Research Paper Medical Science

Inflammatory Myofibroblastic Tumour of Large Intestine -- A Case Report

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ABSTRACT

Inflammatory myofibroblastic tumour(IMT) is a rare benign lesion in adults and children.It is a term used to describe a benign and rare process most commonly involving the lung but found in nearly every site in the body.3 A number of terms have been applied to the lesion, namely, inflammatory pseudotumour, fibrous xanthoma, plasma cell granuloma, pseudosarcoma, lymphoid hamartoma, myxoid hamartoma,inflammatory myofibrohistiocytic proliferation, benign myofibroblastoma, and most recently, inflammatory myofibroblastic tumour.The immunohistochemistry is usually utilized to confirm the myofibroblastic phenotype of the tumour spindle cells which are typically reactive to vimentin, SMA, muscle specific actin and desmin.1 We herein report a case of a 14 year old male presented with pain abdomen since two months and a mass in right iliac fossa. The patient underwent surgical excision of tumour and histology revealed that it was inflammatory myofibroblastic tumour which was confirmed by immunohistochemistry.

KEYWORDS IMT, Desmin, Vimentin

INTRODUCTION: Inflammatory myofibroblastic tumour (IMT) is a rare benign lesion in adults and children. It is an uncommon lesion of unknown cause.[3] It encompasses a spectrum of myofibroblastic proliferation along with varying amount of inflammatory infiltrate. The lungs, liver and gastrointestinal tract are the most common sites for inflammatory myofibroblastic tumour.

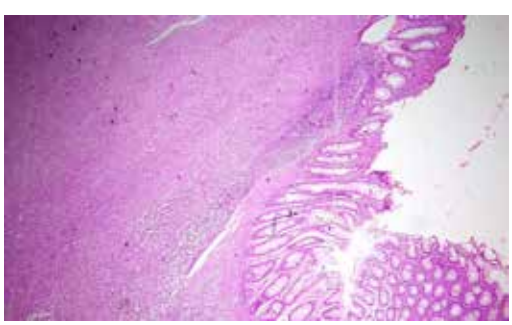
CASE REPORT: A fourteen year old male presented with pain abdomen since two months which was dull aching and gradual in onset and present throughout the day. There was no history of bleeding per rectum, vomiting . On abdominal examination, tenderness present in the right iliac fossa. An ill defined mass was palpable in right iliac fossa with smooth borders and ill defined margins.The tumour was surgically excised and sent for histopathological examination.

GROSS: specimen consisted of four pieces of grey brown tissue largest measuring 12x 8x 6 cm. External surface was bosselated with intestinal mucosa at two ends which was firm in consistency. Cut surface showed well circumscribed homogeneous grey white tumour arising from the mucosa. Another large piece of tissue measured 7x4x3cm. External surface showed part of intestinal mucosa with grey white areas arising from the mucosa. The other two pieces of tissue consisted of intestinal mucosa with fibrofatty tissue measuring 4x3x2 cm each. Five lymph nodes were dissected largest measuring 1x0.5x0.2cm.

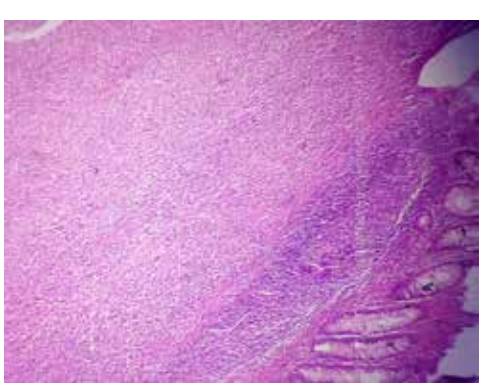
MICROSCOPY: Multiple sections studied showed large intestinal wall with one end showing submucosal lesion extending into serosa and also dissecting few smooth muscle bundles in the muscularis. The lesion consisted of proliferation of spindle cells with dense lymphoplasmacytic infiltration. These spindle cells had good amount of cytoplasm and a small to large nucleus. Some of these nuclei were vescicular with prominent nucleolus. Cells were arranged in fascicles and at places in whorls. Hyperplastic lymphoid follicles were also seen in the lesional tissue. Some areas of lesion showed dense hyalinised stroma with areas of calcification. No atypia in any of the cells seen.

IMPRESSION: Features were suggestive of Inflammatory myofibroblastic tumour.

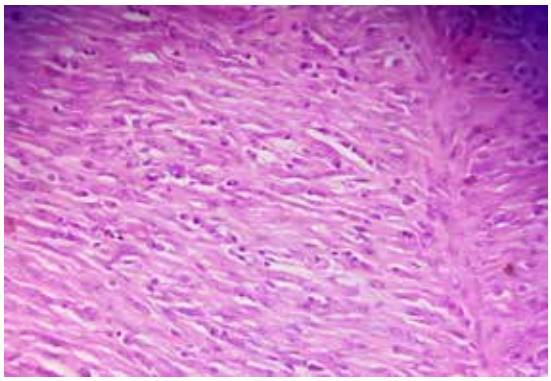
Immunohistochemistry was done which showed vimentin and ALK- 1 positive, Desmin and SMA showed focal positivity . The diagnosis of inflammatory myofibroblastic tumour was confirmed.



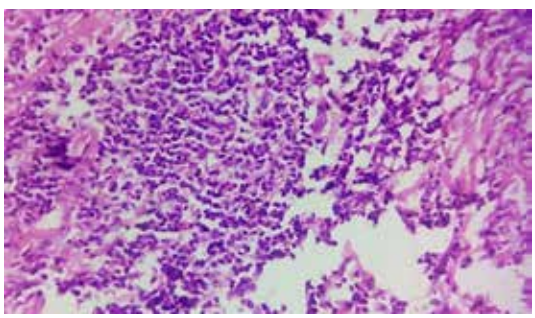
Large intestinal wall with submucosal lesion extending into serosa , X4x, Hand E



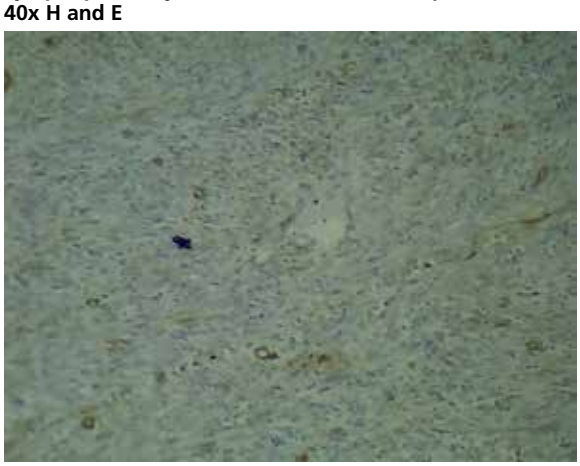
Spindle cells arranged in fascicles X 10x, H and E



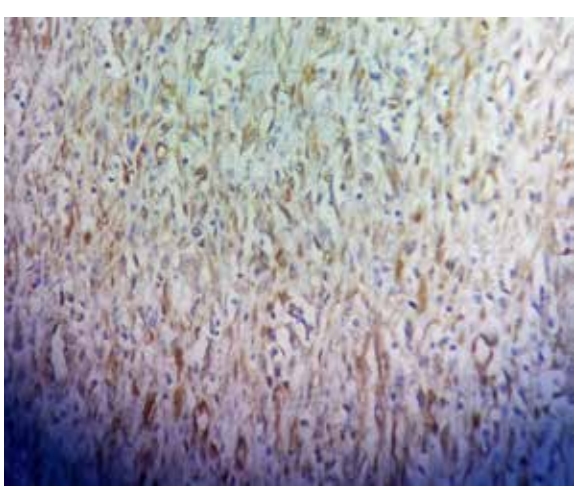
Spindle cells without atypia amidst plasma cells X40x H and E



Lymphoplasmacytic infiltration amidst spindle cells X 40x H and E



SMA positivity seen in spindle cells



Vimentin positivity in spindle cells

DISCUSSION: A number of terms have been applied to this lesion, namely inflammatory pseudotumour, fibrous xanthoma, plasma cell granuloma, pseudosarcoma, lymphoid hamartoma, myxoid hamartoma, myxoid hamartoma, inflammatory myofibrohistiocytic proliferation, benign myofibroblastoma and most recently, inflammatory myofibroblastic tumour. The diverse nomenclature is mostly descriptive and reflects the uncertainty regarding its true biologic nature of these lesions.[1]

Lymphocytes, plasma cells , histiocytes,fibroblasts and myofibroblasts are the basic components of inflammatory myofibroblastic tumour.

Recently, the concept of IMT being a benign reactive lesion has been challenged owing to clinical demonstration of recurrences as high as 37%, the presence of regional metastases and cytogenetic evidence of acquired clonal chromosomal abnormality. However, the issue of reactive versus neoplastic pathogenesis of this lesion remains unsolved.

It is suggested that inflammatory mediators such as cytokines and interleukin-1 are released in response to an insult causing proliferation of fibroblasts, leaky and pro-coagulant endothelium and extravasation of polymorphous cellular infiltrate into the extravascular spaces. Several triggers for its development have been described, including smoking, minor trauma, post-adenoidectomy and infection; however the exact etiology is not known.

The differential diagnosis of IMT comprises low-grade myofibroblastic sarcoma as well as a long list of benign, reactive or neoplastic spindle cell lesions, such as leiomyoma, solitary fibrous tumour, spindle cell carcinoma, nodular fasciitis and peripheral nerve sheath tumour.

In IMTs, spindle cells lack the cytologic atypia and nuclear hyperchromasia of sarcomas. The immunohistochemistry is usually utilized to confirm the myofibroblastic phenotype of the tumour spindle cells which are typically reactive to vimentin(99%), SMA(92%), muscle specific actin(89%) and desmin(69%). Spindle cells may be focally positive to epithelial markers such as cytokeratin, epithelial membrane antigen and CD 68. IMTs are typically negative for myoglobin and S100 protein.[1]

CONCLUSION: Inflammatory myofibroblastic tumour can occur throughout the body and should be considered preoperatively in the differential diagnosis. Surgery is generally required for diagnosis of IMT, but awareness of these lesions may prevent unnecessary radical surgery in some instances. In general IMT follows a benign course with favourable outcome after local excision.[2]

REFERENCES

1.Sangeeta Palaskar, Supriya Koshti, Mahesh Maralingannavar et.al, Inflammatory myofibroblastic tumour- contemporary clinical dentistry: Oct-Dec2011.Vol 2,issue 4. | 2.Madhavi Patnana, Alexander B. Sevrukov,Khaled M.Elsayes et.al, Inflammatory Pseudotumour: The great Mimicker,AJR: 198,March 2012. | 3.Avinash N Katara, Vinod A Chandiramani, Faram D Dastur, Ramesh B Deshpande, Inflammatory pseudotumour of ascending colon: A case report, Indian Journal of surgery. | 4.Kenneth A Iczkowski, et.al,Inflammatory Pseudotumour and Sarcoma of Urinary bladder:Differential diagnosis and outcome in thirty eight spindle cell neoplasms, Mod Pathol 2001;14(10):1043-1051. | 5.Nilufer Onak Kandemir et.al, Intranodal palisaded myofibroblastoma: A case report and literature review: Diagnostic pathology 2010,5:12 |