



CASE REPORT

INFLAMMATORY MYOFIBROBLASTIC TUMOR IN ANTERIOR ABDOMINAL
WALL-A CASE REPORT WITH RADIOLOGICAL REVIEW

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ABSTRACT

Inflammatory myofibroblastic tumor also called inflammatory pseudotumor is a quasi-neoplastic lesion which is a benign lesion. It is often clinically and radiologically mistaken for malignant condition. It is tumor mimic. Radiologist should be familiar with this entity which will help to avoid unnecessary radical surgery. It is a rare entity with higher incidence in younger patients and virtually can occur anywhere in body. Males and females are equally affected. We report a case of 20 year old female presenting with slightly painful swelling in anterior abdominal wall in left side of midline in lower abdomen. Imaging findings were suggestive of inflammatory myofibroblastic tumor or desmoid. Histopathology confirmed inflammatory myofibroblastic tumor.

INTRODUCTION

Case Report

A 20 year old unmarried female presented with firm swelling in left side of midline in lower abdomen since 1 month which was slightly painful. No evidence of fever or trauma. No evidence of swelling anywhere else in body. USG abdomen showed a well defined mixed echoic, predominantly hypoechoic solid lesion of size 2.9 (Transverse) x 2.1 (Antero-posterior) x 2.4 (Cranio-caudal) cm in anterior abdominal wall in midline subcutaneously in hypogastric region showing few tiny echoreflective foci of calcification and minimal vascularity on Doppler (Figure 1).



Figure 1 USG transverse section in anterior abdominal wall just on the left side of midline in hypogastric region - shows well defined hypoechoic solid lesion in subcutaneous plane with fine echoreflective calcific foci with adjoining echogenic fat.

Adjoining subcutaneous fat appeared thickened and echogenic suggestive of panniculitis. No history of alteration in size and pain in relation to menses was noted. Possibility of inflammatory myofibroblastic tumor was given rather than desmoid tumor or endometriotic deposit. CT scan of the abdomen (Plain) showed a well defined soft tissue density mass appearing isodense to muscle on plain study with few foci of central and peripheral calcification with ill defined soft tissue infiltrates in adjoining subcutaneous fat suggestive of fat stranding with no intra-abdominal extension (Figure 2a-b). Possibility of inflammatory myofibroblastic tumor rather than desmoid tumor or spindle cell tumor was given.



Figure 2a



Figure 2b

Figure 2 Plain CT Axial (a), Sagittal MPR(b)-shows well defined solid lesion in anterior abdominal wall just on the left side of midline in hypogastric region in subcutaneous plane nearly isodense with Muscle involving outer muscle fibres of left rectus abdominis With few calcific foci and adjoining fat stranding.

MRI abdomen done with axial T1, T2, T2 HASTE FS, sagittal and coronal T2 HASTE showed a well defined lesion measuring approx. 2.2 (Anteroposterior) x 2.8 (Transverse) x 2.5 (Cranio-caudal) cm in subcutaneous fat involving anterior fibres of left rectus abdominis. It appeared isointense to muscle on T1WI and slightly hyperintense to muscle on T2WI and STIR. Posterior fibres of left rectus abdominis were not involved. No intra-abdominal extension of the mass was seen. Adjoining subcutaneous fat and medial fibres of left rectus abdominis appeared hyperintense on T2 FS suggestive of edema. (Figure 3a-c) FNAC of the mass showed moderate cellularity, singly scattered and clusters of spindle cells showing mild nuclear pleomorphism with no evidence of mitotic activity with inflammatory cells. There was no evidence of frank malignancy. Cytology was suggestive of myofibroblastic tumor (Figure 4).

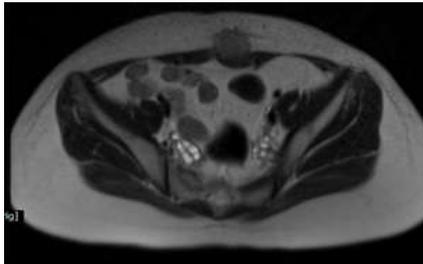


Figure 3a

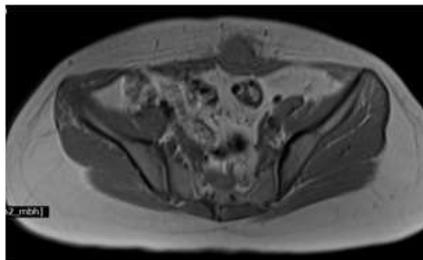


Figure 3b

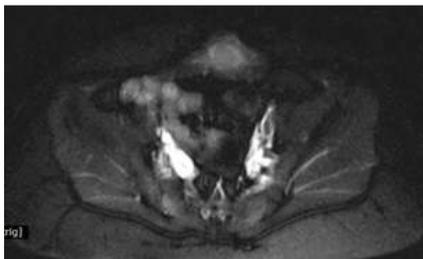


Figure 3c

Figure 3 MRI Axial T1 (a), T2 (b) and T2 HASTE FS(c)-shows well defined solid lesion in anterior abdominal wall just on the left side of midline in hypogastric region in subcutaneous plane nearly isointense to muscle on T1WI, appearing slightly hyperintense on T2WI and T2 HASTE FS Involving outer muscle fibres of left rectus abdominis with adjoining fat stranding.

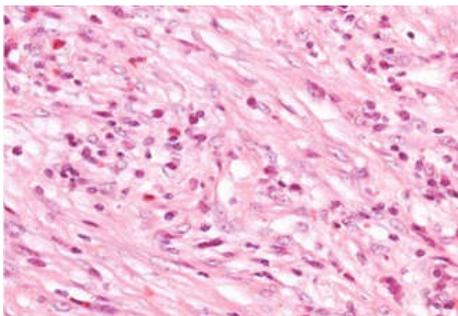


Figure 4 Photomicrograph (Haematoxylin and Eosin x 50) shows spindle cells and inflammatory cells suggestive of Inflammatory myofibroblastic tumor.

Introduction

Inflammatory myofibroblastic tumor (IMT) also called inflammatory pseudotumor. They are heterogeneous group of lesions characterised by variable fibroblastic reaction with inflammatory cell infiltration. These lesions generally show a favourable prognosis with rare incidence of malignant transformation and distant metastasis. They can affect head & neck (most commonly orbit, skull base and temporal bone), infratemporal fossa, nasopharynx, pterygopalatine fossa, soft tissues of neck, paranasal sinuses, cervical esophagus, upper respiratory tract, lung, pleura, heart, mediastinum, liver, spleen, kidneys, skin & soft tissue, tonsil, thyroid, larynx, trachea, ventricles, central nervous system and spinal cord meninges. It occurs in most organs and anatomic sites with few exceptions.¹ It is synonymous with inflammatory pseudotumor, plasma cell granuloma, pseudosarcomatous myofibroblastic lesion, inflammatory myofibrohistiocytic proliferation, histiocytoma, xanthoma, fibroxanthoma, xanthogranuloma, inflammatory fibrosarcoma (urinary bladder), plasma cell histiocytoma complex (lung), plasmacytoma. Various terminology used indicate uncertainty regarding true biologic nature of the lesion in view of recurrence, metastasis and cytogenetic evidence of acquired clonal chromosomal abnormality. The concept of this lesion being reactive is challenged.²

DISCUSSION

IMT was first observed in lung and described by Brunn in 1939 and was named by Umiker *et al* in 1954 as inflammatory pseudo tumor because of its property to mimic a malignant process both clinically and radiologically.³ Pettinato *et al* in 1990 studied 20 lesions of the lung and initially proposed and described the lesion as inflammatory myofibroblastic tumor.¹ It was later designated as post inflammatory tumor. According to World Health Organisation classification of soft tissue tumor, its definition was formalised in 1984 as a tumor composed of differentiated myofibroblastic spindle cells and or lymphocytes.¹ Its cause and pathogenesis is largely unknown. However reports of post traumatic, post surgical and post infections cases can lead to speculation that an initial reactive process can show overt neoplastic transformation. Lesions in liver, spleen and lymph nodes harbour the Epstein-Barr virus in spindle cells.¹ Other infectious agents associated are mycobacterium, klebsiella, pseudomonas, helicobacter pylori, HIV, Human Herpes virus-8, E.coli, bacillus sphaericus. An immune – autoimmune mechanism and infectious cause are also proposed.⁴ Actinomycosis and nocardia are found in hepatic and pulmonary pseudotumor respectively and mycoplasma in pulmonary pseudotumor. It is a mimic of neoplasia. Clinically patient may have varying degree of fever, thrombocytopenia and hypergammaglobulinemia.⁴

IMT in abdomen are rare. IMT can occur at any age though it is usually seen in child and young adults.⁵ Sites of abdominal IMT include liver, spleen, pancreas, mesentery, retro peritoneum, diaphragm, adrenal gland, urinary bladder, kidney, esophagus, stomach, small bowel, colon, appendix, Meckel's diverticulum.⁵ Small bowel, colonic mesentery, retro peritoneum, liver, spleen and other gastrointestinal sites show majority of cases.⁶ Only one pediatric case of IMT arising in abdominal wall has been reported. Only one adult case of IMT of abdominal wall has been reported.⁶ clinically its presence as

a palpable mass with mild to moderate pain, weight loss and fever.⁷

Diagnostic Workup

On USG, it may be well defined or have infiltrative margins. It is solid with heterogeneous echo texture and significant vascularity.⁸ On USG it appears as solid or cystic mass with well or ill defined borders with mixed echo texture. CT appearance of IMT is variable. It has well defined margins, moderate or marked homogeneous or heterogeneous enhancement is seen in solid part of tumor. Calcification, fatty component and non enhancement have been described.⁵ PET scan show high uptake of tracer in IMT. Hence IMT is difficult to differentiate from other neoplasm. MRI is useful to detect extent and involvement of underlying muscle. Laboratory investigations suggest an inflammatory process – leucocytosis, neutrophilia, anemia, thrombocytosis, elevated ESR and CRP, polyclonal hypergammaglobulinemia.

Histopathology

Macroscopically it presents as a solid, elastic, pink whitish mass with a characteristic fibroinflammatory appearance. Basic component of IMT are fibroblast and myofibroblast, lymphocytes, plasma cells and histiocytes in variable proportion. Histopathologically admixture of myofibroblastic spindle cells with inflammatory cell infiltrates of plasma cells, lymphocytes and eosinophils are seen. There is minimal mitotic activity and nuclear pleomorphism. Four basic histologic patterns are recognised-1. Dominant lymphoplasmacytic infiltrates 2. Dominant lymphohistiocytic infiltrates 3. Young and active myofibroblastic process, 4. Predominantly collagenised process with lymphocytic infiltrates. Collagenised IMT is less cellular and resemble desmoids tumor but shows prominent inflammatory infiltrates. The pattern of cellular or stromal elements and growth pattern may vary from one microscopic field to other in the same tumor and in relative proportion from one tumour to other tumor. Immunohistochemistry aids in its diagnosis. Vimentin is invariably positive in spindle cells. Majority of the cases show smooth muscle actin, muscle-specific actin and desmin. CD68 (KP-1), CD30 (Ki-1), cytokeratin and p53 are positive in some case.⁷

IMTs show variable biologic behaviour ranging from the frequently benign lesion to more aggressive variants. Aggressive behaviour can be predicted histologically by presence of cellular atypia, mitotic activity, nucleolar prominence, necrosis, ganglion-like cells, atypical mitotic figures expression of p53 and aneuploidy.⁶ IMT is a benign non-metastasising proliferation of my fibroblast with a potential for recurrence and persistent local growth similar to fibromatosis. Spontaneous regression has been reported in some case of IMT.⁶ Differential diagnosis are benign spindle cell lesion like leiomyoma, solitary fibrous tumor, spindle cell tumor, nodular fasciitis, peripheral nerve sheath tumor, lymphoma and metastatic disease.¹

Treatment

Primary therapeutic approach is surgery with complete resection of tumor if the anatomic location can be amenable. Inadequate resection can lead to recurrence.

Adjuvant approaches using corticosteroids, chemotherapy or COX-II inhibitors and radiation have been attempted in cases of incomplete resection with limited success.⁷ Radiotherapy and chemotherapy consisting of cisplatin, methotrexate and doxorubicin may be indicated.^{5,9}

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Conflict of interest: Nil.

CONCLUSION

IMT of abdominal wall are rare entities in adults. Clinical and radiological differentiation from other soft tissue tumor of abdominal wall is often difficult. Accurate multidisciplinary investigation (including clinical, radiological and histopathological examination) can guide exact diagnosis. Complete resection of tumor along with involved structures is key to successful treatment with reduced risk of recurrence. Long term clinical and radiological following is necessary in view of malignant transformation of IMT despite its benign histological features.

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