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Case Report

INFLAMMATORY MYOFIBROBLASTIC TUMOUR IN ANTIMESENTERIC BORDER OF DESCENDING COLON OF CHILDREN: A CASE REPORT

ISLAM S[1], MORSHED AKMA[2], ISLAM A[3]

Abstract:

Inflammatory myofibroblastic tumour (IMT) occurring at intraabdminal sites in children has rarely been described. Inflammatory pseudotumour is a soft tissue lesion that may be confused with a sarcoma. It is abbreviated as IMT. Inflammatory myofibroblastic tumour, also known as soft tissue tumours ,atypical fibromyxoid tumours, pseudosarcomatous fibromyxoid tumour ,plasma cell granuloma, pseudosarcomatous myofibrotic proliferation, post-operative spindle cell nodules . In this paper, we describe a case of inflammatory myofibroblastic tumour (IMT) with an unusual constellation of clinical, pathological findings. A 10-year- old girl had an 7-cm intraabdominal mass accompanied by severe anemia, fever, constipation, weight loss, thrombocytosis, elevated erythrocyte sedimentation rate. Laparotomy was performed. The final pathologic diagnosis was IMT. At the most recent follow up (12months) after excision of the tumour, the patient was symptom-free and there was no evidence of tumour recurrence.

Key words: Inflammatory myofibroblastic tumour, intra abdominal mass, severe anaemia, children

Introduction:

Inflammatory myofibroblastic tumour is a rare proliferative lesions clinically resembling a malignant neoplasm. Their classification is controversial and confusing. Inflammatory myofibroblastic tumours (IMT) are well described in the lung and upper respiratory tract of young adults and children; but may occur at any age and affect any organ system[1,2,3].

Typically, it is a circumscribed but non-encapsulated lesion containing spindle cells proliferating in a background of fibrosis, with lymphocytes, plasmacytes, histiocytes, foamy macrophages, and occasionally eosinophils and neutrophils. Nuclear pleomorphism and atypical mitosis are absent [4,5]. The histologic differential diagnosis includes calcifying fibrous pseudotumour, inflammatory fibroid tumour and nodular fasciitis. In IMTs, high cellularity with large, plump, active myofibroblasts with prominent nucleoli can cause confusion with malignancy, in particular rhabdomyosarcoma. Howeve the lack of atypia, hyperchromasia and abdominal mitotic figures are pointers toward a benign lesion. IMT should be diagnosed by routine staining because special stains and immunocytochemistry can be misleading. Preliminary biopsy and full histological evaluation is recommended in cases where resection may be particularly hazardous [5,6,7.] Matsubara et al. used the term inflammatory pseudotumour and described three subgroups based on the cell type most encountered in a mass: organizing pneumonia(44%) and lymphoplasmocytic type(12%) [8]. Some studies however suggest that it might be a true neoplasm as some mutations on chromosome 2p23 of anaplstic lymphoma kinase are found to be related to this tumour[9].

Chemotherapy, radiation treatment, nonsteroidal anti-inflammatory drug (NSAID), steroid and cyclosporin-A have been used as treatment modalities, but surgical resection is considered as treatment of choice[10,11].

1. Dr. Shahnoor Islam, Assistant Professor, Department of Pediatric Surgery, Dhaka Medical College Hospital
2. Dr. A. K. M. Amirul Morshed, Associate Professor, Department of Pediatric Hematology and Oncology, Dhaka Medical College Hospital
3. Professor Afiqul Islam, Chairman, Department of Pediatric Hematology and Oncology, BSMMU, Shah-bag, Dhaka, Bangladesh

Correspondence to : shahnoor6989@yahoo.com

According to Kovach *et al* [10], chemotherapy has been reserved for patients for whom resection is neither complete not possible. Radiation treatment showed some benefit in pulmonary IMT[11]. Radiation therapy is used in palliative treatment of this tumor, decreasing the mass effect of IMT and in patients whose tumors are not resectable [10]. The use of steroids is also recommended to reduce the inflammatory process that is surrounding the tumor, especially if the tumor is in the central nervous system [10,11]. Using nonsteroidal anti-inflammatory drugs as a conservative measure for treatment of patients with IMT whose tumors are not amenable to surgical resection[11].

It rarely presents recurrence, metastasis or malignant transformation, and is classified as intermediate neoplasm in the World Health Organization histological typing, but it may recur locally or manifest systemic symptoms. It is for this reason that regular follow-up is necessary even though surgical resection was done[12]. Karnak *et al* reported a case with very high leukocyte count and suggest that this could be a good marker for recurrence[13]. About 18% to 40% of IMTs recur, and most recurrences appear in extrapulmonary lesions that are larger than 8 cm and are locally invasive [14]. Retroperitoneal and mesenteric IMT seem to be associated with more frequent recurrences. Complete surgical resection is the treatment of choice and should be advocated unless prohibited [11].

Case report:

A previously healthy 1o-year old girl presented with a 1-month history of progressive lethargy, pallor, anorexia, intermittent fever, constipation, profound weight loss, left lower abdominal mass . She was found to have severe anaemia, thrombocytosis, elevated erythrocyte sedimentation rate, intraabdominal mass and was referred from pediatric haemato-oncologist to us for excision biopsy. At the start of her hospital stay, she continued to spike fevers upto 103^{o}F in the evenings, while her temperature remained normal throughout the day. Abdominal examination revealed a large, firm, mobile, abdominal mass below the umbilicus just left of midline. Haemogram revealed microcytic hypochromic anemia (haemo globin level 5 g/dl) and thrombocytosis (platelet counts 840x1o^{9}/l), increased ESR(115mm in first hour-Westergren), eosinophilia(08%), without neutrophilia. Lactate dehydrogenate (358u/l), alpha-feto-protein (0.67ng/ml), beta-human chorionic gonadotropin (<1.00mlu/ml) were normal. Widal test was negative. Ultrasonography demonstrated one large irregular outline mass measuring about 7.2x6.2cm was present at left iliac region, extending into left lumbar region, below the anterior abdominal wall . No hepatosplenomegaly, pelvic or para-aortic lymphadenopathy, abnormal collection or any other abnormality was seen. A provitional diagnosis of intraabdominal mass was given on the basis of ultrasonographic findings. Chest skiagram was normal. Ultrasono-guided fine needle aspiration cytology(FNAC) from lower abdominal mass yielded scanty blood mixed material. Microscopic smears show hypocellular material composed of few round oval cells, spindle cells and blood. No malignant cell was seen. After a short period of investigations, a laparotomy was performed for excision biopsy. Four units of blood were transfused preoperatively and postoperatively. A long pedunculated (5cm) , encapsulated, multilobulated mass measuring about 8x7x3cm was found arising from the antimesenteric border of the descending colon without local adhesion. The tumour was excised completely without any complication. After surgery, the child's clinical condition improved, laboratory findings returned to normal, and her fever resolved. She discharged on seventh postoperative day after her surgery. It is currently twelve months of follw-up after surgery without evidence of recurrence.

Macroscopically, specimen consisted of a nodular piece of tissue measuring 8x7x3cm. Cut surface was gray- white and solid. Microscopically specimen showed that a tumour composed of spindled myofibroblast, fibroblasts and inflammatory cells including eosinophils, plasma cells and histiocytes. These spindle cells have vesicular nuclei and abundant cytoplasm. No cellular atypia, hypercromatism was seen. A final diagnosis was given on the basis of histological findings. These findings were compatible with an IMT.

Discussion:

IMT has been reported at various sites such as mesentery, stomach, small intestine, large intestine, liver, mediastinum, retroperitoneum and bladder . The most common site of IMTs is the lung and the most common sites of extrapulmonary inflammatory myofibroblastic tumor are the mesentery and omentum. Among extrapulmonary IMT, 43% arose in

the mesentery and omentum. Although it occurs primarily in children and young adults (mean age, approximately 10 years), in more recent years a broad age range has been documented . There is no difference in incidence between females and males, though each report has a difference in mean age and gender ratio[11]. In a review of 44 confirmed diagonsed cases, only 3 cases occurred in colon [13].

Until 1992, a total of 21 children predomi- nantly female, with IMTs affecting intraabdominal sites have been reported in the literature. These tumours are often large. Multicentric lesions are rare. There are no reports of malignancy arising in an IMT; nevertheless, the clinical, radiological and histological features of these may cause confusion with malignant lesions[2,4]. Our case also was 10 yrs old female with intrabdominal mass. Most patient presented with fever , severe anemia, thrombocytosis, hyperglobulinemia and weight loss in previous case reports. These systemic features resolve after tumour excision[2,4]. These were also features of our patient. Misdiagnosis has led some patients to be inappropriately treated with chemotherapy and radical surgery. It had not happened in this case.

Other reasons for confusion of IMTs with malignant neoplasms include their capacity for local tissue infiltration, occasional rapid growth and the development of local recurrence. Mediastinal and esophageal involvement from adjacent pulmonary pathology appears to be particularly aggressive. Their course is complicated, which ranges from spontaneous regression through gradual enlargement to rapid growth with local invasion.

Conclusion:

IMT is a benign neoplasm rarely presented with malignant features such as local invasiveness, recurrence, distant metastasis, or malignant transformation. IMT can be suspected preoperatively through some hematologic abnormalities and radiologic findings, but precise diagnosis should be made on the basis of histologic findings. Complete surgical resection and close follow-up are all necessary for appropriate treatment to avoid recurrences as well as unnecessary and potentially harmful therapy. The optimal management of locally aggressive and recurrent forms should be decided individually for each patient. This underlines the importance of complete surgical resection and long-term follow –up.

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