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RESEARCH ARTICLE

INFLAMMATORY MYOFIBROBLASTIC TUMOR OF THE MEDIASTINUM IN A PEDIATRIC PATIENT: A RARE CASE PRESENTATION AND MANAGEMENT REVIEW

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Abstract

Mediastinal tumors in pediatric patients encompass a broad spectrum of pathologies, ranging from benign to malignant entities. We present a case of a 10-year-old boy who initially presented with persistent back pain followed by nocturnal coughing, asthenia, and significant weight loss over a year. Upon admission, physical examination was unremarkable except for a mediastinal opacity noted on radiographic evaluation, leading to further characterization by thoracic CT scan, revealing a sizable right paracardiac mediastinal mass. Surgical excision was performed successfully, with en bloc resection of the mass adherent to the middle and lower lung lobes, and involving regional lymph nodes. Postoperative recovery was uneventful over a two-month follow-up period.Histopathological analysis confirmed the diagnosis of an inflammatory myofibroblastic tumor (IMT), a rare entity within the mediastinum Our case underscores the challenges in diagnosing and managing mediastinal IMTs, emphasizing the importance of a multidisciplinary approach involving clinical suspicion, diagnostic imaging, and definitive histopathological evaluation.Continued reporting and study of such cases contribute to our understanding of this rare pathological entity, guiding optimal management strategies and long-term surveillance protocols for affected pediatric patients. This case report highlights the need for heightened awareness and research efforts to improve diagnostic accuracy and therapeutic outcomes for mediastinal IMTs in pediatric populations.

Introduction:-

Mediastinal tumors in children a diverse range of pathologies, including both benign and malignant forms. [1]A comprehensive understanding of the different compartments of the mediastinum is essential for conducting relevant etiological investigations. These tumors may be discovered incidentally or manifest with symptoms that impact prognosis. Imaging plays a crucial role in the diagnostic process. [2]Masses located in the anterior mediastinum are primarily associated with lymphomas and germ cell tumors, while neurogenic tumors predominate in the posterior mediastinum. [9]

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Case Presentation

We report the case of a 10-year-old boy, who presented with back pain persisting for 6 months prior to admission. The progression was marked by the onset of nocturnal coughing one month before admission, all within the context of asthenia and a weight loss of 10 kg over a year. On admission, physical examination revealed a conscious child, stable in terms of respiratory, hemodynamic, and neurological parameters. Examination of the pleuropulmonary and cardiovascular systems was normal,and peripheral lymph node examination was unremarkable.

Laboratory findings revealed microcytic hypochromic anemia on complete blood count (CBC), while electrolytes, infectious workup, and tumor markers (Bhcg and Afp) were within normal limits. Radiographic evaluation **(FIGURE1)** demonstrated a mediastinal opacity in the lower part of the right lung field, partially obscuring the ipsilateral diaphragmatic dome. A thoracic CT **(FIGURE2)** scan further characterized a sizable right paracardiac mediastinal mass, well-defined and lobulated, measuring 72 x 66 mm. The mass displayed regular borders, some fine central calcifications, heterogeneous enhancement following iodinated contrast injection, and was in contact with the right pulmonary artery, nestled between the two superior and inferior pulmonary veins. It displaced the inferior vena cava, came into contact with the right atrium and ventricle without signs of invasion, and reached the ipsilateral diaphragmatic dome without penetrating it.

Abdominal ultrasound for staging revealed no abnormalities. The decision was made to proceed with surgical excision, during which a mediastinal mass measuring 7x7 cm, adherent to the middle and lower lobes and involving two lymph nodes, was identified. The mass was totally excised **(FIGURE3)** with preservation of lung parenchyma, and the postoperative course was uneventful over a follow-up of 2 months. Histopathological studies confirmed the presence of an inflammatory myofibroblastic tumor.

Discussion:-

Inflammatory myofibroblastic tumors (IMTs) are rare neoplasms, particularly uncommon in the mediastinal region. [1]The literature highlights their benign nature, although their occurrence within the mediastinum remains infrequent. [2] Our patient, a 10-year-old male, exemplifies the unique demographics associated with this condition. The pathogenesis of IMTs remains poorly understood; however, autoimmune and infectious mechanisms have been proposed by several authors. Clinically, symptoms are often nonspecific, leading to incidental diagnoses in many cases. Our patient presented with respiratory symptoms, including coughing, along with signs of asthenia and weight loss [3]

Radiologically, IMTs can present with diverse appearances. The most commonly described features include solitary or rarely multiple nodules or masses, often well-demarcated and located peripherally, with a predilection for lower lung lobes [4]. However, aggressive radiological signs, such as in our patient's case, may also be observed. Differential diagnosis via imaging must consider pulmonary lymphoma, cryptogenic pneumopathies, and solid mediastinal tumors like mature teratomas. Confirmation of diagnosis typically relies on histopathological examination postoperatively, mirroring the course of diagnosis in our case.

Surgical excision remains the cornerstone of treatment for IMTs, with a necessity for complete resection to achieve optimal outcomes. Fortunately, our patient's postoperative course was uneventful. Subsequent surveillance, primarily through CT and chest X-ray, is imperative. Following histologically confirmed complete excision, our patient underwent postoperative imaging without complications, as recommended for future annual follow-ups. [5]

Despite the benign nature of IMTs, the possibility of recurrence exists, with potential recurrence occurring even many years after initial surgery, emphasizing the importance of extended surveillance. Long-term follow-up for up to 15 years may be warranted due to the potential for late recurrence, as noted in the literature. [6,7]

This case underscores several key points regarding IMTs in the mediastinum. Firstly, the rarity of this tumor in this anatomical location highlights the need for continued awareness and research. The nonspecific clinical presentation of IMTs necessitates a high index of suspicion, particularly when encountering persistent respiratory symptoms, asthenia, or unexplained weight loss. Radiologically, the variability in imaging findings underscores the challenge of accurate preoperative diagnosis, necessitating histopathological confirmation postoperatively. [8]

The successful management of our patient's case through surgical resection and subsequent uneventful recovery aligns with current literature advocating for complete excision as the primary treatment modality. Furthermore, the emphasis on long-term surveillance is critical given the potential for delayed recurrence.

In conclusion, while inflammatory myofibroblastic tumors of the mediastinum are rare entities, their clinical management requires a multidisciplinary approach involving clinical suspicion, diagnostic imaging, and definitive histopathological assessment. Continued reporting and study of such cases contribute to our understanding of this intriguing pathological entity, guiding optimal management strategies and long-term surveillance protocols for affected patients.

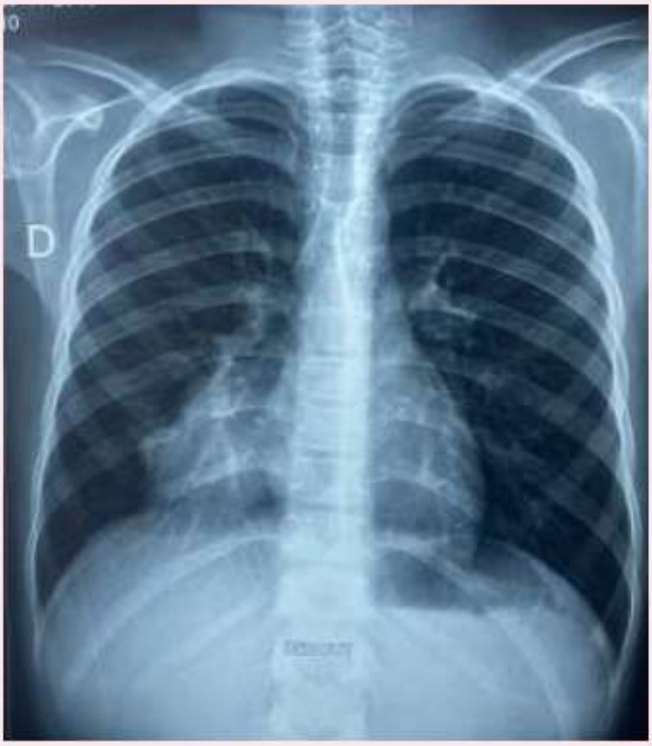


Figure 1:- Pulmonary X-Ray.

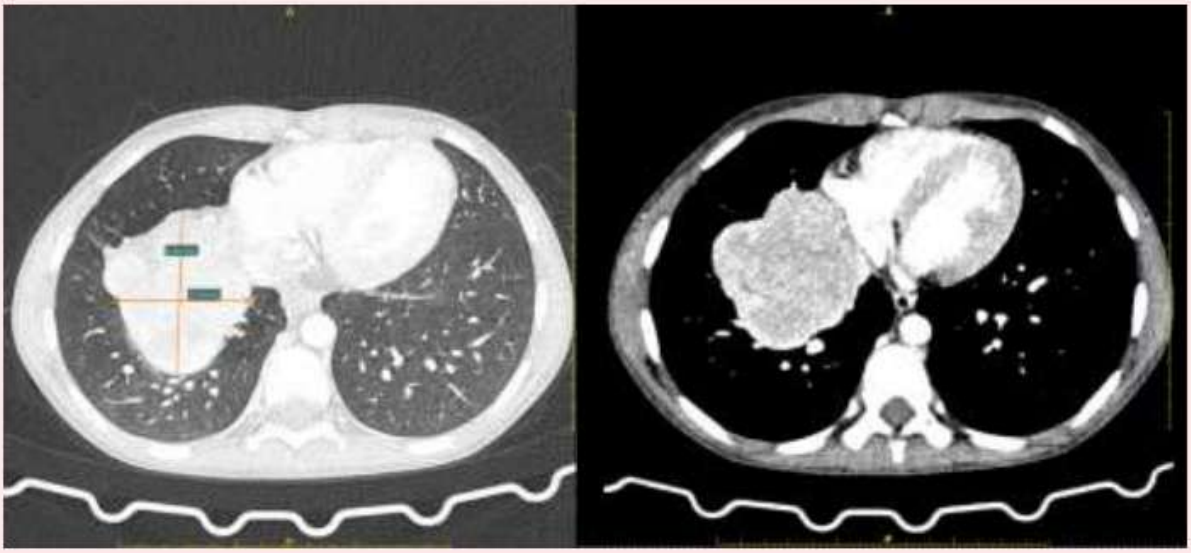


Figure 2:- Chest CT scan showing the mass.

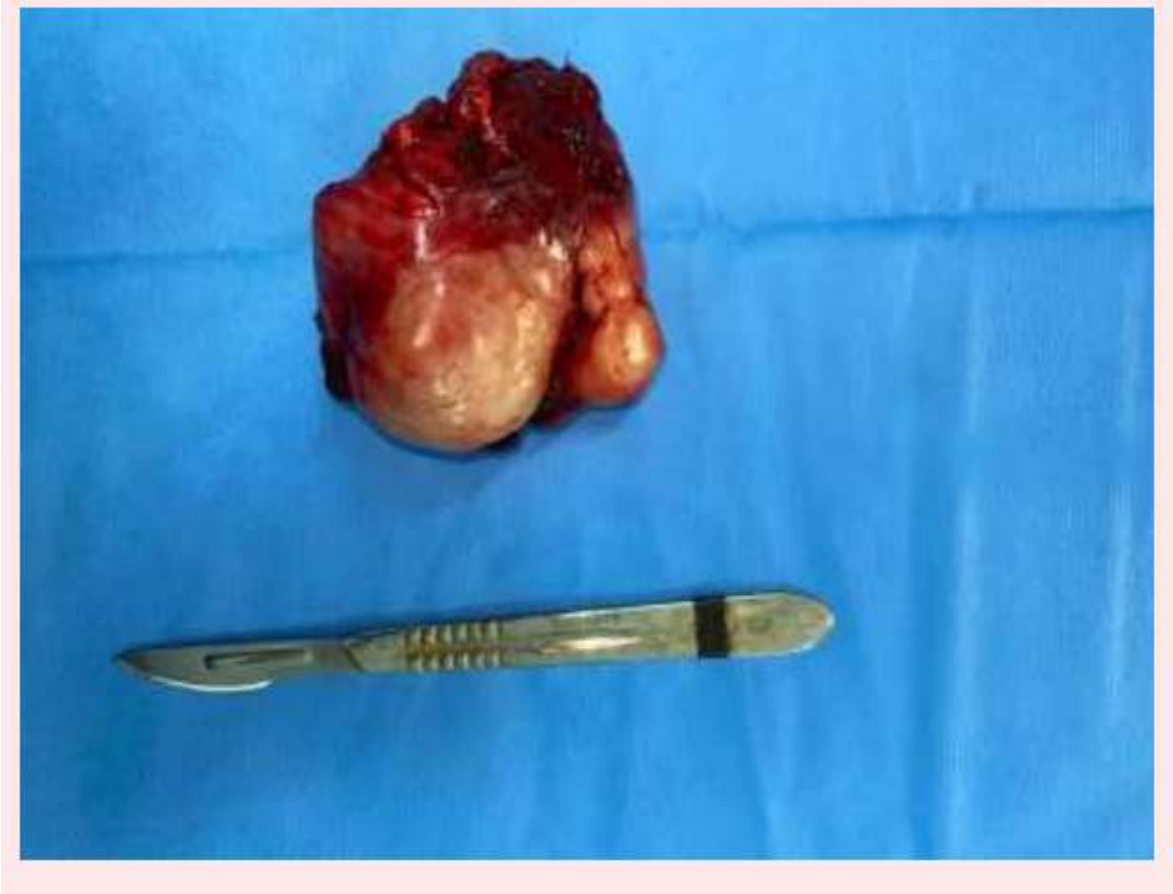
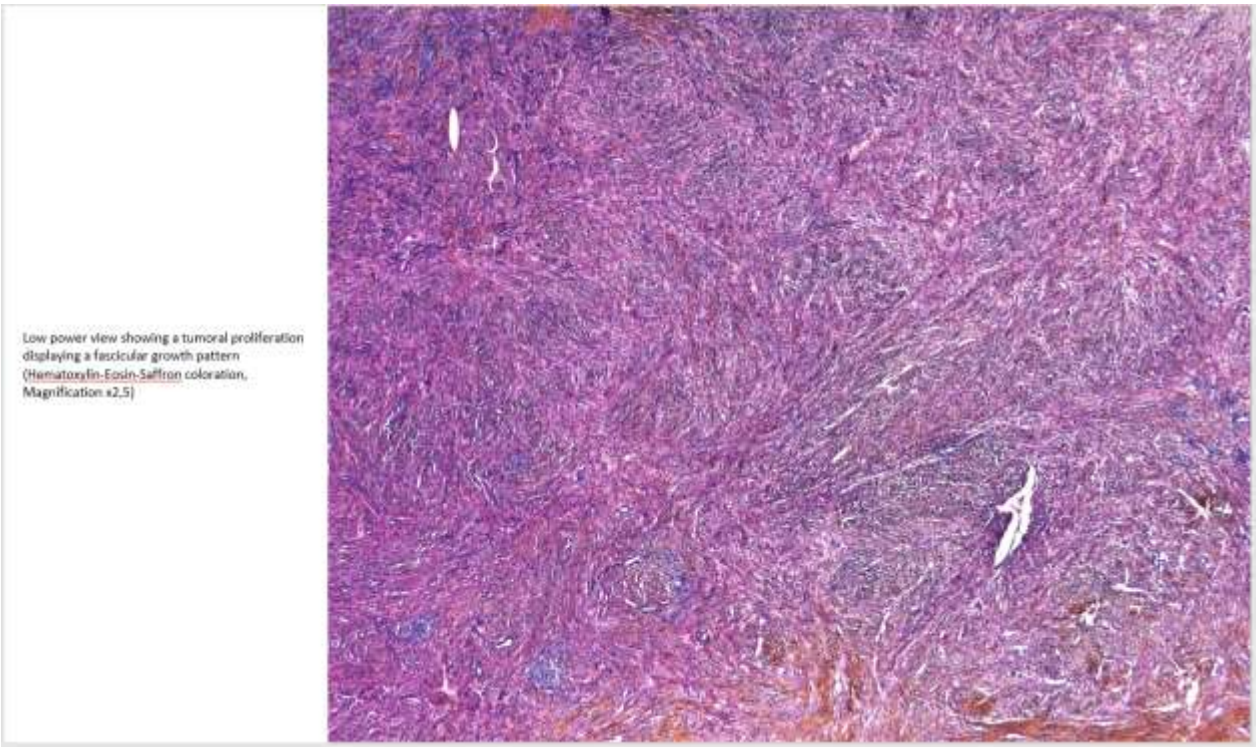
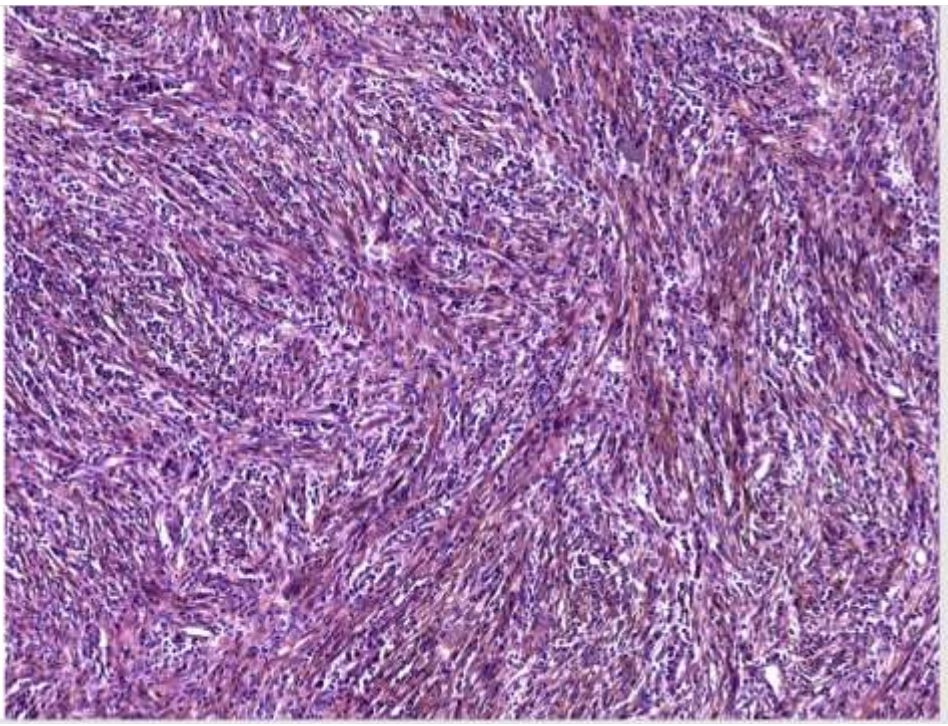


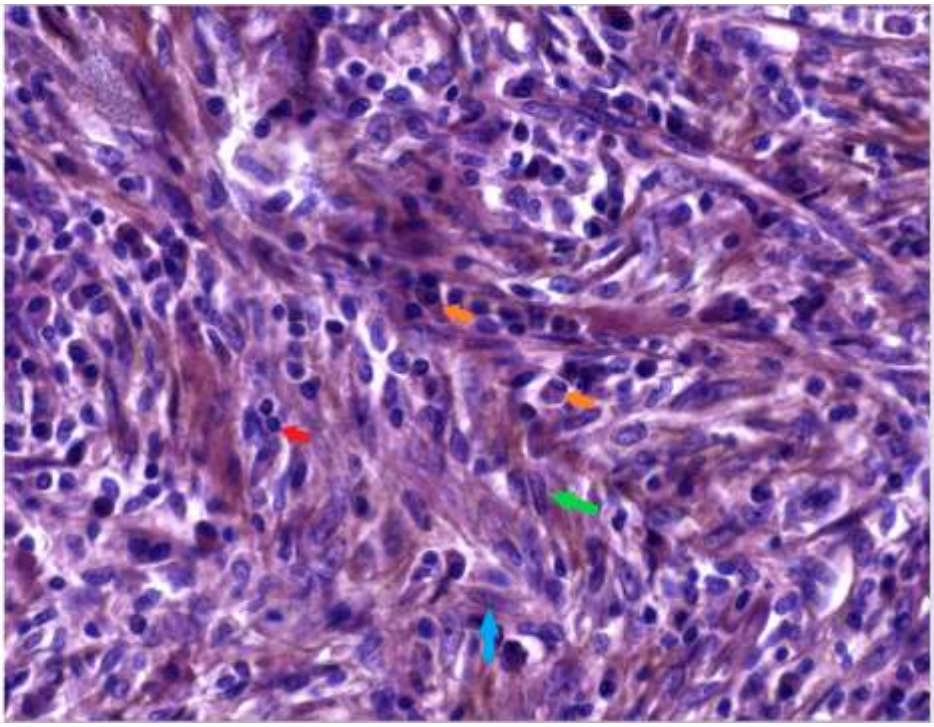
Figure 3:- Macroscopic aspect of the mass.

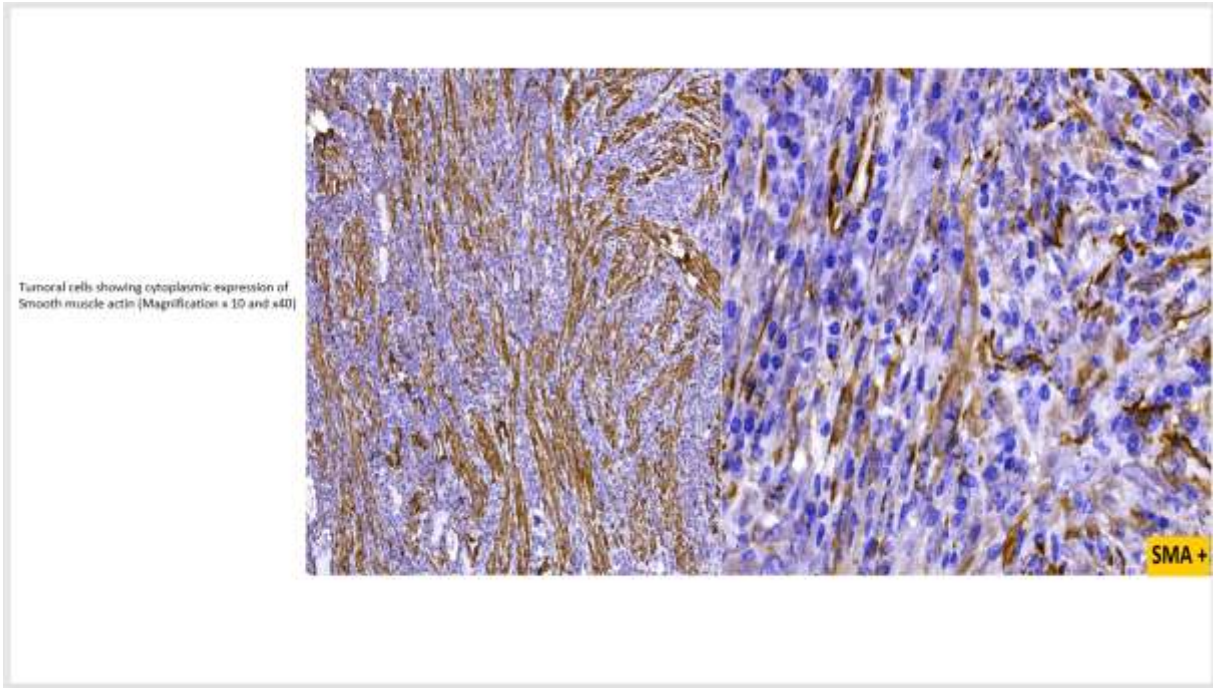


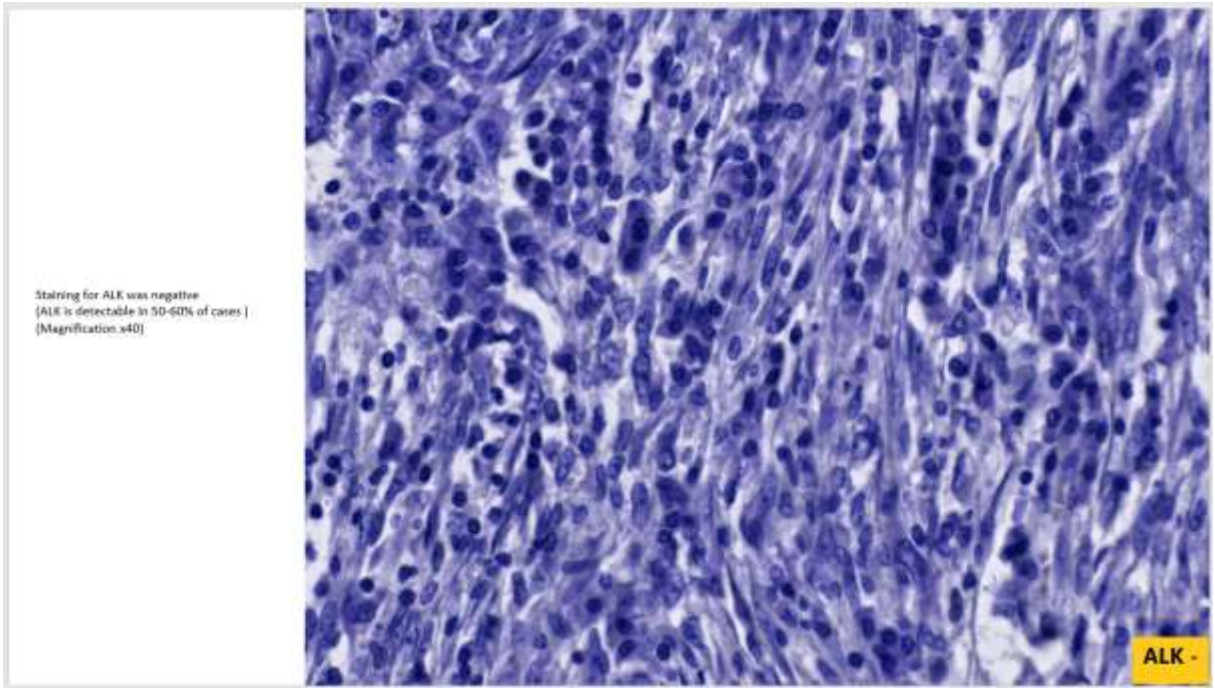
The tumor is highly cellular, formed of interlacing fascicles of tumoral cells, associated with a mixed chronic inflammatory infiltrate (Hematoxylin-Eosin-Saffron coloration, Magnification x10)



The tumoral cells are mainly spindle-shaped (Blue arrow) and occasionally epithelioid (Green arrow) with bland nuclei, fine chromatin and inconspicuous nucleoli . The tumoral stroma is densely inflammatory, typically rich in plasma cells (Orange arrow) and lymphocytes (Red arrow) (Hematoxylin-Eosin-Saffron coloration, Magnification x40)







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