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Omental – Fibrous Myxoid Hamartoma a Rare Lesion Presenting as Malignancy in a 3 Yrs Old Male Child (a Case Report)

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Abstract

Background: The Omental fibrous hamartoma is a very rare lesion, mainly seen in children characterised by single/multiple, Omental/mesenteric large solid mass, which may be confused with malignant neoplasm. Microscopically these are composed of fibroblast in bundles separated by myxomatous stroma. Lesion has benign clinical course without recurrence.

Case Presentation: We present a 3 yrs old male child with history of complaints of abdominal lump since 2 yrs, which was increasing progressively in size over period of time. No history of nausea, vomiting, fever etc. Contrast enhanced computed tomography (CECT) revealed large well-demarcated heterogeneously hyperdense lesion in right hypochondriac, right lumbar and umbilical region displacing bowel loops on either side without penetration & showing minimal enhancement on contrast study. Histological examination of entire mass confirmed the diagnosis of OMENTAL FIBROUS MYXOID HAMARTOMA. No evidence of recurrence noted during follow up period of 2 yrs.

Conclusion: Fibrous hamartoma of omentum is a very rare lesion and its aggressive appearance needs to differentiate from malignancy. The clinical picture of our case also led to high suspicion of malignancy. However by consideration of histological findings we could achieve the correct diagnosis.

Keywords: Omental–Fibrous Myxoid Hamartoma; Omentum; Abdominal mass; Childhood.

Introduction

Omental-mesenteric myxoid hamartoma (OMMH) is the entity initially described by Gonzalez-Crussi *et al* in 1983. There work described three infants who presented with multiple nodular tumours of the omentum and mesentery characterized Histologically by plump mesenchymal cells in well vascularised Myxoid stroma. Electron microscopy of one tumor revealed reticulated inclusions in dilated cisterna of endoplasmic reticulum. All of these patients survived with no evidence of recurrence following excision.[1] OMMH shares the morphologic features with inflammatory myofibroblastic tumor (IMT) and, in other words it may represent a variant of IMT.[2] These lesions may show features of immaturity and high cellularity that can be confused with a true malignancy. If surgeons are more aware with this rare disease, the accurate diagnosis can be made earlier, thus avoiding some unnecessary postoperative options.

Case presentation

A 3 year old male child presented with lump in abdomen since 2 years with history of occasional post prandial discomfort, no history of nausea, vomiting, distension, constipation etc. Clinical examination revealed nontender,

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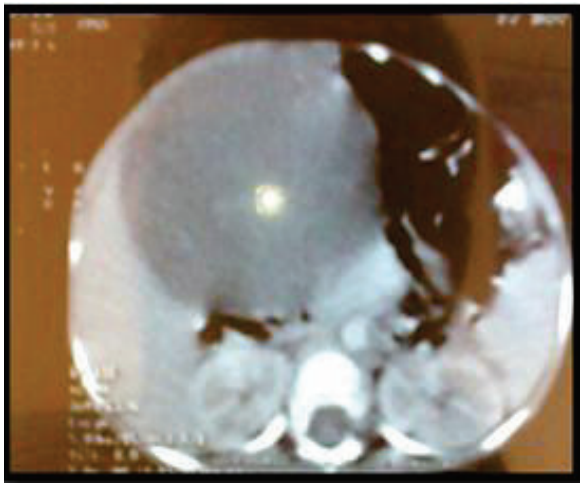
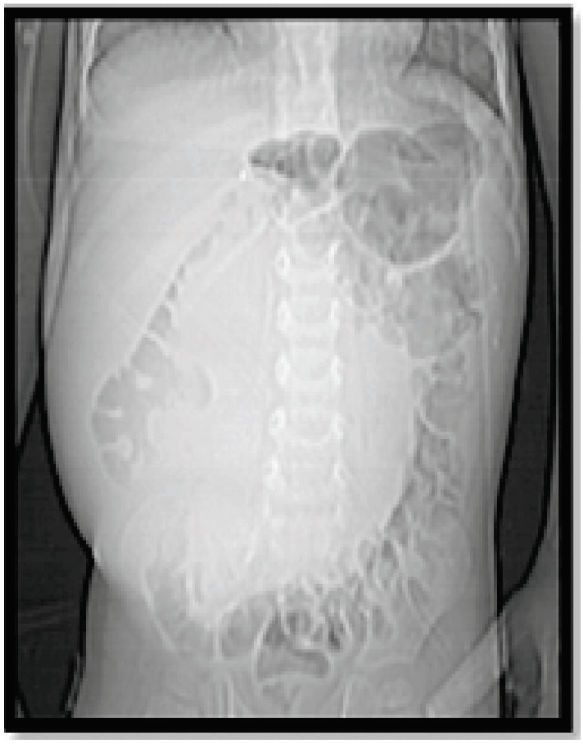
Fig 1: CECT of abdomen



firm mass in right Hypochondrium extending to umbilical and right lumbar region. Ultrasonography showed well defined heterogeneously hyperechoic lesion in right Hypochondrium and lumbar region extending up to umbilical area, displacing bowel loops on either side and measuring 11.5 cm x 11.8 cm.

Contrast enhanced computed tomography revealed a huge well demarcated hyperdense mass which displaced bowel loops without

Fig 2: Reconstructed coronal Section



obvious penetration to the Intestinal walls and normal intrabdominal organs. There was no paraaortic lymphadenopathy, no contrast enhancement (Fig 1 & 2). The laboratory studies were all within normal limits.

With suspicion of malignant neoplasm the patient underwent aspiration biopsy of the

Mass which was inconclusive. Thus a

Fig 3: Tumor on exploration

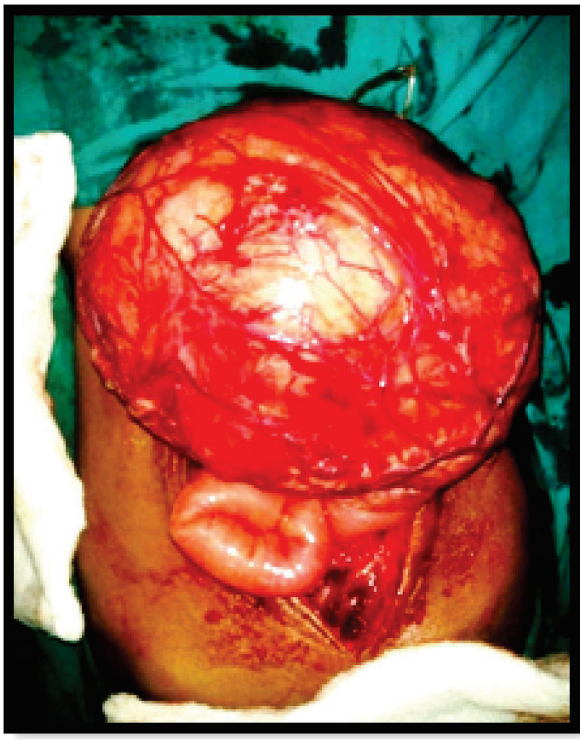


Fig 4: Tumor origin from Greater omentum

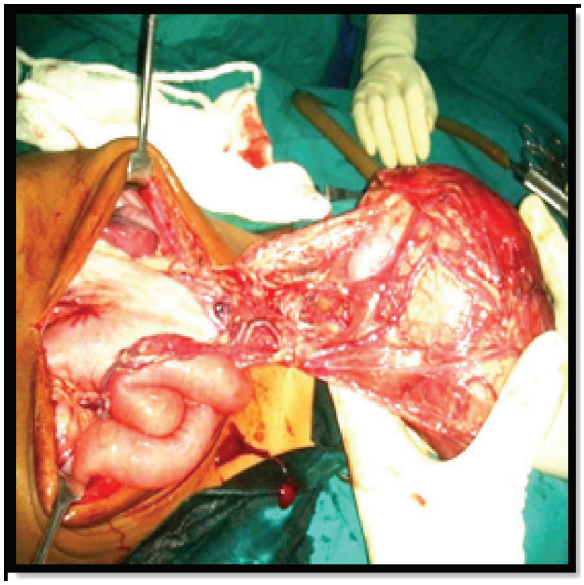
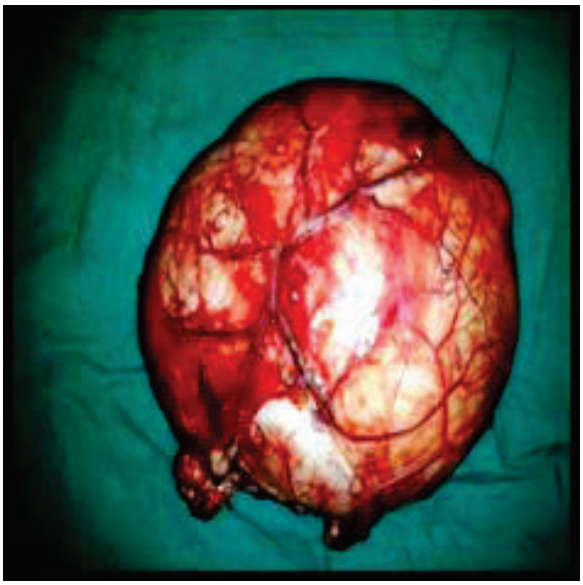


Fig 5: Well circumscribed Excised tumor mass.



presumptive diagnosis of neuroblastoma was made.

The patient underwent laparotomy; a huge, smooth surfaced, well-circumscribed tumor was found within peritoneal cavity (Fig 3). Origin of tumor was from greater omentum without any attachment to internal organs (Fig 4). It was accompanied by mesenteric lymphadenopathy. Tumor mass was removed completely with mesenteric lymph node biopsy.

Gross examination revealed a huge (12x 14 cm), solid, smooth surfaced, well encapsulated mass weighed 1100 grams (Fig 5 & 6).

Fig 6: Cut surface of tumour showing Solid and Myxoid areas.

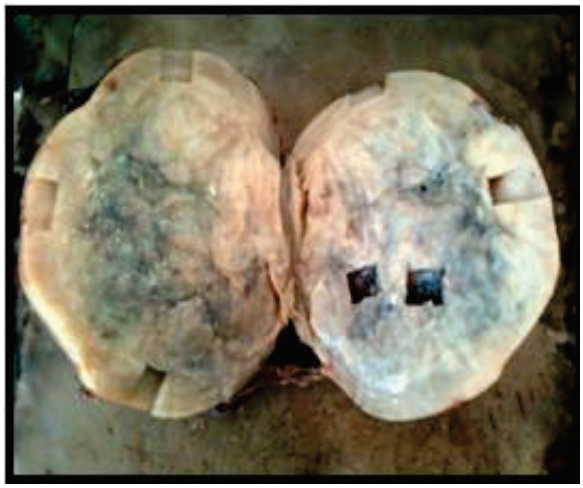
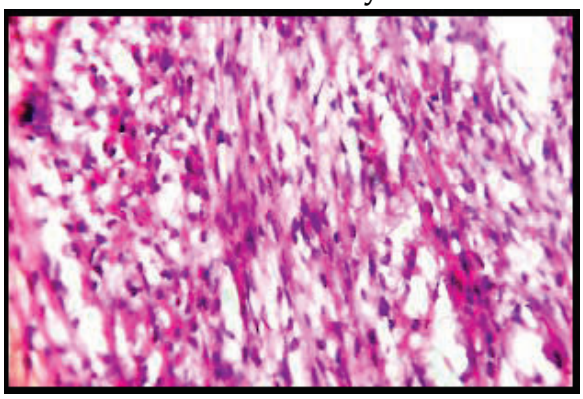


Fig 7: High magnification – fibroblasts in bundles separated by myxomatous stroma, spindle cells arranged in bundles, with a few inflammatory cells



Histologically tumor showed fibroblasts in bundles separated by myxomatous stroma, spindle cells arranged in bundles. Lipocytes admixed with spindle cells. Presence of few inflammatory cells but no evidence of neuroblastoma & malignant cells (Fig 7). There was no evidence of mitosis or necrosis.& secondaries in lymph nodes. Immunohisto-chemmically, tumor cells were strongly positive for vimentin and desmin. They were negative for EMA, NSE and MYO-D1.

All these findings confirmed the diagnosis of Omental- fibrous myxoid hamartoma. The patient had an uneventful postoperative course and was discharged from the hospital on the 10th post-operative day. No evidence of recurrence was noted during 2 years follow up.

Discussion

OMMH is a very rare lesion seen mainly during childhood [2]. As per previous available literature, only thirteen cases, including ours, have been reported. OMMH is considered as a variant of IMT, term used for nonspecific chronic inflammatory expansive lesions. Variants of IMT include : plasma cell granuloma or pseudotumor, inflammatory myofibrohistiocytic proliferation, myofi-broblastoma and OMMH[3].

Three basic tissue patterns are considered in IMT: a) myxoid/vascular pattern, b)

compact spindle cell pattern, and c) hypocellular fibrous pattern [4,5]. The clinical presentation seen is usually abdominal mass accompanied by variable symptoms of malaise, anorexia, vomiting, and abdominal distension, abdominal lump (the same as in our case).

The gross appearance of OMMH is a soft to firm, lobulated, smooth, pinkish white circumscribed mass with myxoid areas accompanied by numerous smaller myxoid masses in peritoneum.[4,6]

Microscopically, these lesions consist of a richly vascularised myxoid stroma with plump basophilic mesenchymal cells having vesicular nuclei and prominent nucleoli. Cellular foci alternate with areas of collagenisation. Occasionally, vacuolated and multinucleated cells are seen. Mesenchymal cells are immunoreactive for vimentin, smooth muscle actin and sometimes for desmin and S- 100 protein.[5]

Clinically OMMH is not a malignant neoplasm despite its cellularity, immature appearance, and resemblance to myxoid liposarcoma and other myxoid sarcomas. Lipoblasts are not a feature of Omental-mesenteric myxoid hamartoma and their absence differentiates it from lipoblastomatosis.[4] Other tumours which may be considered in the differential diagnosis of (OMMH) are atypical gastrointestinal stromal tumor, malignant fibrous histiocytoma, leiomyosarcoma, neuroblastoma and solitary fibrous tumor.[5]

However, the histological and immunohistochemical findings of our patient's tumor suggest the above-mentioned diagnosis.

Complete surgical resection is the treatment of choice for OMMH. In all previous reported cases of OMMH, no recurrence was noted with favourable prognosis.[5] Same was true for our case. All of the reported cases have survived with no evidence of recurrence following excision[5], OMMH is considered as a variant of IMT which can show unpredictable course with local recurrences, and hence long-term postoperative follow-up is recommended.[7]

Conclusion

Lesions of OMMH show features of aggressiveness along with immaturity and high cellularity that are more likely to be confused with a true malignancy. OMMH is a very rare lesion and because of its aggressive appearance, differential diagnosis with malignancy should be considered. The initial clinical picture of our case also led us to high suspicion of malignancy. However the histological and immunohistochemical findings helped us to achieve exact diagnosis and save the patient from unnecessary postoperative adjuvant treatments.

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