic and on evaluation was found to have a mediastinal leiomyosarcoma.

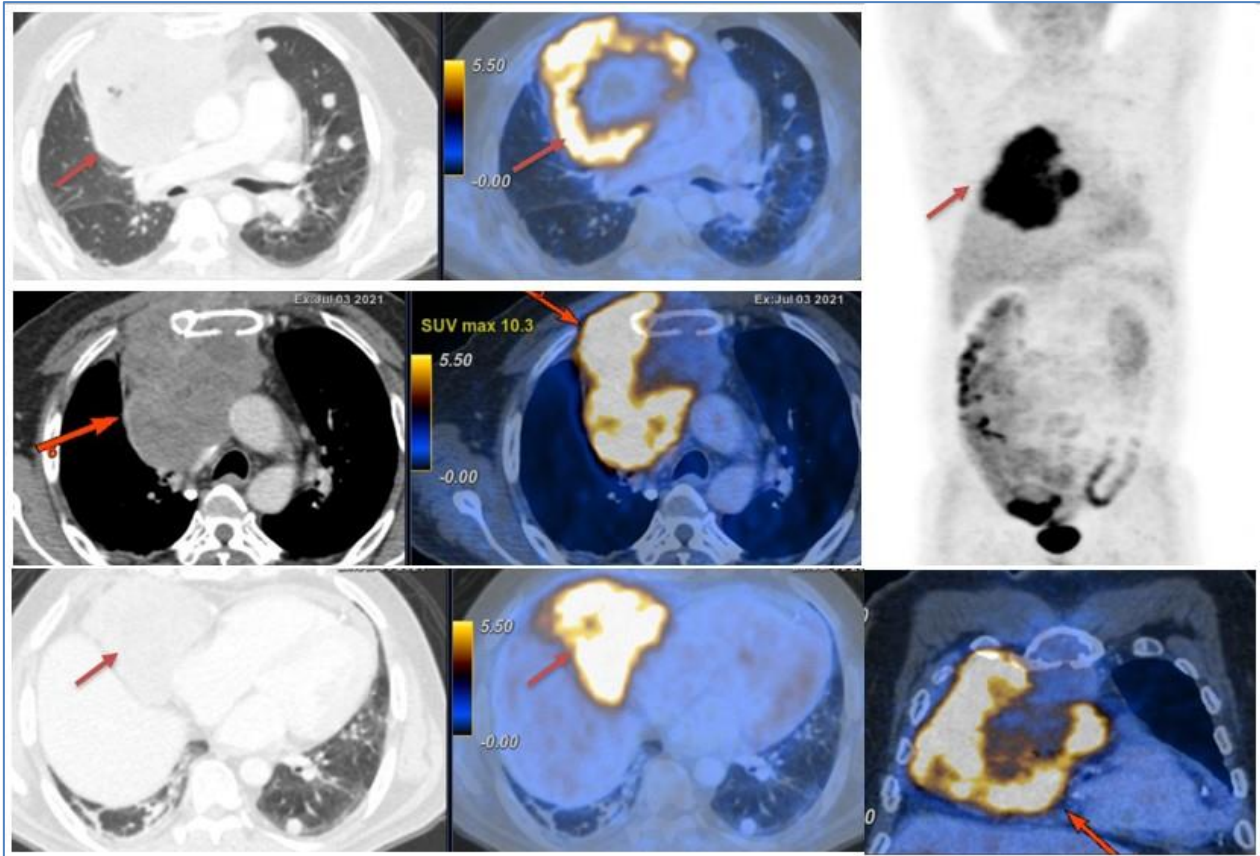
## CASE REPORT

62-year-old male, reformed smoker (smoking Index-140), a known case of Primary hypertension, Type-II Diabetes Mellitus presented with history of fever, dry cough and generalised malaise of four days duration. On evaluation his throat swab for RT-PCR was positive for SARS-CoV2. His haematological,

renal and liver parameters were within normal limits however his inflammatory markers in form of C-Reactive protein (CRP), serum ferritin and D-dimer were raised. He was managed with intravenous steroids (inj dexamethasone 6 mg once daily), low molecular weight heparin and oxygen therapy via nasal prongs. He showed good response to therapy and was later discharged to home. He reported to our post- COVID clinic 4 weeks later for review. He was symptomatic with dry cough and breathlessness, retrospectively he gave history of weight loss for 6 months and he had significant desaturation on 6 Minute walk tests. On evaluation, his high-resolution computed tomography (HRCT) of chest showed a large well defined soft tissue density mass (10.5 X 12.4X 13 centimetres) in anterior mediastinum predominantly on right side. There were also multiple hypodense areas suggestive of cystic degeneration with few tiny specs of calcification within. The mass was seen abutting anterior chest wall, manubrium and body of sternum; laterally it was seen abutting adjacent part of right upper and middle lobe while medially it was abutting adjacent aorta and right atrium. There was loss of intervening fat planes with right atrium. The mass was also compressing the SVC and was also seen crossing the midline extending into adjacent pericardial fat. There were discrete small nodular lesions in left upper lobe and lingula and

segmental collapse of anterior segment of right upper lobe and medial segment of right middle lobe with associated volume loss. He underwent whole body PET scan which confirmed FDG avid (SUV max 10.3) heterogeneously enhancing soft tissue mass in anterior

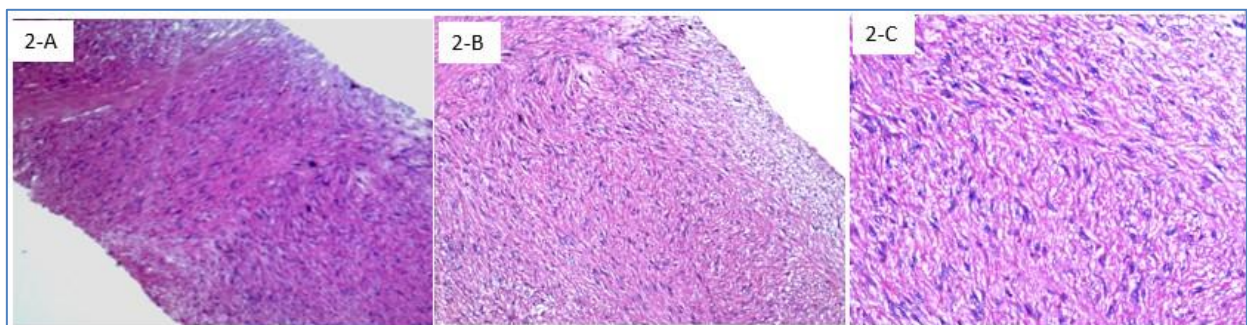
mediastinum, predominantly on the right side with necrotic component within. Anteriorly, chest wall infiltration through right 2<sup>nd</sup> intercostal space to pre-sternal region, erosion of manubrium sternum and body of sternum was also seen.



**Fig-1: Showing representative CT and PET images of the patients showing FDG avid anterior mediastinal mass**

The patient underwent a CT guided biopsy which showed a cellular tumour in herring bone pattern and interlacing fascicles comprising of atypical spindle shaped cells with increased N: C ratio, pleomorphic nuclei, vesicular chromatin and inconspicuous nucleoli.

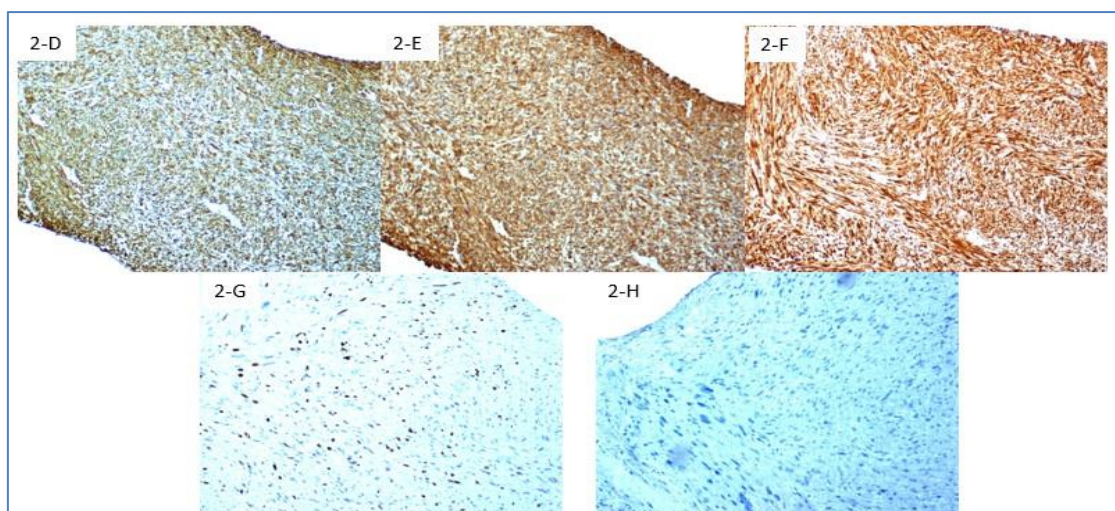
Proliferation of blood cells and mononuclear infiltration was also noted at places. There were also foci of bizarre nuclei and mitosis (1-9 /10 high power field). (Fig-2-A, B, C).



**Fig-2-A, B, C: Showing microscopic picture of the tumor composed of fascicles of spindle shaped cells in x5 (A), x10 (B) and x40 (C) magnification.**

On Immunohistochemistry, the tumour cell was positive for Vimentin, Desmin, SMA, Ki67 (30%)

and negative for Pan-CK, S-100, B-Catenin, BCL-2 and CD34. (Figure-2-D, E, F, G, H).



**Fig-2-D, E, F, G, H: Showing Immunohistochemical features of the tumour - Positive for SMA (D), Vimentin (E), Desmin (F) and Ki-67 (G) and negative for S100 (H) in x10 magnification**

The patient was diagnosed as a case of Leiomyosarcoma of grade-2 (FNCLCC grade). The patient was reviewed by oncophysician and he was started on chemotherapy as per MAID protocol. The MAID regimen consists of injectable Mesna 1500 mg/m<sup>2</sup>/day for 4 days, Doxorubicin 15 mg/m<sup>2</sup>/day for 3 days, Ifosfamide 1500 mg/m<sup>2</sup>/day for 3 days and Dacarbazine 250 mg/m<sup>2</sup>/day for 3 days which is administered every 4 weeks. The patient was later discharged and is being kept on follow-up.

## DISCUSSION

STS are group of heterogenous tumours which accounts for less than 1% of adult tumours. Leiomyosarcomas accounts for 25% of sarcomas. They arise mainly form embryonic mesoderm (Mangla *et al.*, Micheal CH) LMS shows a predilection for soft tissues and abdomino- pelvic organs. Mediastinal leiomyosarcoma is a rare entity and there are even fewer cases reported of anterior mediastinum leiomyosarcoma (Ishikawa A *et al.*, Eroglu A *et al.*) The leiomyosarcoma can be classified into multiple groups including retroperitoneum, abdominal cavity, somatic soft tissue and of vascular origin subgroup. The somatic subgroup like in our case generally has been associated with better prognosis but those with vascular origin are associated with worse prognosis.(Mangla *et al.*, Ishikawa A *et al.*, Mestiri S *et al.*).

The origin of LMS is still not understood and there are no definite identifiable risk factors. The most common organ involved is uterus, however LMS originating in the mediastinum is rarely seen. The pathophysiology of LMS is very heterogenous due to inconsistent cytogenetic and molecular changes. Classically LMS arise from smooth muscle cells and on cytology show intersecting groups of spindle cells with elongated, hyperchromatic nuclei and abundant eosinophilic cytoplasm (Mangla *et al.*, Micheal CH, Mestiri S *et al.*).

The presentation of LMS can be with non-specific symptoms, most commonly as mass compressing the nearby organs. Imaging in form of computed tomography or Magnetic resonance imaging can confirm the diagnosis and stage of involvement which is important as the tumour mainly spread through haematogenous route (Micheal CH, Ishikawa A *et al.*, Eroglu A *et al.*)

The diagnosis can be confirmed on histopathology and these tumours generally show staining of both smooth muscle and actin and desmin, especially the well differentiated tumours. The Immunohistochemistry can also be used to differentiate LMS from other spindle cell sarcomas and nerve sheath tumours (Mangla A *et al.*, Ishikawa A *et al.*).

The treatment option for LMS is limited with and surgery can be offered to patients with limited disease, however the disease is known to metastasize early. The patients with metastatic disease can be managed with cytotoxic chemotherapy including doxorubicin and ifosfamide (Mangla A *et al.*, Ishikawa A *et al.*, Mestiri S *et al.*, Burt M *et al.*) Our patient was given three cycle of chemotherapy as per MAID protocol which consists of (1) mesna 1500 mg/m<sup>2</sup>/day x 4 days; (2) doxorubicin 15 mg/m<sup>2</sup>/day x 3 days; (3) ifosfamide 1500 mg/m<sup>2</sup>/day x 3 days; (4) dacarbazine 250 mg/m<sup>2</sup>/day x 3 days which is administered intravenously every 4 weeks. The patient is presently under our follow-up.

## CONCLUSION

Our case report highlights the rarity of LMS originating in the anterior mediastinum. COVID-19 pandemic has caused unprecedented mortality worldwide and has also affected the presentation of other diseases causing delay in reporting and further management. The treating physician should keep the

diagnosis of LMS in mind while working up a case of mass in anterior mediastinum.

### Statements

### Acknowledgement – Nil

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### Statement of Ethics

The patient had given written informed consent to publish their case and the study protocol was approved by the departmental and institute's committee of medical research.

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