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A rare cause of intraabdominal tumor in childhood: Omental mesenteric myxoid hamartoma☆

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ABSTRACT

Omental masses are rarely seen in childhood. Omental mesenteric myxoid hamartoma is a very rare and a new entity, first described by Gonzalez-Crussi et al. The tumor originates from the omentum and mesentery and presents as multiple nodules. It shares many morphologic features with inflammatory myofibroblastic tumor and therefore, may be considered as a variant of inflammatory myofibroblastic tumor. However, the clinical course and prognosis of this tumor is different. This rare and relatively new pathology in childhood, mimicking malign tumor and sharing many histopathological features with inflammatory myofibroblastic tumor, is presented to emphasize that it is a clinically different entity in terms of clinical picture and prognosis.

Omental masses are rarely seen in childhood. Omental mesenteric myxoid hamartoma (OMMH) is a very rare and a new entity, first described by Gonzalez-Crussi et al. [1]. It is the tumor of the omentum and mesentery, presenting as multiple nodules. It is histologically characterized by plump mesenchymal cells in myxoid, well-vascularized stroma [1,2]. OMMH shares many morphologic features with inflammatory myofibroblastic tumor (IMT) and therefore, may be considered as a variant of IMT [3]. However, the clinical course and prognosis of the tumor is different from that of IMT. This rare and relatively new pathology in childhood, mimicking malign tumor and sharing many histopathological features with IMT, is presented to emphasize that it is a clinically different entity from IMT in terms of clinical picture and prognosis.

1. Case report

A 15-year-old girl admitted with a rapidly growing abdominal mass in the last 3 months which completely filled the left upper quadrant. There was no history of trauma, abdominal pain, nausea or vomiting.

Physical examination revealed a firm, immobile solid mass filling the left upper quadrant and epigastric region. Other examination findings were normal. Routine laboratory findings and erythrocyte sedimentation rate were also normal. Abdominal Doppler ultrasonography showed a 8 × 6 cm, vascularized, hipoechoic solid mass with necrotic foci adjacent to the left kidney, the spleen and the tail of pancreas and that the tumor was compressing these organs. Abdominal magnetic resonance imaging revealed a solid mass of 93.7 × 87.7 mm in diameter in the coronal plane and 79 × 71 mm in the axial plane (Fig. 1). The tumor was displacing the left kidney and the spleen. Thoracal computed tomography was normal. The tumor markers (CA 15-3, CA 19-9. CA 125, βHCG) were negative.

The patient was operated on with the prediagnosis of gastrointestinal stromal tumor or soft tissue sarcoma. At laparotomy, a well-circumscribed tumor was found in the greater omentum. The mass was rubbery, glistening and oedematous in nature and 8 × 10 cm in size. The reddish-yellow tumor which was mainly solid with some cystic elements showed regular modularity and increased vascularization (Fig. 2). The tumor was adherent to spleen and stomach and it displaced them superiorly and laterally. However, there was no adhesion to adjacent organs and structures. The mesenterium was normal. There was no obvious regional and mesenteric lymph node enlargement or no intraperitoneal fluid. The mass was totally excised.

Histologically, the tumor consisted of a richly vascularized myxoid stroma with plump mesenchymal cells (Fig. 3a). There was no evidence of mitosis or necrosis. Immunohistochemically, tumor

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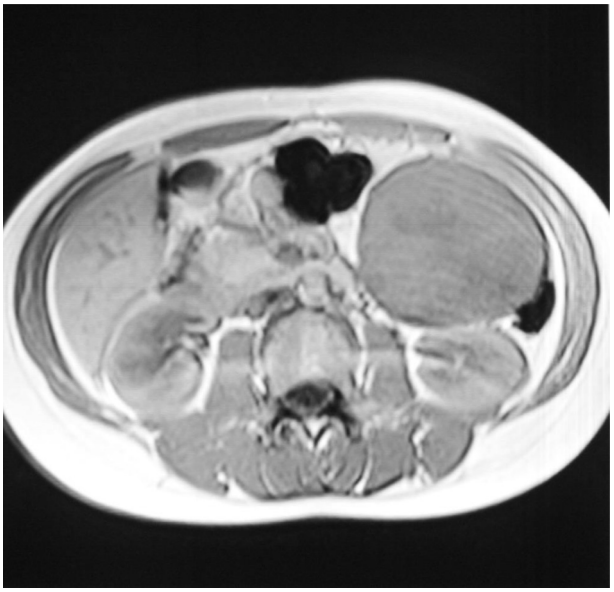

Fig. 1. Abdominal magnetic resonance imaging of the tumor compressing the left kidney and the spleen.

cells were positive for vimentin, negative for S-100, CD34, CD117, smooth muscle actin and ALK (Fig. 3b). The histopathological examination revealed that the mass was omental mesenteric myxoid hamartoma.

The postoperative course was uneventful. No early recurrence was observed during one-year follow-up.

2. Discussion

OMMH is a very rare tumor of gastrointestinal system. 10 cases have been reported in the literature up to now [1–9]. It is a tumor characterized with multiple omental and mesenteric nodules. Although there are no large series about omental mesenteric myxoid hamartoma in the literature, it is considered as a benign tumor. Therefore, complete surgical resection is the only choice of treatment. In the presented case, the tumor was totally and completely resected with no complication.

Some authors report that the basic tissue patterns in IMT are myxoid/vascular pattern, compact spindle cell pattern and hypocellular fibrous pattern. Tumors growing in a grape-like fashion in a myxoid/vascular pattern are considered indicative of OMMH [4]. Especially these tumors may be multinodular and be localized

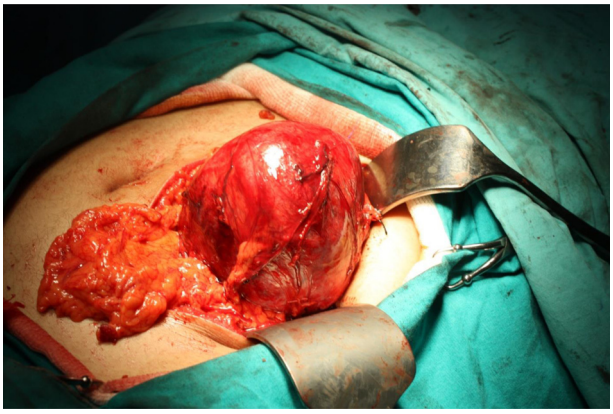

Fig. 2. Intraoperative picture of the omental tumor.

inside the abdomen like OMMH. One of the most important clinical differences between OMMH and IMT is the localization of the tumor. The other difference is the prognosis. IMTs are more commonly localized in lungs, eye, brain and pericard. They are rarely seen in the alimentary tract [10,11]. OMMT, however, originate from the gastrointestinal system. In the IMT cells, there may be deceptive features of immaturity, high cellularity and aneuploidy. Besides, associated chromosomal anomalies and recurrences may be observed in IMT. No recurrence has been reported in OMMT up to now and they are considered as benign tumors [1,2,4–9]. In this presented case, no recurrence was observed in one-year follow-up.

Histologically, OMMH consists of plump basophilic cells containing vesicular nuclei and prominent nucleoli in a richly vascularized myxoid stroma. The myxoid matrix contains acid mucopolysaccharides and the mitotic rate is low. Cellular foci alternate with areas of collagenization. Rarely, vacuolated and multinucleated cells are seen [1,2,12]. Mesenchymal cells are immunoreactive for vimentin, smooth muscle actin and sometimes for desmin and S-100 protein [4]. In the presented case, tumor cells were positive for vimentin, negative for S-100, CD34, CD117, smooth muscle actin and ALK.

Ultrastructural examination reveals immature mesenchymal cells, fibroblasts and vacuolated cells with dilated cisternae of rough endoplasmic reticulum and dense structures resembling lysosomes [1]. In some studies, the presence of clonal chrosomal aberrations in the tumor has been reported to indicate that OMMH is a neoplastic proliferation [2]. Gastrointestinal stromal tumors (GISTs) originating from the omentum resemble omental mesenteric myxoid hamartoma histopathologically and immunohistochemically. Microscopic examination dominantly reveals spindle cell lesions and in less than 15%, myxoid and epitheloid tumors. It is

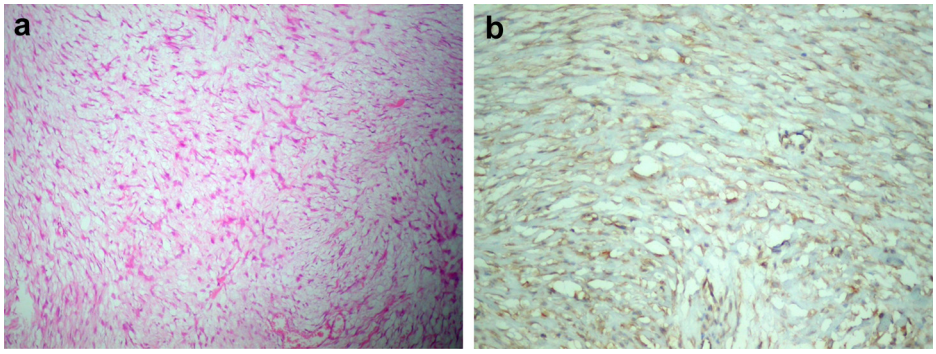

Fig. 3. a: Plump mesenchymal cell proliferation in myxoid background (H&E, ×20). b: Immunohistochemistry demonstrates positive staining of these cells with antibodies to Vimentin (×200).

important to distinguish OMMH from omental GISTs, because complete surgical resection is curative in OMMH. However, further medical treatments such as tyrosine kinase inhibitors are usually required together with surgery in omental GISTs. Moreover, differential diagnosis of inflammatory malignant fibrous histiocytoma, leiomyosarcoma, solitary fibrous tumor, cell-rich gastrointestinal autonomic nerve tumor, myxoid sarcoma and myxoid liposarcoma from OMMT should also be made [4,13].

3. Conclusion

Although OMMH is clinically a benign neoplasm, the clinical picture and histological findings may mimic malign tumors of the gastrointestinal system. The cellularity of the tumor and immature appearance resemble malign tumors. The experience of both the surgeon and the pathologist about this entity prevents unnecessary postoperative adjuvant treatments. Omental myxoid hamartomas must be considered in the differential diagnosis of abdominal masses in childhood. Patients with OMMH may not need long-term follow-up and the parents of the patients can be informed about the prognosis more clearly.

Conflict of interest and sources of funding statement

Authors claim no conflict of interest and no financial conflicts.

Consent

Written informed consent was obtained from the parent's of the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the editor-in-Chief of this journal on request.

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