Imaging features of inflammatory myofibroblastic tumor (IMT) and literature review.

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Purpose

Our purpose is to review thoracoabdominal imaging findings of inflammatory myofibroblastic tumor (IMT) through the analysis of our database and to assess a comprehensive overview of the related literature available.

Methods and Materials

A retrospective analysis of 13 histological proved thoracoabdominal IMT cases over a 10-year period was undertaken at our hospital. Patients were analyzed for clinical data, namely symptoms and previous diseases. Imaging findings were evaluated focusing on its anatomical distribution and on its patterns of presentation. CT was performed in 11 patients; only two of them underwent MRI. Core biopsy was performed in all cases. A review of the current literature is provided.

Results

Results:

- We report four cases of lung IMT’s, one of them with endobronquial origin and progressive transbronquial involvement. A thoracic wall IMT is illustrated and described. A cardiac IMT also occurred during this period.
- The abdominal IMT’s occurred in the liver, retroperitoneum, mesentery, small bowel, bladder and urachus.
- Most of them presented as solitary and sharply circumscribed masses with heterogeneous attenuation and variable patterns of enhancement; these imaging findings were consistent with those described in the reported cases available in literature.
- Histological characterization confirmed the diagnosis of IMT in all cases, except one, for which a definite diagnosis between retroperitoneal IMT and retroperitoneal fibrosis couldn’t be established.
- Most of these tumors were successfully removed with no further recurrence of the disease. There was no increase in size and no malignant transformation for the unresected remain tumors, and in the case of the hepatic IMT, there has been a significant spontaneous size reduction over time.

Literature review
• **Definition**

IMT is a rare and distinctive entity composed of myofibroblastic spindle cells and polyclonal inflammatory infiltrates.

• **Synonyms**

A lot of names have also been used to describe this condition, like plasma cell granuloma (heart), xanthomatous pseudotumor, plasma cell-histiocytoma complex (lung) and Inflammatory fibrosarcoma (bladder).

• **Demographics**

It can be detected at any age, but it has a higher incidence in the first two decades of life.

• **Location**

Although it has a widespread location, lungs, orbit, mesentery and omentum are the most common sites of involvement.

• **Etiology**

It is unknown, but some authors describe it as low-grade fibrosarcomas with inflammatory cells. It has also been associated with prior occult infection, minor trauma and surgical procedures.

• **Presentation**

The presence of symptoms varies with tumor location and size.

• **Imaging findings**

There are no specific imaging findings associated with this disease. **Ultrasound**: it commonly appears as a well or partially defined solid lesion.

**CT scans**: it is has a heterogeneous attenuation and enhancement and might have internal calcifications. When it is large, it can be centrally hypodense due to central necrosis.

**MR imaging**: it is usually hypointense on T1-weighted images, hyperintense on T2-weighted images, and heterogeneously enhanced after contrast intravenous administration.

• **Definitive diagnosis**

Histological characterization is required for a definitive diagnosis and to prevent unnecessary radical surgery procedures, once it can simulate malignant conditions, either clinically or radiologically.
• **Prognosis**

Good prognosis, although it has a propensity to be locally invasive and, in rare cases, it can undergo malignant transformation. Spontaneous remission of the disease has also been reported.

• **Treatment**

Surgical excision is the treatment of choice and local recurrence can occur in 25% of cases.

Benign fibrous tumors (fibromatosis, sclerosing inflammatory conditions, nodular fasciitis)

Soft-tissue sarcomas

Lymphoma

Metastatic disease

Embryonal rhabdomyosarcoma (mainly in children)

Gastrointestinal stromal tumor

Inflammatory malignant fibrous histiocitoma

**Differential diagnosis of IMT**

**Images for this section:**

![Images](image1.png)

**Fig. 1:** Fig. 1- Right upper lobe mass in a young child with chest pain and dyspnea, representing a pulmonary IMT. Chest radiograph (A) showed a mass at the right upper lobe that simulated a lobar pneumonia. Coronal MR images (B,C,D) confirmed the presence of a heterogeneous mass surrounded by reactive edema. The tumor was successfully resected with no further relapse of the disease.
Fig. 2: Fig. 2- Endobronquial IMT in a symptomatic man. Chest radiograph (A) showed right hilar enlargement and loss of volume of the homolateral lung. Axial NECT (B,C) identified a right obstructive endobronquial mass with transbronquial extension; mucous plugs at the upper lobe bronchi were also detected. The referred lesion was removed by surgery with an excellent outcome for the patient.

Fig. 3: Fig. 3- Chest wall IMT. Axial CT images before (A) and after (B) contrast administration showed a soft tissue enhancing mass at the right lateral chest wall. Coronal (C) and sagittal (D) CECT reformations are also shown.
**Fig. 4**: Liver IMT in an asymptomatic woman. US (A) showed a right lobe hepatic heterogeneous and very hypoechoic mass. The patient underwent MR imaging which detected a well-defined solid lesion that showed intermediate signal intensity on T2-weighted images (B). Hypervascularity of the lesion was demonstrated after gadolinium administration during arterial (C), venous (D) and late (E) phases. Coronal T2-weighted reformation is also assessed (F). Hystological characterisation was required for the correct diagnosis. A wait and see approach was chosen with partial reduction in size over time. Images courtesy of Otilia Fernandes.
Fig. 5: Small bowel IMT in a woman with abdominal pain. Axial NECT (A) showed a partially-obstructive intraluminal ileal mass which was furtherly surgically removed; small bowell proximal dilatation and little intraperitoneal fluid were additional findings.
**Fig. 6:** Fig. 6- Omental IMT in a patient with recurrent abdominal pain. Ultrasound (A) showed a hypoechoic ill-defined omental mass. Axial (B), coronal (C) and sagittal (D) CECT showed an oval, encapsulated, hypodense lesion lying adjacent to the descending colon. Fat infiltration of the surrounding omentum was also evident. The patient denied tumor removal after biopsy and there has been little size variation since the diagnosis.

![Fig. 6 Images](image1)

**Fig. 7:** Fig. 7- Urachal IMT. Axial T1-weighted (A), T2-weighted (B) and T1 Fat Supressed contrast-enhanced (C) images showed a large and heterogeneously enhancing urachal mass depressing the dome of the urinary bladder and displacing the uterus. Coronal T2-weighted (D) and sagittal T2-weighted and T1 Fat Supressed contrast-enhanced (E) images show the referred mass ( ); a uterine myoma is an additional finding (*). The urachal tumor was totally removed and no further recurrence occurred.

![Fig. 7 Images](image2)
**Fig. 8:** Fig. 8- Retroperitoneal IMT versus retroperitoneal fibrosis. Axial (A,B), coronal (C) and sagittal (D) abdominal CT images illustrated a rind of soft tissue density surrounding the aorta, the inferior vena cava and the iliac vessels. Abdominal plain radiograph (E) shows the bilateral stents, which had been placed for previous bilateral hydronephrosis. Although medial ureteral deviation and ureteral obstruction are classic findings of retroperitoneal fibrosis and unusual for IMT, histological analysis wasn’t able to differentiate these two conditions.
Conclusion

Although IMT is a rare entity, it includes a wide range of either benign or malignant conditions that can be found within numerous organs. Despite the correct diagnosis cannot be made upon its clinical and imaging features, which are quite nonspecific, the radiologist must be aware of them, once they are commonly mistaken for malignant diseases. Definite diagnosis, essential to preclude unnecessary radical surgery, is based on histopatological analysis.

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