GAstrIC MIOFIBROBLASTIC INFLAMMatory TuMOR IN A 4-YEAR OLD FEMALE

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ABSTRACT

Summary: We present the case of a 4-year old female admitted for management of a gastric tumor. Following the routine preoperative assessment, the patient underwent exploratory laparotomy with partial gastrectomy and removal of a 15-cm tumor. Histochemistry analysis using antibodies against CD117, vimentin, desmin, smooth muscle, S100 and Ki67 made the diagnosis of inflammatory myofibroblastic tumor (IMT). The patient received no adjuvant chemotherapy. No recurrence was detected during the 9-month follow up. It is reiterated that the clinical presentation of gastric IMT is not specific and only histochemistry can differentiate between IMT and other high-grade malignancy conditions.

Key words: tumor, gastric, inflammatory, myofibroblastic, child.

Introduction

The Inflammatory Myofibroblastic Tumor (IMT) was initially considered as a soft tissue inflammatory pseudo-tumor of the young adult patient (1). The pulmonary localization of IMT was described in 1937 (2). Similar lesions were also described in the abdomen, head and upper limbs (3-8). Murphy (1994) performed a review of 1,403 gastric tumors and identified only three cases in children, one of which was diagnosed as ‘plasma cell granuloma’ (9). Cho, in 2002, reported the case of a 2-year old boy with a myofibroblastic tumor originating in the stomach. He also made a literature review and found 13 similar cases of tumors originating in the stomach (10).

Case report

A 4-year old girl was admitted for non-specific complaints, which included: nausea and vomiting decreased appetite, weight loss, dysuria and hematuria. Physical examination revealed an abdominal tumor localized in the left middle and lower quadrants, respectively. The mass was mobile and could also be intermittently palpated in the epigastrium, but looked to ’drop’ in the LLQ when the patient was standing up. The mass was firm, not painful and had the size of a grapefruit. Patient’s previous history was not significant. She was a second born child to a normal pregnancy, with no complications during or after delivery. Birth weight was 3,300 g; Apgar score was 10 at 1 and 5 min, respectively. Weight and height at present admission were age-appropriate.

Laboratory tests were within normal limits, except for a moderate degree of thrombocytosis. Several imagistic studies were performed, as follows: chest X-ray was normal; abdominal X-ray showed gastric distension and increased liver size. Abdominal US identified an 11-cm size round-shaped hyperechogenic tumor located in the left middle and lower quadrants, respectively. Abdominal CT Scan identified a 7 cm x 11, 5 cm x 12 cm tumor located in the upper and middle abdominal areas, well delineated and with increased density after i.v. contrast dye injection. The tumor was compressing the left liver lobe, the stomach and the small bowel loops, but had fairly distinct margins.

Figure 1: The tumor found on the anterior side of the stomach

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There was no obvious retroperitoneal lymphadenopathy. The tumor was considered to arise from the stomach (lymphoma versus leiomyoma of the greater curvature) and surgery was decided.

Laparotomy was performed by the senior author (GA). A round shape tumor with a 2 x 2 cm implantation base on the anterior side of the stomach was identified (Fig. 1). The base of the tumor was located 3 cm away from the greater curvature and 4 cm away from the pylorus. It then invaded the greater omentum and the gastro-colic ligament. The tumor had a firm consistency and irregular surface, suggestive of malignancy (Fig. 2). It also displayed an extensive superficial vascular network, but there were no apparent areas of hemorrhage or necrosis. Large resection of the tumor with a 2-cm gastric wall margin was performed, as well as dissection of retrogastric lymph nodes.

In what concerns pathology, frozen sections showed a tumor pattern composed of fusiform cells, consistent with gastro-intestinal stromal tumor (GIST). However, permanent hematoxilin-eosin (HE) and tricromic stains identified a different pattern of fusiform cells arranged in fascicles and a rich inflammatory cell proliferation composed of lymphocytes, plasmocytes, histiocytes and few granulocytes (Fig. 3). The tumor cells had a stelate appearance within the myxoid areas, but lacked both nuclear/ cytoplasmic atypical features and significant mitoses. Immunohistochemistry stains were strongly positive for vimentin, slightly positive for desmin, and negative for CD117 (c-kit) and S100 protein (Fig. 4). Ki67 also showed a weak positive stain (< 5% of tumor cells).

The postoperative course was uneventful. The patient was admitted in the ICU for 2 days after surgery. No chemotherapy was given during the postoperative course. The patient resumed oral diet on the 5th postoperative day and was discharged home on the 7th postoperative day. She was then followed in our clinic at 1, 3, 6 and 9 months after surgery, respectively. Abdomen US, flexible endoscopy and Abdominal CT Scan results were negative for residual or recurrent tumor.

Discussion

The inflammatory myofibroblastic tumor (IMT) is a rare, yet distinctive pseudosarcomatous inflammatory lesion, which primarily occurs in the soft tissue and abdominal organs of children and young adults. The etiology of IMT is not clearly established. Some investigators believe IMT is a true neoplasm, while others consider it as an immunologic response to aggression by infectious or noninfectious agents (15). The histological appearance is of fascicles of band myofibroblasts located inside a variable collagenous stroma. A prominent inflammatory component of lymphocytes, eosinophiles, and plasma cells is also present. Its distinctive histological appearance has given this lesion a wide variety of names, including inflammatory pseudotumor, pseudosarcomatous myofibroblastic proliferation, inflammatory sarcoma, plasma cell granuloma, and inflammatory myohistiocytic proliferation (11–13).

Immunohistochemistry studies were able to confirm the mesenchymal nature of the tumor proliferate with positive reactivity for vimentin and smooth muscle actin protein, while stains for desmin, myoglobin and S100 protein are negative (10). In order to assess the degree of tumor aggressiveness, additional markers like PCNA, bcl-2 and p53 can also be used. These markers however were not shown to correlate with the rate of local recurrence, distant metastasis or death (14).

Cytogenetics and FISH studies confirmed the neoplastic nature of the mesenchymal proliferation in IMT. The inflammatory component of the tumor is only reactive in nature and there looks to be no actual inflammatory neoplasia process. Several chromosomal aberrations were identified in the 2p23 area, which involved the ALK gene (12). TPM 3-ALK and TPM 4-ALK oncogenes were identified in IMT cases, but also in cases of anaplastic lymphoma (21). This finding could support the hypothesis of both tumors having a common origin. The clinical picture of IMT varies depending on the site of the tumor. Abdominal IMTs present as a mass associated with inflammatory symptoms, impaired growth and symptoms related to lo-

Figure 2: The tumor with a firm consistency and irregular surface, suggestive of malignancy

Figure 3: Microscope findings - pattern of fusiform cells arranged in fascicles and a rich inflammatory cell proliferation (lymphocytes, plasmocytes, histiocytes and few granulocytes)
cal compression such as pain and vomiting (4, 10, and 16). Very few cases with gastric location were described; most (69.2%) of the patients were females and younger than 6 year old by the time of diagnosis (10). Clinical presentation for gastric tumors is non-specific, including: fever, weight loss, pallor, malaise, abdominal pain or mass, UGI bleeding, nausea and vomiting, decreased appetite, GER, growth retardation (5, 9, and 10). No specific symptoms were described so far for the IMT cases with gastric location.

Diagnosing a gastric IMTs with the help of conventional imaging (US, CT scan) or endoscopy studies is difficult. It was found that some of these tumors had extragastric adherence to spleen, pancreas, porta hepatitis or pericardium (17). The most common preliminary diagnosis in these cases is gastric lymphoma.

Current treatment for gastric IMT is complete surgical resection. Chemotherapy has not been proven effective for this tumor (18). Recurrence was only mentioned in just one case of gastric IMT (9), while retroperitoneal or mesenteric IMTs were reported to have a 15–37% local recurrence rate (20). An increased tendency for local recurrence seems to be related to the initial extrapulmonary site of the tumor.

Conclusions

The paper presents the case of a 4-year old girl with a gastric inflammatory myofibroblastic tumor (IMT). The clinical presentation was non-specific and malignancy could not be ruled out until pathology became available after surgery. The treatment only consisted of laparotomy and complete tumor resection; no chemotherapy or radiotherapy was given. There was no recurrence identified after 9 month follow up. We concluded that the initial diagnosis work up for a gastric tumor should include IMT. Pathology exam should include immunohistochemistry tests to confirm the mesenchymal nature of the tumor proliferates.

REFERENCES