

Inflammatory Myofibroblastic Tumour Mimicking Malignancy: does imaging help?

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Learning objectives

Inflammatory myofibroblastic tumor (IMFT) is a rare neoplasm of mesenchymal origin, primarily in soft tissues seen in many different anatomical locations. Our objectives are:-

- To review the spectrum of inflammatory myofibroblastic tumour in different organs.
- To illustrate the multi-modality imaging workup of such lesions.
- To increase awareness and recognition of uncommon presentations and locations of these lesions.

Background

Inflammatory pseudotumor was first observed in the lung and described by Brunn in 1939 and was so named by Umiker et al in 1954 because of its propensity to clinically and radiologically mimic a malignant process. The term "inflammatory pseudotumour" has been used to describe a wide range of reactive and neoplastic lesions, including inflammatory myofibroblastic tumour (IMFT), pseudosarcomatous myofibroblastic proliferations of the genitourinary tract, infectious and reparative processes, and inflammatory pseudotumours of lymph node, spleen and orbit. Over the last two decades, IMFT has emerged as a distinct entity with characteristic clinical, pathological and molecular features. However, confusion remains regarding the distinction of these tumours from other lesions in the "inflammatory pseudotumour" family, as well as from non-neoplastic fibrosclerosing processes and malignant neoplasms with a prominent inflammatory infiltrate.

Inflammatory pseudotumour more common affects the lung, where it was considered a reparative postinflammatory condition rather than a neoplastic process. But it has been reported to occur in nearly every site in the body.

IMFTs have been reported to occur mainly in children and young adults. IMFTs in the pediatric patient have clinical importance because the lesions often mimic malignant neoplasms, such as sarcomas, lymphomas, or metastases. Histologically, the lesion is composed of myofibroblastic spindle cells, accompanied by plasma cells, lymphocytes, and eosinophils. Immunohistochemically, the myofibroblastic spindle cells can be positive for vimentin (99%), smooth muscle actin (92%), muscle-specific actin (92%), desmin (69%), cytokeratin (36%), CD68 (KP-1) (24%), and CD30 (Ki-1) (6%). Cytoplasmic reactivity for ALK has also been demonstrated in approximately 50% of these lesions.

Ultrastructurally, the myofibroblastic cells display poorly developed Golgi, abundant rough endoplasmic reticulum, extracellular collagen, and intracytoplasmic thin filaments

and dense bodies. Genetic rearrangements involving 2p23, to which the ALK receptor tyrosine kinase gene has been mapped, are also often demonstrated.

The biological behavior of inflammatory myofibroblastic tumors is somewhat controversial. World Health Organization classification places inflammatory myofibroblastic tumors in an intermediate category (rarely metastasizing, in 5%) between benign and malignant.

The treatment options are varied and consist of surgery, high-dose steroids, irradiation, and chemotherapeutic agents.

To the best of our knowledge, the radiologic features of IMFTs in the pediatric patients have been described in only a limited number of patients.

Findings and procedure details

The radiologic features of IMFT are variable and nonspecific, possibly because of the amount of fibrosis and cellular infiltration.

- **US:** lesions can be hypoechoic or hyperechoic with either ill defined or well-circumscribed borders. These lesions often have increased vascularity during color or power Doppler examinations.
- **CT:** - shows varying appearances; lesions can have low, equal, or high attenuation compared with the surrounding tissue.
- **MRI:** - It usually has low signal intensity on both T1- and T2-weighted images, which may reflect the fibrotic nature of these lesions.
- **Contrast-enhanced CT and MRI** may show a homogeneous or heterogeneous lesion. Delayed imaging often shows increasing enhancement due to the presence of fibrosis.

1. 1. Lung IMFT

Inflammatory pseudotumor first observed in the lung and described by Brunn in 1939, was so named by Umiker et al in 1954 because of its propensity to clinically and radiologically mimic a malignant process.

Patients with IMFT are usually asymptomatic, with a solitary pulmonary nodule or mass detected on routine chest roentgenogram. It can behave as a malignant tumor both clinically and radiologically. Cough, fever, dyspnea, and hemoptysis are the usual presenting symptoms.

Radiological features of IMFT are variable and nonspecific. Although many pseudo tumors can be diagnosed presumptively on chest radiographs, a CT scan of the thorax may be necessary for a definitive diagnosis. The diagnosis of a pulmonary pseudotumor should be considered any time a lenticular opacity is identified superimposed on the central portion of the lung on a chest radiograph. CT features of pulmonary IMFT has been found to be typically a solitary, peripheral, sharply circumscribed mass with an anatomic bias for the lower lobes. Local invasion and primary involvement of the mediastinum and hilar structures are unusual manifestations. CT calcifications within the lesion occur more frequently in children than in adults. They are useful for differential diagnosis if present, but they are usually non-specific in shape and configuration. If present calcification pattern ranges from an amorphous, mixed, or fine flecklike pattern to heavy mineralization. Atelectasis and pleural effusion may occur. Cavitations and lymphadenopathy are very rare. Multiple lung lesions, pneumonic consolidation, atelectasis, hilar masses, and cavitations are unusual findings. CT and MRI are helpful for demonstrating the extent of the potentially aggressive disease. Lung IMFT is an uncommon cause of solitary lung nodule (0.7 % of lung tumors).

1. **2. liver IMFT**

Hepatic involvement by inflammatory pseudotumors was first described in 1953 by Pack and Baker. The majority of hepatic inflammatory pseudotumors occur in children and young adults. Most cases have been solitary solid tumors, mainly arising from the right hepatic lobe. In a few cases, inflammatory pseudotumor has involved the porta hepatis or bile ducts, which results in obstructive jaundice.

Patients generally present with a mass or nonspecific symptoms, including vague abdominal pain.

The hepatic IMFT mass appears heterogeneous echogenicity on sonography.

Low attenuation with a peripheral faint enhancing rim on contrast enhanced CT images. (Fig. 2).

The other form of hepatic IMFT appears as ill-defined, heterogeneously enhancing infiltrative lesion at the porta hepatis, with adjacent intrahepatic bile duct dilatation on CT images (Fig.7).

1. **3. Splenic IMFT**

A splenic inflammatory pseudotumor was first described in 1984 by Cotelingam and Jaffe, who categorized the lesion as a spectrum of non-neoplastic, inflammatory and reparative

changes. The main differential diagnoses of splenic IMFT include hamartoma, vascular neoplasm, follicular dendritic cell tumor and malignant lymphoma.

Splenic IMFT are discovered incidentally upon screening and symptoms roughly correlate with the lesion size. Symptomatic patients most often manifest left upper quadrant pain, fever of unknown origin, weight loss, malaise, and occasionally idiopathic thrombocytopenic purpura.

Laboratory findings are also non-specific include leukocytosis, anemia, thrombocytopenia, hypergammaglobulinemia, an elevated erythrocyte sedimentation rate, hypercalcemia, and/or elevated serum levels of soluble interleukin (IL)-2 receptor.

The splenic mass is most often appeared as low density with variable enhancement via CT, hypoechoic by US (Fig. 3), and either hypovascular or avascular following angiography. The most common preoperative diagnosis is lymphoma. Splenectomy is both diagnostic and curative, and results in an excellent prognosis.

Mesentery or omentum-

IMFTs that involve the mesentery or omentum present with abdominal masses that are associated with fever and abdominal pain. Fever may be a manifestation of an inflammatory response, and abdominal pain maybe related to the compression effect of the tumor.

Mesenteric or omental IMFTs appeared as well-defined solid, mixed-echogenic masses within the mesentery as seen on sonography and prominent vascularity as depicted on Doppler sonography. On CT scans, a mesenteric IMFT shows typically heterogeneous attenuating enhancement. (Fig. 4). These lesions were considered sarcomas, lymphomas, or IMTs.

Maxillary IMFT

Maxillary IMFT has aggressive appearance on imaging than does its counterpart in the orbit (Fig.6). It appears as soft-tissue mass associated with bony changes of erosion, remodeling, sclerosis, and thickening. This imaging appearance can mimic an antral malignant tumor. Often, multiple biopsies are necessary to establish the diagnosis of inflammatory pseudotumor. The diagnosis is one of clinical-radiologic- pathologic exclusion. We reported two different pattern of IMFT in maxilla.

- The first one is a rather defined soft tissue mass lesion expanding the maxillary sinus destructing the medial wall and extending into the nasal

cavity, showing homogenous enhancement, destructing the medial wall and extending into the nasal cavity on CT. The lesion has signal intensity of soft tissue (hypointense T1WI, hyper intense T2WI) with vivid post contrast enhancement. ADC images show low signal denote restricted diffusion reflect hypercellular compact lesion. (Fig. 5)

- The second pattern is expansile lesion at the angle of the maxillary bone showing internal calcific foci, with no associated soft tissue masses. The lesion appears hot on the bone scan (Fig.7).

Images for this section:

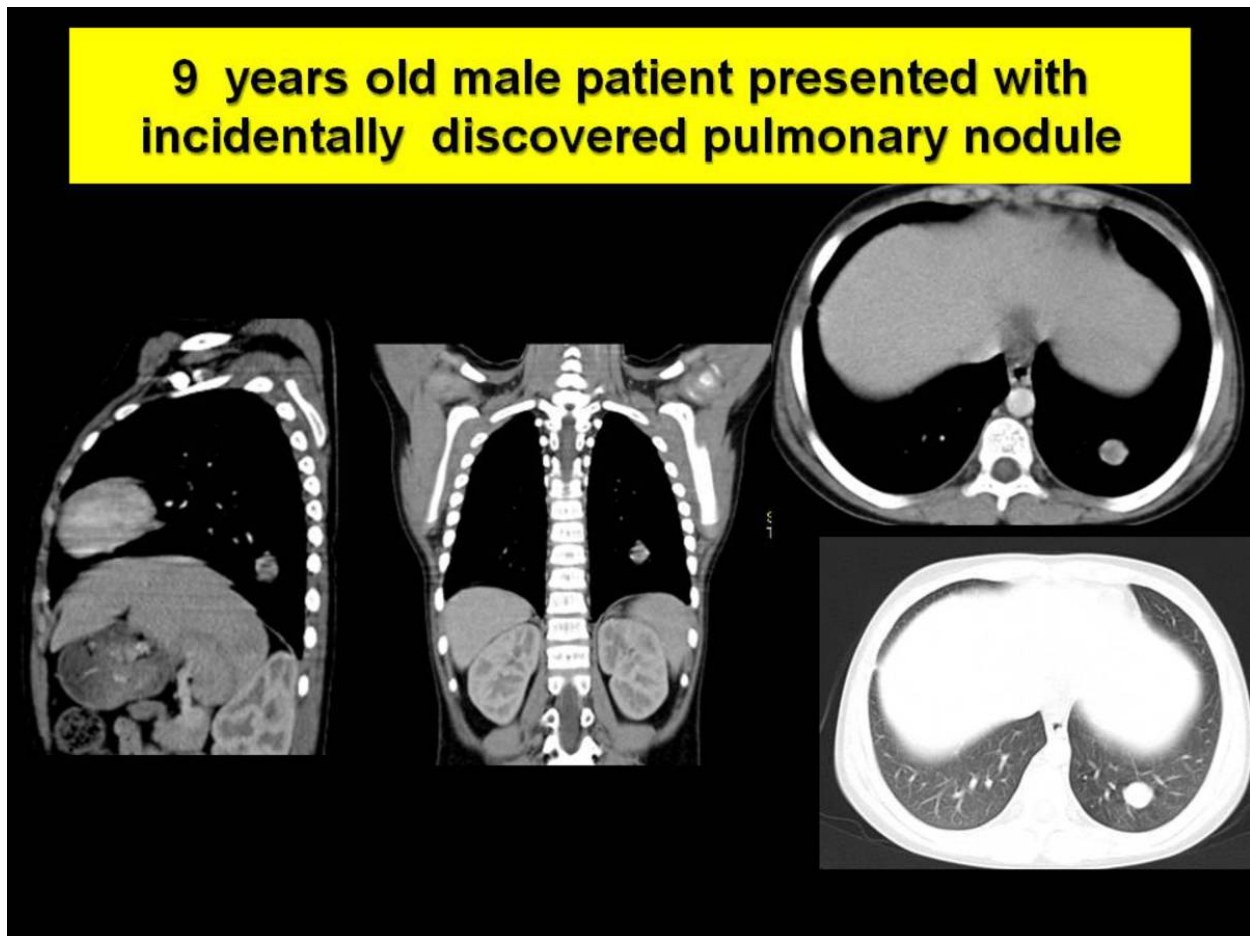


Fig. 1: A well defined hypo dense pulmonary nodule seen at the left lung base . No internal calcification , cystic degeneration or fatty component

17 years old male patient presented with incidentally discovered hepatic focal lesions

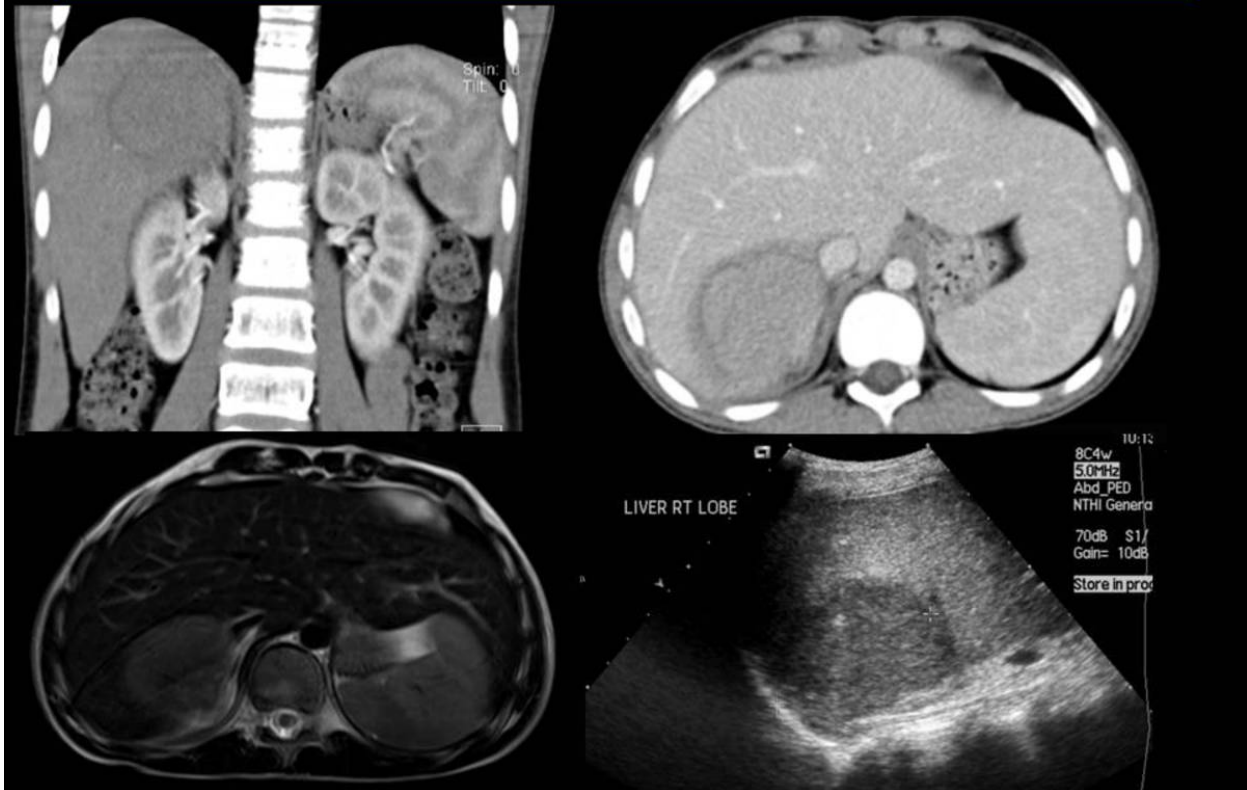


Fig. 2: A well defined oval shaped hepatic focal lesion is seen at segment 6 . It is seen less enhanced than the normal hepatic parenchyma. It shows hypo intense signal on T2wi MRI and hypoechogenicity on US . Pathological DX : IMFT

3 year male patient presented with abdominal mass.



Fig. 3: Large left hypochondrial mass lesion is seen exophytic from spleen compressing left kidney showing heterogeneous enhancement with no definite internal calcification . On US the lesion is echogenic with small cyst . No associated enlarged lymph nodes.

1 year old boy presented with abdominal discomfort .

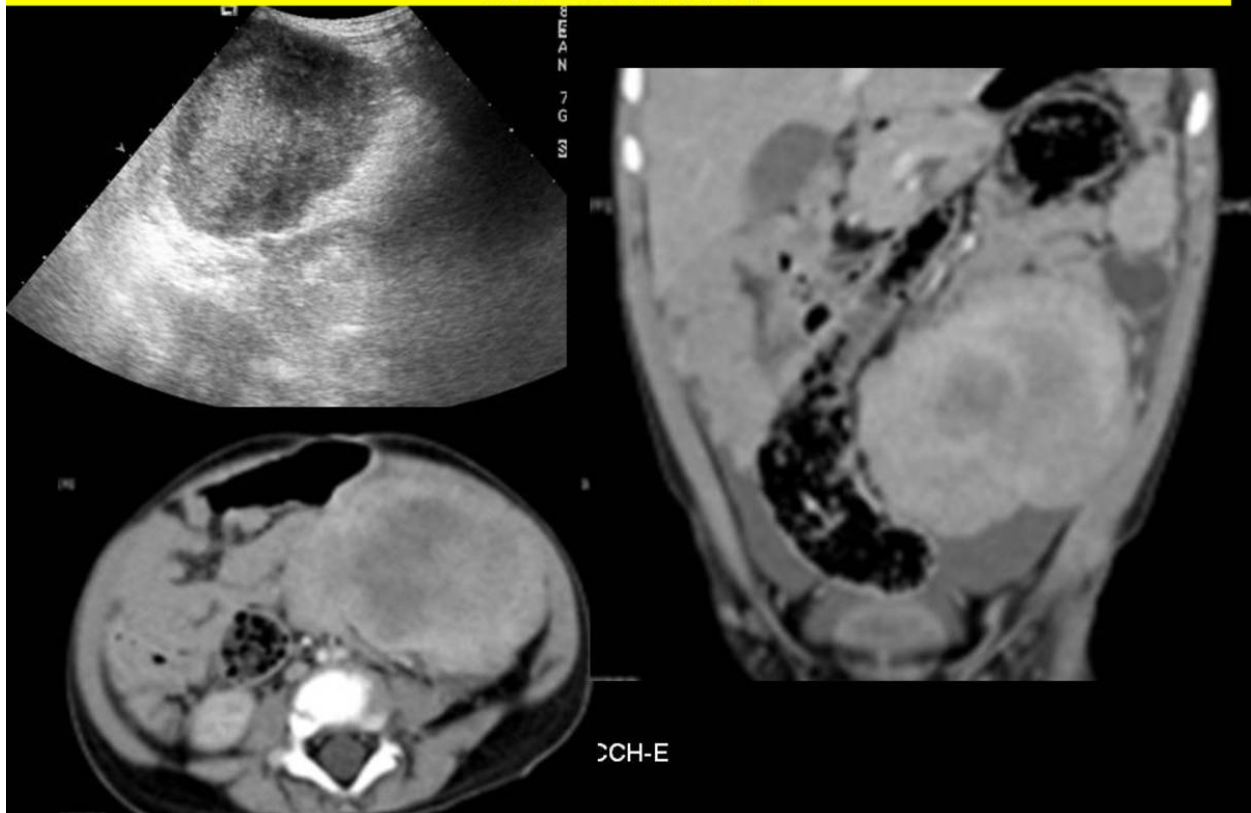


Fig. 4: A well defined enhanced mesenteric soft tissue mass lesion is seen at the left hypochondrial region . Diagnosed as a lymphoma . Surgical resection and pathological assessment revealed IMFT

6-year-old male presented with large left cheek painless slowly growing mass.

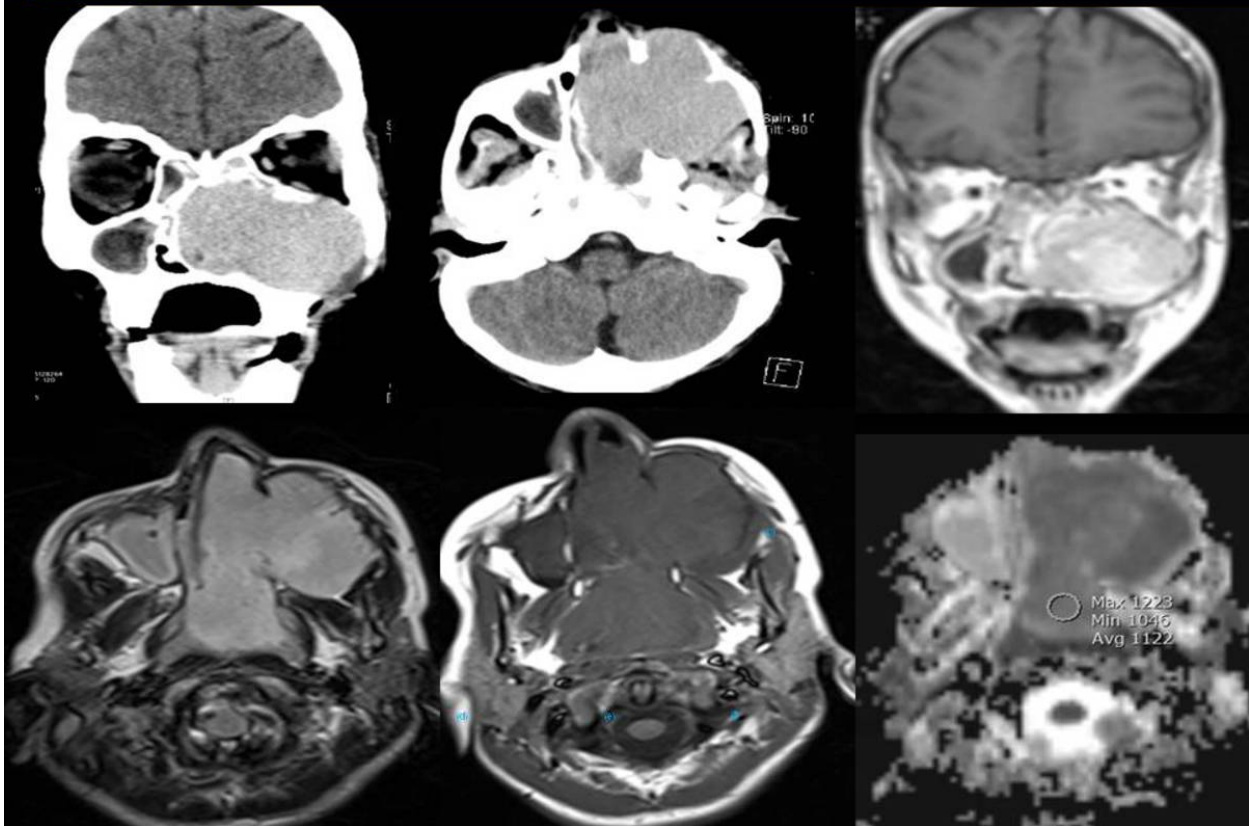


Fig. 5: CECT study of the facial region demonstrates a large , fairly well defined , enhancing soft tissue mass lesion occupying the left maxillary sinus, with destruction of its medial wall and extending into the nasal cavity. MRI shows:- the lesion occupying the left maxillary sinus, extending into the left nasal cavity medially, and to the sphenoid sinus posteriorly. The lesion has signal intensity of soft tissue (hypointense T1WI , hyper intense T2WI) with vivid post contrast enhancement. ADC images show low signal denote restricted diffusion reflect hypercellular compact lesion.

7 year female patient with left orbital swelling.

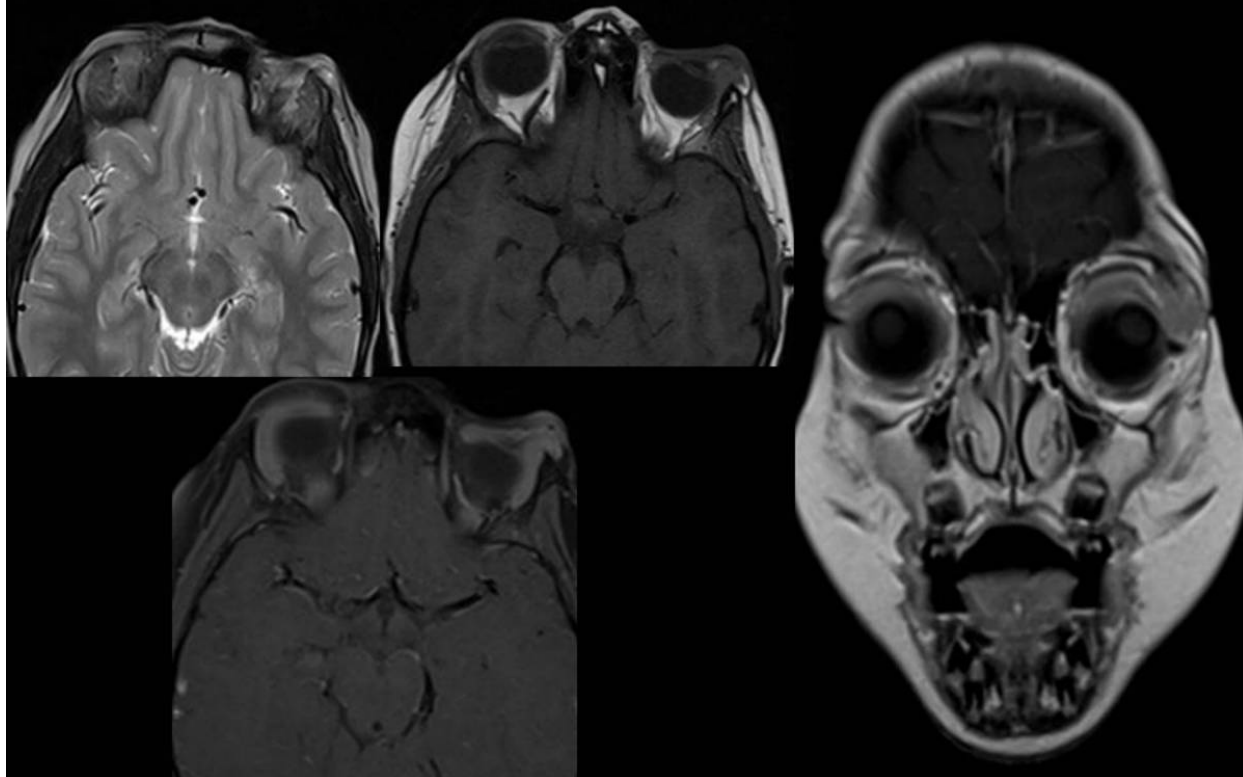


Fig. 6: A soft tissue sheet like is seen at the superior aspect of the left orbit , no underlying bone affection , no invasion of the eye globe . The lesion elicits hypo intense on T1WI & T2WI with uniform enhancement on post Gd series

13 years old male patient present with bony pains



Fig. 7: Multiple polyostotic expanding bony lesions seen at the right maxilla and sternum (not shown). These lesions appear hot on bone scan. Heterogeneous density of liver and enlarged caudate lobe is also noted. Multiple bilateral pulmonary nodules are seen on a background of ground glass opacification of both lungs. Pathological dx: IMFT

Conclusion

Although an IMT is essentially a benign disease, an IMT in the pediatric abdomen can be problematic from both a diagnostic and a therapeutic standpoint. In spite of differentiation from malignant lesion is not clear-cut, the radiologist should be aware of these entities to avoid unnecessary radical surgery.

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