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Case Report

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# A Case Report on Retroperitoneal Inflammatory Myofibroblastic Tumor - A Relatively Rare Neoplasm

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## Abstract

Inflammatory mypfibroblastic tumor (IMT) is a rare tumour with unpredictable clinical presentation. It belongs to neoplastic inflammatory spindle cell lesion. People with IMT can be asymptomatic, some may have nonspecific respiratory symptoms, fever, or pain. Here we present a case of 56-year-old male who was incidentally detected with lump in the abdomen on ultrasound abdomen during evaluation for fever. On further evaluation on imaging with abdominal CT (Computerized tomography) angiogram, it was diagnosed as a highly vascular retroperitoneal mass lesion. Total surgical resection of the retroperitoneal tumour was done. Histopathological examination of the specimen was in favor of inflammatory myofibroblastic tumour.

## Keywords

Inflammatory mypfibroblastic tumor, Retroperitoneal mass

## Introduction

World Health Organization (WHO) 2020 has defined IMT as a distinctive, rarely metastasizing neoplasm constituting myofibroblastic and fibroblastic spindle cells accompanied by an inflammatory infiltrate of plasma cells, lymphocytes and/or eosinophils [1]. IMT is distinct from mesenchymal tumor with malignant potential that is frequently observed in patients who are < 16 years old and is rarely observed in adults [2]. The definitive diagnosis is made by histological examination [3-5] and surgical resection is the treatment of choice [3, 5, 6].

## Case Report

A 56-year-old male presented to the hospital with complaints of fever with chills and rigors of three days duration. No urinary complaints/ENT complaints/cough/joint pains. No history of abdominal pain, no altered function of bowel and bladder, no weight loss, and no anorexia was detected. The Patient was treated for viral pyrexia and recovered well.

During evaluation, a lump in the abdomen was noted near the inferior surface of the left kidney. CECT (Contrast enhanced computerized tomography) and PET (Positron emission tomography) CT of abdomen showed large retroperitoneal mass in left paraaortic location below left renal hilum. Result of CECT and PET CT are shown in figure 1 and figure 2, respectively.

On further evaluation of the detected lump, no history of headache, palpitation, excessive sweating, no organomegaly, and no fluctuations in serial monitoring of blood pressure was detected. On clinical examination no lump was palpable per abdomen, and rectal and systemic examination was normal. Testis were normal in size, surface, and texture and had no palpable nodule.

Figure 3 shows abdominal CT angiography which revealed a well circumscribed heterogeneously hypodense lesion measuring 5.6 x 5 x 5.1 cm (CCxAPX-

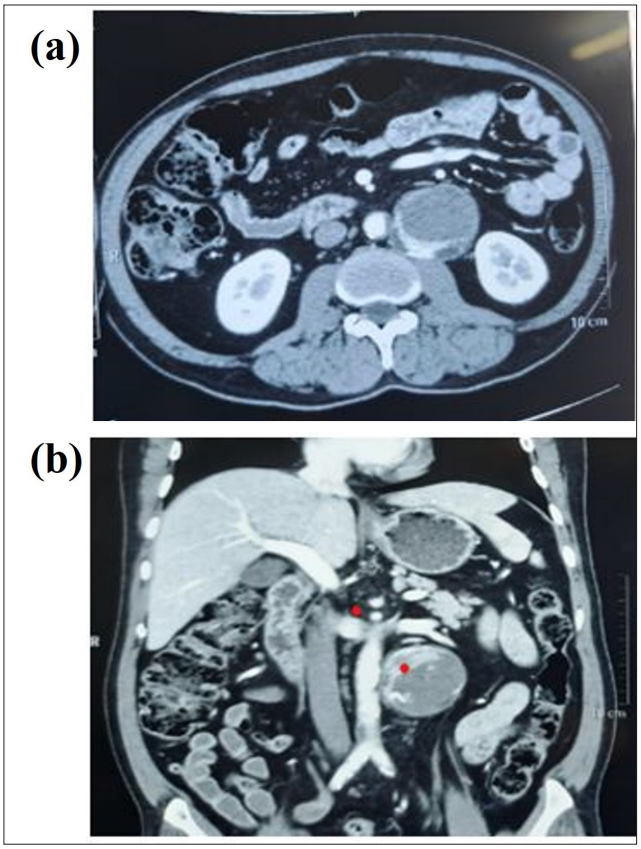


**Figure 1:** CECT abdomen showing large retroperitoneal mass in left paraaortic location below left renal hilum. **(a)** axial and **(b)** coronal views.

Tra) in the retroperitoneal space adjacent to abdominal aorta just below the left renal artery and inferior pole of left kidney at the level of L1/2. There was loss of interphase/fat planes between the left psoas muscle and the adjacent abdominal aorta. The lesion showed enhancing areas in the arterial phase and pooling of contrast in the subsequent phase - suggestive of highly vascular retroperitoneal mass lesion. No definitive communication with adjacent abdominal aorta was visualized. Treatment was done by exploratory laporotomy and total surgical resection of retroperitoneal tumour. As is shown in figure 4, a 7 cm x 5 cm firm, solid, and highly vascular soft tissue swelling in para-aortic region below the left renal vessels was resected.

Tumor has pushed the left renal vein superiorly, pushing the left kidney and left gonadal vessels laterally. Tumor was adherent to aorta medially with multiple feeder vessels from aorta and multiple lumbar veins draining the tumor.

The resected tumour is shown in figure 5, was sent for histopathology. As shown in figure 6, diagnosis was spindle cell tumour with necrosis favoring inflammatory myofibroblastic tumour. Further, immunohistochemistry examination was done. The immunohistochemical examination revealed negative staining for ALK and CD34, and positive result for vimentin (90%), Ki 67 < 1% (Low proliferative index), and SMA, as shown in figure 7.

## Discussion

IMTs are uncommon mesenchymal neoplasms which exhibit low to intermediate malignant potential and are com-

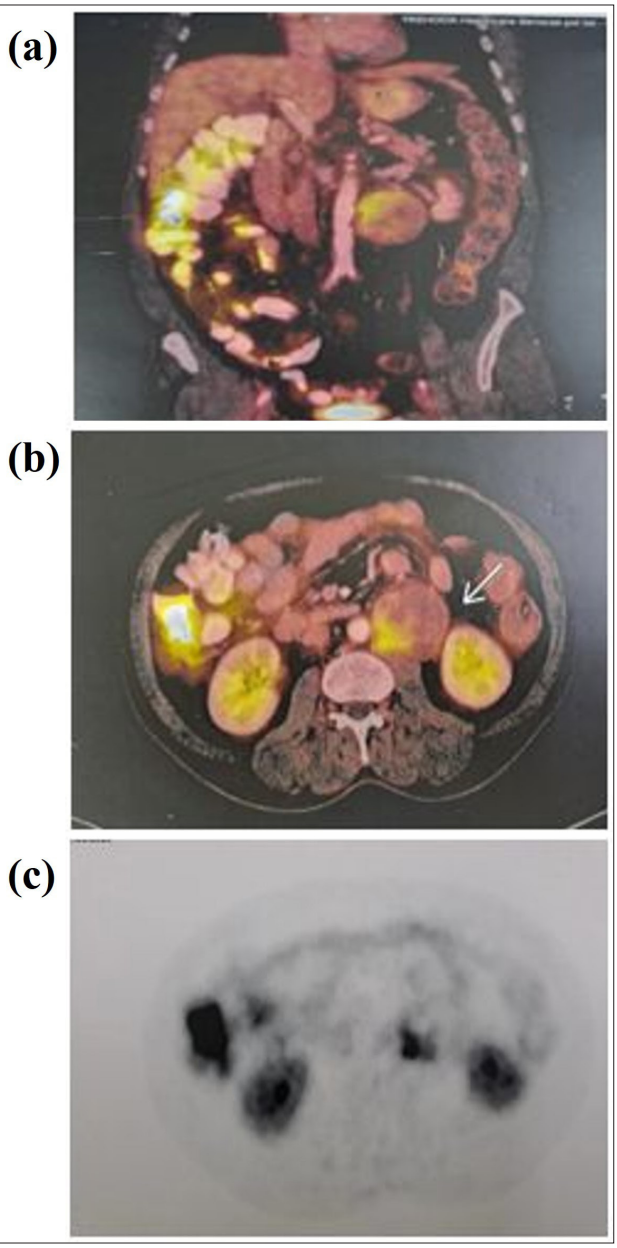


**Figure 2:** PET CT suggestive of paraganglioma.

posed of spindle shaped myofibroblasts often accompanied by the presence of inflammatory cells [7, 8]. IMT generally presents in patients < 16 years old [2], but also affects adults. Lungs are the most common anatomical location of IMT; however, any other site may be involved, including the mesentery, retroperitoneum, mediastinum, somatic soft tissues, larynx, uterus, bone, and central nervous system [7].

The etiopathogenesis of IMT is not well known. Injury, surgery, inflammation, and infection may contribute to the development of IMT [9]. Studies in genetics demonstrate chromosomal translocation and genes fusion [3, 5, 10-13].

Patients usually are present with single mass and nonspecific symptoms, which vary according to the location, size, and growth pattern of the tumor. Radiological features are also non-specific [7]. About 5 to 10% of the patients present with systemic and physical manifestations such as fever, weight loss, anorexia, microcytic and hypochromic anemia, hypergammaglobulinemia, and thrombocytosis [4, 5].

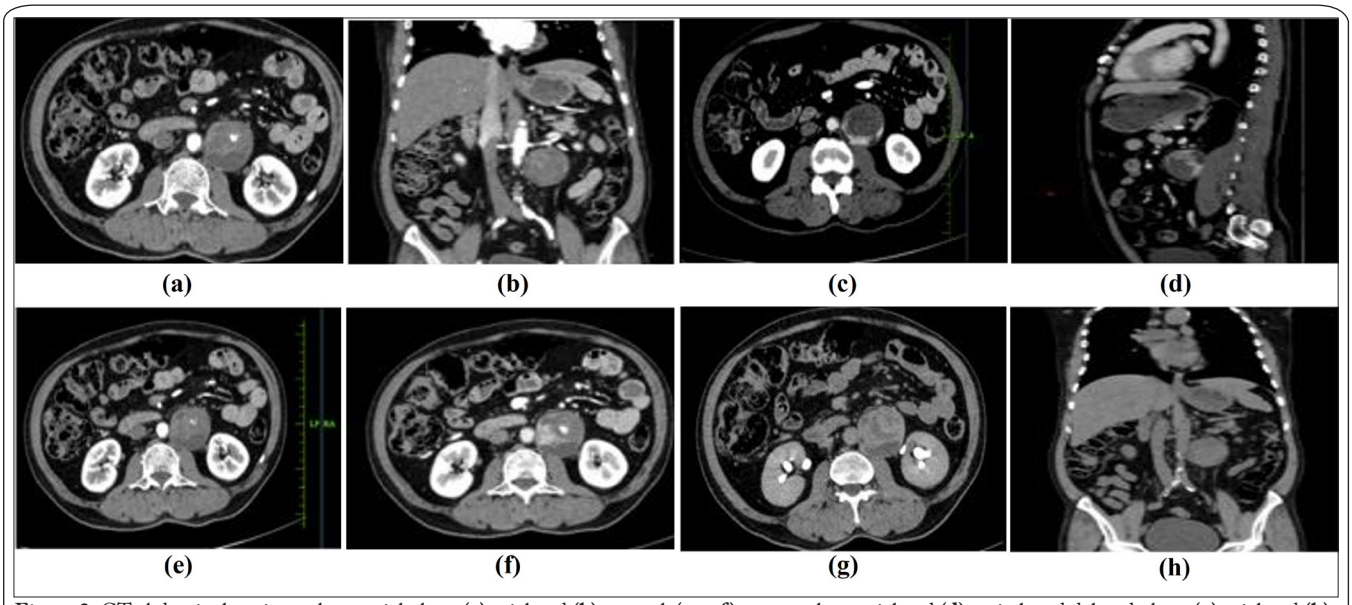


**Figure 3:** CT abdominal angiography arterial phase **(a)** axial and **(b)** coronal, **(c, e, f)** venous phase axial and **(d)** sagital, and delayed phase **(g)** axial and **(h)** coronal images suggestive of highly vascular retroperitoneal mass lesion, which is showing no obvious communication or leak from the abdominal aorta communicating with the para-aortic mass lesion.

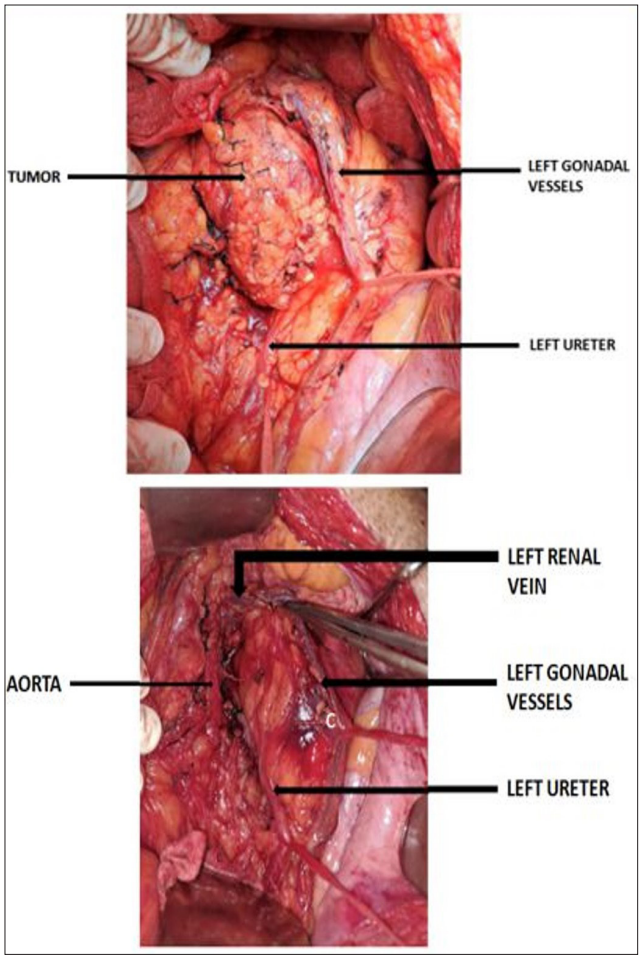


**Figure 4:** Intra OP images.

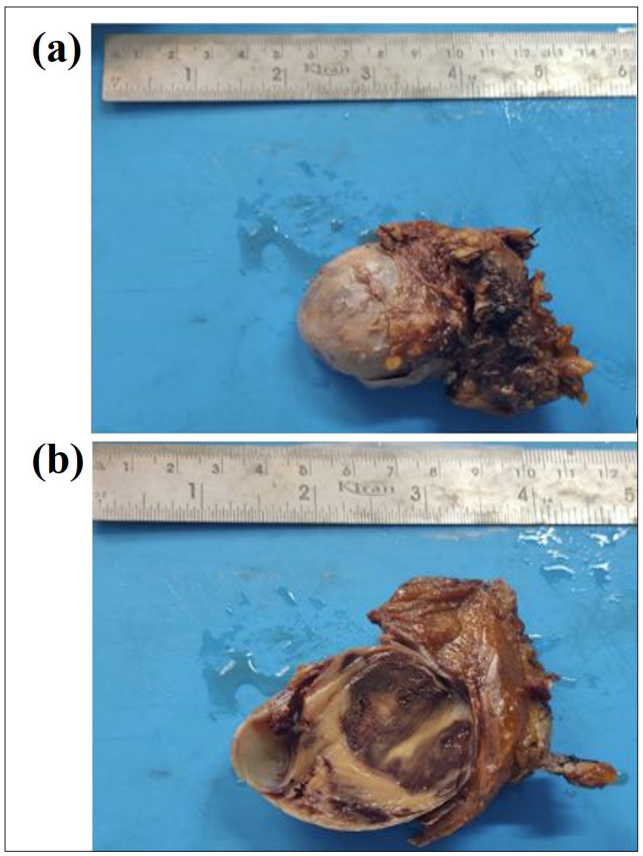


**Figure 5:** Gross specimen showing a well circumscribed lesion measuring 11 x 5 x 3 cm with attached pad of fat **(a)** on cut section it has variegated solid and cystic areas **(b)** solid areas show haemorrhage and necrosis and cystic areas drained haemorrhagic fluid.

Nonspecific radiologic findings of IMT in various locations have been reported in the literature. Variable density with homogenous or heterogeneous masses may be seen on CECT. Central necrosis may be seen in larger lesions. Varying degrees and patterns of contrast enhancement occur [14, 15]. Variable patterns of echogenicity with ill-defined or well-circumscribed margins may be seen on ultrasonography, while Doppler may reveal prominent vascular flow [15]. The appearance of these tumors is also variable on magnetic resonance images. They usually present as hypointense relative to skeletal muscle on T1-weighted images, hyperintense on T2-weighted images and heterogeneously enhanced after administration of contrast material [16]. These variable radiologic findings may

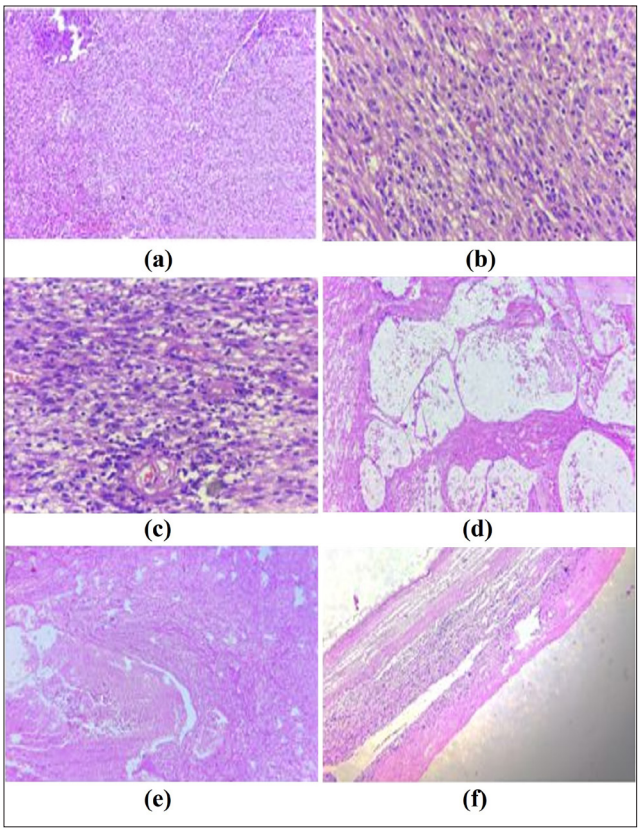


**Figure 6:** On histopathology the solid area showed a cellular tumour composed of spindle cells arranged in short fascicles **(a)** on higher magnification spindle cells are uniformly fair with vesicular nucleus and had adjacent inflammatory infiltrate consisting of lymphocytes and plasma cells, also seen are blood vessels **(b, c)**. Abundant dilated vasculature and extensive necrotic areas are noted **(d, e)** and cystic spaces showed fibrocollagenous cyst wall with infiltration of lymphocytes and plasma cells **(f)**.

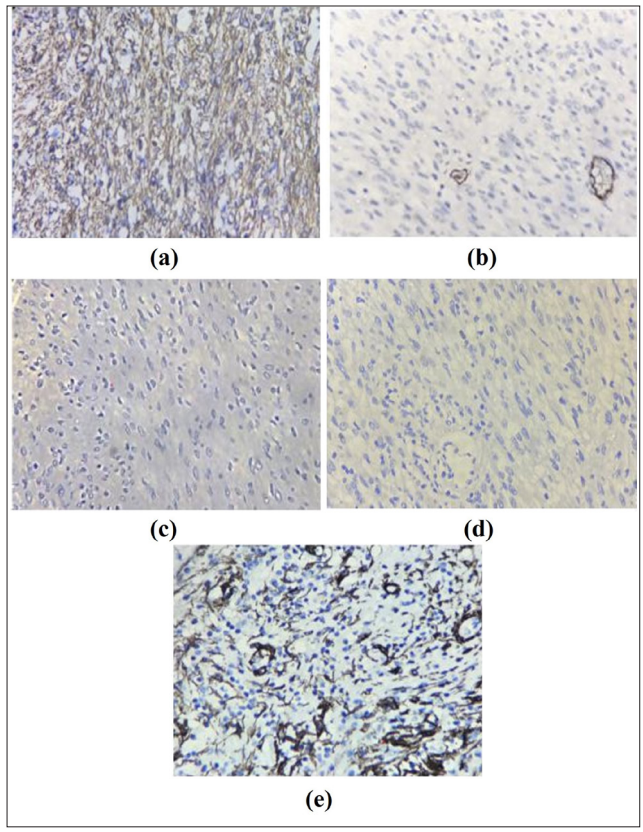


**Figure 7:** IHC vimentin 90% positive **(a)** negative for CD34 **(b)** negative for ALK **(c)** low Ki 67 (< 1%) **(d)** and SMA positive **(e)**.

be attributed to histologic complexity with the duration of the disease process, the amount of fibrous tissue and the degree of cellular infiltration. The differential which could be considered based on the imaging findings are solitary fibrous tumor, paraganglioma, myxofibro sarcoma, myxoid liposarcoma, pleomorphic liposarcoma, leiomyoma, leiomyosarcoma, and rhabdomyosarcoma.

Immunohistochemistry analysis was required after surgery to confirm the pathological diagnosis. The histological features of IMT include variable spindle cell proliferation in a myxoid to collagenous stroma, with prominent inflammatory infiltrate comprising primarily of plasma cells and lymphocytes, with the occasional presence of neutrophils and eosinophils [7]. Study of Immunohistochemistry is necessary to confirm the diagnosis. Mesenchymal cells are usually immunoreactive for vimentin, desmin, alpha-SMA, and S1008 protein and negative for C-Kit [17].

About 71% of these tumors are positive for ALK-1 [12, 17], which makes IMT more susceptible to drug treatment than those which do not express it [4], but with a higher recurrence rate [12]. Differential diagnosis includes benign lesions such as giant cell granuloma, solitary fibrous tumor, myoepithelioma, myxofibroma [11, 18], and malignant tumors such as low-grade myofibroblastic sarcoma, teratomas, rhabdomyosarcomas, and lymphomas [3, 11].

The management of IMT depends on its location, the ALK expression, its behavior, and the feasibility for surgical resection. The role of radiotherapy and chemotherapy is unclear [4, 17]. Since it has not shown a definitive benefit [5], surgical resection is treatment of choice [3, 5, 6, 12, 17].

Recurrence occurs in 25 to 40% patients and is often in extra-pulmonary lesions and in the first year after resection [3, 5, 11]. The recurrence rate is less than 10% (If the removal is complete and the lesion shows tumor-free margins) [3]. Metastases are infrequent and around 2% [12, 17]. The factors like female gender, age over 25 years, abdominopelvic location, large size, multinodular mass, incomplete resection, and ALK negative, predict worse prognosis of tumour with recurrence and metastasis probability [17]. Monitoring with ultrasound is suggested for follow up after resection, at 3, 6, and 12 months [3, 6].

## Conclusion

IMT is categorized as an intermediate behavioral myofibroblastic tumor. The clinical manifestations of IMT are diverse and will be determined by the affected anatomical area. Management depends on its location, ALK expression, behavior, and feasibility of surgical resection.

## Acknowledgements

None.

## Conflicts of Interest

None.

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