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Inflammatory myofibroblastic tumour of abdomen: A case report

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Abstract

Inflammatory myofibroblastic tumours (IMTs) constitute a rare group of neoplasms composed of a mixture of spindle-shaped myofibroblasts or fibroblasts and a variable amount of inflammatory cells. We reported a case of Inflammatory myofibroblastic tumor in 12 year old female patient.

Keywords: Inflammatory myofibroblastic tumors, Neoplasms, Cell

1. Introduction

Inflammatory myofibroblastic tumours (IMTs) constitute a rare group of neoplasms composed of a mixture of spindle-shaped myofibroblasts or fibroblasts and a variable amount of inflammatory cells (eosinophils, plasma cells and lymphocytes). [1] Many different terms have been used to refer to these tumours: plasma cell granuloma, inflammatory myofibrohistiocytic proliferation, fibroxanthoma, xanthogranuloma. However, nowadays IMT is the most accepted. The most frequently affected organs are lung and orbit, but they have been described in nearly every organ. [2]

Different etiologies have been proposed for IMT, being different chronic infections, autoimmune diseases and trauma the most accepted. Specific inflammatory diseases, such as IgG4 disease, have also been recently associated. Little information exists about the natural history of this entity. In some cases, an aggressive behaviour with metastases has been described. [3]

Since IMTs can arise from various anatomic locations, they concern almost every subspecialty in surgical oncology. The management of these tumours can be challenging because there are no established medical treatment protocols, and tumours can be irresectable owing to their proximity to vital structures. [4] The present article highlighted a case report of IMT in 12 year old female patient.

2. Case Report

A 12 yrs old female presented to the surgery OPD with lump lower abdomen for 3 months, extending above the umbilicus. Patient also complained of pain in lower abdomen which was sudden in onset, intermittent, colicky type, aggravated during bowel movements & relieved with analgesics & after passing stools. There was no H/O malena, hematemesis, hematochezia, nausea, vomiting and anorexia. Patient had not yet attained menarche. The lump was initially of the size of cricket ball which gradually increased to attain the present size. There was no significant past medical/surgical history & no significant family history.

On Examination, patient was thin built, pallor was present. There was no jaundice, edema, cyanosis and lymphadenopathy present. All vitals were within normal limits. On local examination, whole abdomen was distended especially lower abdomen. There were no dilated veins, no visible pulsations, no previous scar mark was present, umbilicus was downwardly placed with vertical slit.

There was large lump measuring 12X10cm which was intraperitoneal, extending above the umbilical region & below upto the hypogastrium, with rounded margins, smooth surface, firm & uniform in consistency, mobile in both cranio-caudal & side to side direction, mildly tender, dull on percussion (Fig-1). On palpation no pulsations were felt. Per vaginal examination was not done as patient was unmarried. On per rectal examination, sphincter tone was normal & no signs

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of hemorrhoids, rectal mucosa were mobile & normal, no growth was present, no fecalith were present. Pouch of Douglas was full, cervix was felt & was normal, no growth felt around the cervix, uterus was not felt.

All hematological and biochemical investigations were within normal limits. CA 125 were within normal limits. Ultrasonography revealed a well defined mildly heterogeneous mass of size- 10.6*3.6 cm is seen in lower abdomen.

MRI whole abdomen (Plain) revealed a large lobulated soft tissue signal intensity mass lesion in the lower abdomen & pelvis with associated moderate ascites. The differential possibility include small bowel Gastrointestinal stromal tumor (GIST), mesenteric lesion like Desmoid tumor, GIST, mesenchymal neoplasm (leiomyoma, neurofibroma, leiomyosarcoma), exophytic colonic adenocarcinoma, liposarcoma, dermoid tumor, ectopic spleen, ovarian tumor and lymphoma. Histopathology was suggestive of inflammatory myofibroblastic tumour (Fig- 2).

Patient was managed on IV fluids & IV antibiotics & kept nil per orally for 5 days. 2 units of fresh whole blood was transfused & 2 units of 20% albumin was also given in the subsequent post operative period. Patient was given liquids & semisolids from 6th post- operative day & she recovered completely. Whole postoperative period was uneventful. Patient was discharged from the hospital on 8th post- operative day.

3. Discussion

IMTs have a predilection for children and adolescents, although they may arise as late as the eighth decade of life. The most common anatomical locations are the abdominopelvic region, lung, and retroperitoneum, but virtually any site may be involved, including the somatic soft tissues, bone, larynx, uterus and central nervous system. Accurate data regarding the incidence and anatomical distribution of IMT are difficult to obtain due to the use of the terms “inflammatory pseudotumor” and “IMT” interchangeably in the literature.^[5]

Clinical presentation of IMTs depends on the organ in which they arise, but they frequently associate general inflammatory symptoms as fever or malaise. Patients generally present with a mass or nonspecific symptoms, including vague abdominal pain or gastrointestinal complaints for intra-abdominal lesions, and cough, chest pain, or, less often, haemoptysis for pulmonary tumours. A constitutional syndrome consisting of fever, weight loss and malaise is seen in 15–30% of patients, and laboratory evaluation may reveal microcytic anemia, a raised erythrocyte sedimentation rate, thrombocytosis, and/or polyclonal hypergammaglobulinemia. In some cases, the mass may be found only after an extensive workup for fever of unknown origin or growth failure.^[6]

Radiological appearance of IMTs is unspecific and they are often misdiagnosed as malignant neoplasms. Many of them are incidentally discovered when an imaging technique (computed tomography [CT], ultrasonography [US] or magnetic resonance imaging [MRI]) is performed for any other reason. Their most common radiological presentation is as solid, irregular, well-defined masses.^[7]

Histological studies are critical to properly diagnose them. Immunohistochemical studies of T- and B-cell subpopulations, CD34, CD117, S-100 and c-Kit may be helpful in distinguishing IMTs from other soft-tissue neoplasms. At the molecular level, approximately half of IMTs contain a clonal cytogenetic aberration that activates the anaplastic lymphoma kinase (ALK). Positive immunohistochemical staining of ALK is in approximately 40–100% of IMTs, depending on the anatomical

sites where they arise. This finding suggests a possible neoplastic cause of IMTs. Furthermore, ALK expression could be a prognostic factor of aggressiveness for IMT. According to an update based on the new World Health Organization (WHO) classification, IMTs are considered as neoplasms which may recur or metastasize in as many as 5% of cases.^[8]

Most of these tumours are discovered incidentally during radiological studies. There are no established decisive criteria for differential diagnosis. The current histopathological definition of an IMT is a distinctive neoplasm composed of myofibroblastic mesenchymal spindle cells accompanied by an inflammatory infiltrate of plasma cells. Nevertheless, tissue samples obtained by computed tomography-guided fine-needle or tru-cut biopsies, and even analysis of perioperative biopsies, are occasionally not enough to establish a diagnosis and there is no specific immunohistochemical staining for IMTs. As a result, the pathologist usually asks for the whole specimen.^[9]

Currently, surgery is the mainstay of the treatment for IMTs. Complete removal of the tumour generally provides resolution of all symptoms and laboratory abnormalities. However, tumours in intra- or retroperitoneal locations tend to invade adjacent structures, preventing curative resections and breeding local recurrences. These patients require further management. Unfortunately, chemotherapy and radiotherapy are not successful in most patients.^[10] Recently, researchers have published promising results with anti-inflammatory agents and anti-tumour necrosis factor- α binding antibodies.

Fig 1: Abdominal lump

Fig 2: Histopathological photograph

4. Conclusion

Although IMTs in some organs are not uncommon, they are not usually included in the differential diagnosis of masses. Their radiological features suggest malignant neoplasms, whereas they are not.

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