

Case Report

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Intra Abdominal Fibromatosis Mimicking GIST; A Case Report and Review of the Literature



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Abstract

A 22 years old female patient, presented with a painless abdominal mass of 2 weeks duration, without other symptoms. A mobile, non-tender globular mass was felt at the left upper quadrant. A computed tomography revealed a mass lesion, measuring 8.3*8.5cm, without signs of bowel obstruction. At surgery, a large, 8cm mass was found. The mass involved the transverse colon, with its mesocolon and adhered to the greater curvature of the stomach. The mass was excised completely along with a wedge resection of part of the transverse colon and part of the greater curvature. Pathological report revealed an intra-abdominal fibromatosis.

Keywords: GIST; Intra-abdominal fibromatosis; Tyrosine kinase inhibitor; Familial polyposis coli

Introduction

Intra-abdominal fibromatosis (IAF) is a rare, benign mesenchymal tumor, that can develop at any age. Although rare, it is the most common primary tumor of the mesentery [1]. It can be solitary or multiple with remarkable local aggressiveness. The tumor usually occurs in the mesentery or retroperitoneum, but can occur throughout the gastrointestinal tract [1-3]. IAF is usually secondary to trauma or hormonal stimulation or associated with familial polyposis coli or Gardner's syndrome [2,4]. Clinical, radiological and histopathological appearance might mimic the appearance of gastrointestinal stromal tumor (GIST), thus leading to misdiagnosis [5]. Discrimination between the two entities is very important, due to the fact that they have different biological behaviors and different therapeutic strategies; IAF is a benign and with exclusive aggressiveness, usually treated by means of surgical excision with a wide margin of uninvolved tissue, whereas GISTs are usually malignant and may lead to distant metastases, with a significant role of tyrosine kinase inhibitors in addition to surgery in their management [5,6].

As mentioned previously, IAF usually treated by means of surgical excision with a wide margin of uninvolved tissue; however, local recurrence due to incomplete resection have been

described in an estimated 16% of cases, leading to intestinal obstruction and death in 6% [3]. We report a case of a primary intra-abdominal fibromatosis treated in our department.

Case Report

A 22 years old, female patient, presented to our emergency department complaining of a painless, palpable abdominal mass at the left upper quadrant of 2 weeks duration. She denied anorexia, weight loss, vomiting or change in bowel habits. Her past medical and surgical histories were unremarkable. She had no family history of malignancy. On admission; her vital signs were within normal limits and on physical examination, a non tender, mobile mass at the left upper quadrant was palpated. Her abdomen was soft, not distended and no other masses were detected. Complete blood count, urinalysis, liver and kidney function tests were all within normal limits. A CT scan of her abdomen and pelvis (Figure 1) revealed a hyper dense lesion with high vascularity, measuring 8.3cm*8.5cm*6.6cm, adhering to the transverse colon and large curvature of the stomach, without evidence of bowel obstruction or intraperitoneal spread. These findings raised a suspicion of gastrointestinal stromal tumor (GIST) at first. The patient was admitted to our department for further work up. Chest CT scan was unremarkable. Endoscopic

ultrasound (EUS) revealed heterogenous mass lesion, about 86mm, behind the stomach without involvement of the pancreas. EUS- guided FNA was performed, yielding a paucity of material, not enough for cytological analysis. At colonoscopy, a submucosal lesion, compressing the lumen externally was identified about 55cm from the anal verge. Tumor markers -CA19-9, CEA and CA125 were within normal limits. Subsequent exploratory laparotomy revealed a large mass (Figure 2), about 8cm in diameter, adhering to the transverse colon with its mesocolon and also to the greater curvature of the stomach, without involvement of the pancreas. The mass was excised with the transverse colon and its mesentery and a wedge resection of the stomach. The patient recovered well from the operation. Pathology report revealed an intra-abdominal fibromatosis (IAF), and the cells were stained positive to Beta-catenin and negative to CD117 and DOG-1.

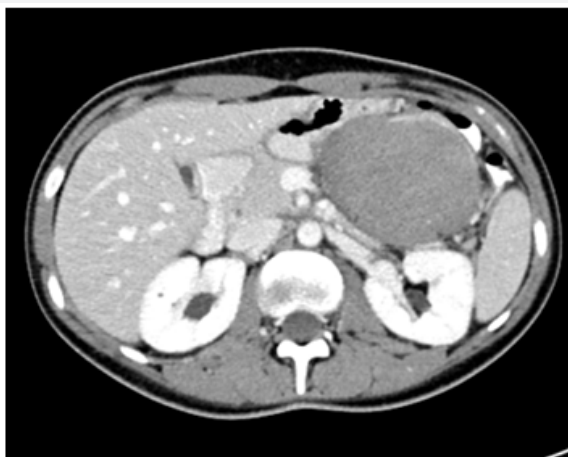


Figure 1: CT scan showing the mass lesion.



Figure 2: The mass after excision.

Discussion

An intra-abdominal fibromatosis (IAF), although known as the most common primary tumor of the mesentery, is a rare

benign neoplasm [1]. It is usually secondary to trauma or hormonal stimulation or associated with familial polyposis coli or Gardner's syndrome, or it could be primary, which is a rare variant of benign stromal neoplasms of fibroblast/myofibroblast origin [2-5]. Diagnosis of IAF is fraught with difficulties, especially when it presents as a tumor adhering to the intestinal wall, thus the diagnosis of GIST may be considered. Comparing to GIST, in which the tumor is usually soft and lobulated with hemorrhage, necrosis or cystification, patients with IAF tend to be younger and the tumor is firm, tan and homogenous. These entities can also be distinguished primarily by their light microscopic and ultrastructural features, but there is a notable overlap in their immunohistochemical profiles (in one study: 88% of GIST positive to CD117 while 75% of IAF positive to CD117) [6]. As mentioned above, distinguishing between the two entities is important due to the different treatment protocols and biological behavior. IAF is a low grade neoplasm, treated usually by surgical excision, that may recur, but never metastasizes. Recurrence is usually related to incomplete excision. Tamoxifen has been used in the management of a recurrent or unresectable tumor, but it needs to be investigated more. Prognosis of IAF is excellent because it never metastasizes.

Conclusion

A primary Intra-abdominal fibromatosis is a very rare benign tumor, which usually affects the mesentery. The mainstay of treatment is surgical excision, with recurrence mostly related to incomplete excision.

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