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APPENDICEAL INFLAMMATORY MYOFIBROBLASTIC TUMOUR: A RARE POSTOPERATIVE FINDING

General Surgery

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ABSTRACT

Introduction- Inflammatory myofibroblastic tumour (IMT), which usually affects children and young adults, is a rare neoplastic lesion of unknown aetiology.[1–5]Once thought to be reactive, now these lesions are considered to be neoplastic.[2] Definitive diagnosis of IMT is confirmed by histopathological analysis and immunohistochemical tests of surgically removed specimen.[1–5] According to the World Health Organization, IMT is a distinctive lesion composed of myofibroblastic spindle cells accompanied by an inflammatory infiltrate of plasma cells, lymphocytes, and eosinophils.[1–6] Inflammatory myofibroblastic tumour is a relatively new histopathological term. In the literature, this entity was formerly known as inflammatory pseudotumor, plasma cell granuloma, plasma cell pseudo-tumour, pseudo-sarcomatous myofibroblastic proliferation, reactive pseudo-sarcomatous response, omental-mesenteric myxoid hamartoma, and inflammatory fibrosarcoma. This unification of descriptive terms was achieved on the basis of considerable morphological and clinical overlap, combined with both clinical and genetic evidence of its neoplastic nature.[2,3,5,7] IMT was first described in 1937 as a primary lung tumour [1,2,3,5,7] but it was subsequently recognized that virtually any anatomical site can be involved.[1,4,6,7] Most of the extrapulmonary IMTs are localized in the mesentery of the small intestine, omentum, and retroperitoneum.[1,4] To this date, IMTs of the gastrointestinal tract are rare.[1,3,8] Most of these reported cases are adolescent or adult males.[2,3] **Case Report-** A 13 years old male patient presented to surgery casualty with complaints of pain in abdomen vomiting for 4 days on clinical examination patient have tachycardia and tenderness in right iliac fossa. Initial USG A+P was suggestive of perforated appendix with early lump formation. Patient was managed conservatively and discharged after 3 days. But after 2 weeks patient again presented with similar complaints and progressively increasing lump in right lower abdomen. Lump was 15x8cm in right iliac fossa extending to hypogastric and right lumbar region firm to hard in consistency with mild tenderness on deep palpation. Ultrasound of abdomen suggestive of organised haematoma. CECT abdomen and pelvis suggestive of inflamed appendix with suspicious perforation and well-defined heterogenous collection of max volume 340cc could represent organised haematoma. Patient was posted for elective exploratory laparotomy with intra op findings of 1) mass of size 15x10x8cm located in right lower quadrant 2) whole specimen weighed 350gms 3) appendix showed diffuse inflammation and adherent to mass. Appendicectomy along with excision of mass was done and sent for histopathology. HPE Report-Benign spindle cell lesion highly suggestive of inflammatory myofibroblastic tumour. **Conclusion-** IMT is a rare lesion that can represent an unexpected intra-operative finding at emergency surgery. Generally, IMTs have a good outcome but they have a tendency for local recurrence and small risk of distant metastasis. IMT can be observed in appendices even though it is rare. Complete surgical resection should be the method for curative treatment. Definitive diagnosis of IMT is confirmed by histopathological analysis and immunohistochemical tests of surgically removed specimen.

KEYWORDS

INTRODUCTION:

Inflammatory myofibroblastic tumour (IMT), which usually affects children and young adults, is a rare neoplastic lesion of unknown aetiology.[1–5] Once thought to be reactive, now these lesions are considered to be neoplastic.[2] Definitive diagnosis of IMT is confirmed by histopathological analysis and immunohistochemical tests of surgically removed specimen.[1–5] According to the World Health Organization, IMT is a distinctive lesion composed of myofibroblastic spindle cells accompanied by an inflammatory infiltrate of plasma cells, lymphocytes and eosinophils.[1–6] Inflammatory myofibroblastic tumour is a relatively new histopathological term. In the literature, this entity was formerly known as inflammatory pseudotumor, plasma cell granuloma, plasma cell pseudo-tumour, pseudo-sarcomatous myofibroblastic proliferation, reactive pseudo-sarcomatous response, omental-mesenteric myxoid hamartoma, and inflammatory fibrosarcoma. This unification of descriptive terms was achieved on the basis of considerable morphological and clinical overlap, combined with both clinical and genetic evidence of its neoplastic nature.[2,3,5,7]

IMT was first described in 1937 as a primary lung tumour [1,2,3,5,7] but it was subsequently recognized that virtually any anatomical site can be involved.[1,4,6,7] Most of the extrapulmonary IMTs are localized in the mesentery of the small intestine, omentum, and retroperitoneum.[1,4] To this date, IMTs of the gastrointestinal tract are rare.[1,3,8] Most of these reported cases are adolescent or adult males.[2,3]

Case Report:

A 13 years old male patient presented to surgery casualty with complaints of pain in abdomen vomiting for 4 days on clinical examination patient have tachycardia and tenderness in right iliac fossa. Initial USG A+P was suggestive of perforated appendix with early lump formation. Patient was managed conservatively and discharged after 3 days. But after 2 weeks patient again presented with similar complaints and progressively increasing lump in right lower abdomen. Lump was 15x8cm in right iliac fossa extending to hypogastric and right lumbar region firm to hard in consistency with mild tenderness on deep palpation. Ultrasound of abdomen suggestive of organised haematoma. CECT abdomen and pelvis suggestive of inflamed appendix with suspicious perforation and well-defined heterogenous collection of max volume 340cc could represent organised haematoma. Patient was posted for elective exploratory laparotomy with intra op findings of 1) nmass of size 15x10x8cm located in right lower quadrant 2) whole specimen weighed 350gms 3) appendix showed diffuse inflammation and adherent to mass. Appendicectomy along with excision of mass was done and sent for histopathology. HPE Report -Benign spindle cell lesion highly suggestive of inflammatory myofibroblastic tumour.

Patient was postoperatively stable, and he was discharged on postoperative day 15 after removal of all sutures and with reports. Now one year after his initial diagnosis, this gentleman continues to be monitored for local and distant recurrence of disease but patient stable with no history any recurrence.

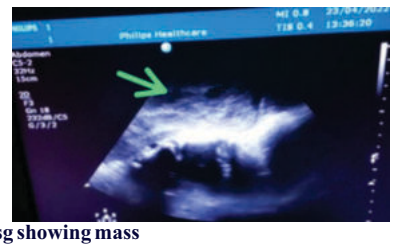

Figure 1: usg showing mass

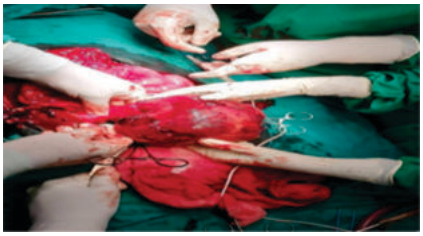

Figure 2: Intraoperative image

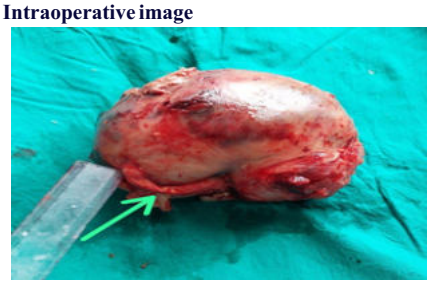

Figure 3: Specimen with adherent appendix

DISCUSSION:

IMT is a rare inflammatory pathological entity that most affects children and young adults.[1–3] IMTs can be pulmonary and extrapulmonary. Extrapulmonary IMTs are most frequent in the abdomen including the mesentery and omentum.[2,3] IMT is still referred to as a lesion of unknown etiopathogenesis.[1–4] It may arise due to an exaggerated or aberrant tissue response to chronic inflammatory process occurring after infection, trauma or surgery.[1,3,4] Studies have shown Coxiella burnetti, Klebsiella pneumoniae, Epstein–Barr virus, and Campylobacter jejuni as possible infectious agents.[1,2,7] Association with Castleman's disease and Hodgkin's disease was suggested.Microscopic sections show well circumscribed tumour with tumour cells arranged in storiform pattern,vaguely nodular, The tumour cells are spindle shaped the nuclei are elongated with oval to spindle shaped .Stroma is loose and myxoid with congested numerous blood vessels with inflammatory infiltrates of lymphocytes, plasma cells ,neutrophils and few mast cells, Appendix is been adherent to the tumour mass and is congested.In our case symptoms related to acute appendicitis had been present 2 weeks before that acute inflammation may be the etiological factor for IMT in this case. The clinical presentation of IMT varies markedly, depending on the site at which the tumors originated.[1–4] Hence appendiceal IMT may present with features of acute appendicitis.[1,3] Patients usually present with non-specific, vague symptoms such as abdominal pain, nausea, and vomiting. Because of the lack of specific clinical or imaging signs, IMT still offers a great deal of diagnostic challenge. Thus, it is very difficult to diagnose these tumors pre-operatively.[1–3,6–8] Complete surgical resection is the method for curative treatment even in those with multiple recurrences.[1,2,4–6] An appendectomy is adequate in those cases restricted to the appendix. As such, the definitive diagnosis is possible via histopathological assessment.[1,3–6] IMTs are essentially cellular, fascicular fibroblastic, or myofibroblastic proliferations accompanied by a prominent infiltrate of chronic inflammatory cells, particularly plasma cells. The spindle cell component typically has plump nuclei, and the mitotic rate is variable. IMT has the similar immuno histochemical phenotype as other myofibroblastic tumors. They show positivity for vimentin, smooth muscle actin, and/or muscle-specific actin in the majority of the cases and may also show staining for desmin.The neoplastic nature of IMT was first suggested in 1991 by Meis et al.[2,8,9] IMTs are among the increasing number of diseases in which aberrant anaplastic lymphoma kinase gene (ALK) has been implicated in their pathogenesis.[1,7–9] Tsuzuki et al. demonstrated ALK gene translocation in IMT, which is not present in other spindle cell tumors and demonstrated that ALK-1 expression is highly specific for IMT and confirmed its neoplastic nature by fluorescence in situ hybridization (FISH).[9] Recent studies reported that 36–50% of IMTs are positive for ALK and show up to 67% ALK rearrangements by FISH.[2,6] ALK expression is more prevalent in younger patients, but is not restricted to this population.[2,9]

The clinical course is usually benign, but local recurrence following complete excision can occur in 2–25%. Risk of <5% for distant metastasis and the 5-year survival is about 87%.[2,4,7] Metastatic disease is usually identified at presentation or within a year of diagnosis, but occasional patients develop metastases as many as 9 years following resection. The most common sites of metastasis are lung and brain, followed by liver and bone.[7] There have been cases reported in the literature where spontaneous regression has been described and good results have been reported with steroid treatment among recurrent cases.[6] Long-term follow- up is highly recommended to detect local recurrence or possible metastasis.[4–6]

CONCLUSION:

IMT is a rare lesion that can represent an unexpected intra-operative finding at emergency surgery. Generally, IMTs have a good outcome but they have a tendency for local recurrence and small risk of distant metastasis. IMT can be observed in appendices even though it is rare. Complete surgical resection should be the method for curative treatment.Definitive diagnosis of IMT is confirmed by histo pathological analysis and immunohistochemical tests of surgically removed specimen. In this study, we aimed to examine the literature as well as to describe a case that we operated on that presented as an appendiceal mass.Upon histopathological examination, the lesion was identified as an IMT.

Awareness of this kind of tumor in the differential diagnosis of appendiceal masses avoids extensive surgical resection and subsequent morbidity and highlights the need for a long-term clinical and radiological follow-up because of relatively high frequency of local recurrence (up to 25%) and the possibility to metastasize (<5%).

Disclosure:

No portion of the manuscript, including images,contains patient-identifiable information.

Conflict of Interests:

The authors declare that there is no conflict of interests regarding the publication of this paper.

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