ORIGINAL RESEARCH PAPER | Pathology | Volume : 6 | Issue : 12 | December 2016 | ISSN - 2249-555X | IF : 3.919 | IC Value : 79.96

Inflammatory Myofibroblastic Tumor of Caecum Presenting As Acute Appendicitis-A Rare Case With Rare Presentation With Immunohistochemical Study

KEYWORDS

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Inflammatory myofibroblastic tumors (IMT) also known as inflammatory pseudotumor is a histologically benign entity that primarily occurs in the viscera like lung, orbit, mesentery and omentum. (1) We have encountered a rare case of intraabdominal caecal IMT masquerading as acute appendicitis.

A 45 year old female presented with right iliac fossa pain. There was history of fever, nausea and vomiting. Abdominal x ray showed few dilated loops of ileum. Ultrasonography showed mass lesion with thickening of wall of caecum and dilated and elongated retrocaecal appendix. Clinically diagnosis of acute appendicitis was made.

On laparotomy, appendix was swollen and adherent to caecal mass. Right hemicolectomy was performed to excise the mass. On two year follow up, patient is in good health.

Gross examination revealed an ill circumscribed firm to hard caecal mass of size 6 x 4x 2 cm, grayish white to yellow in colour. The appendix was elongated and dilated, measuring 10 cm in length .(Figure 1A)

Microscopic examination showed lesion extending from subepithelium to serosa. It showed elongated cells with spindle shaped nuclei arranged in haphazard bundles and short fascicles with collagenous background. Abnormal mitoses and nuclear pleomorphism was absent. The stroma showed variable amount of infiltration by lymphocytes forming focal dense collections, plasma cells and few eosinophils.(Figure 1B) Section through appendix showed thickened muscular wall and lymphoplasmacytic infiltrate. Immunohistochemical studies showed strong positivity with smooth muscle actin (SMA) and desmin respectively.(Figure 1C,1D) CD117 was negative and CD3 and CD20 was focally positive.

Inflammatory pseudotumor of the caecum is an uncommon mesenchymal neoplasm with a variable histologic appearance.(2)They are often confused with malignancy clinically. The incidence of inflammatory pseudotumor is higher in younger patients, both genders being equally affected.(3)

It has been postulated that IMTs are associated with a variety of infectious agents (4). However in our case there was no past history of inflammation. Few studies suggest a potential for low grade neoplasia.(5).They also have tendency to recur. IMTs are often confused with highly malignant neoplasms clinically. Laboratory investigations are of little help. Radiological appearance of these tumors is variable often presenting with varying degrees of enhancement, areas of calcifications. Histopathology of tissue confirms the diagnosis.

Complete excision or right hemicolectomy is ideal treatment. We present this case to emphasize that this entity should always be kept in mind when a well circumscribed lesion of caecum is encountered at laparotomy.

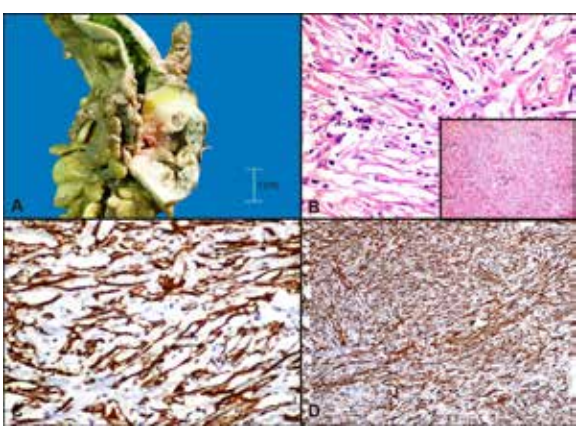

Figure Legends

Figure 1A- Gross Specimen showing an ill circumscribed firm to hard caecal mass of size 6 x 4x 2 cm, grayish white to yellow in colour with elongated and dilated appendix.

Figure 1B- Photomicrograph(40 X) showing elongated cells with spindle shaped nuclei arranged in haphazard bundles and short fascicles and infiltration by lymphocytes forming focal dense collections, plasma cells and few eosinophils. Inset-(10 X)

Figure 1C- (40 X) Immunohistochemical stain SMA(Smooth Muscle Actin) showing strong positivity.

Figure 1D- (40 X) Immunohistochemical stain Desmin showing strong positivity.

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