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Primary mesenteric fibromatosis: Report of a sporadic case

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Abstract

Mesenteric fibromatosis or intra-abdominal desmoid tumour is a rare neoplastic, monoclonal myofibroblastic proliferative disease affecting the mesentry which is prone to aggressive local recurrences, but lacks metastatic potential. Primary or spontaneous mesenteric fibromatosis is rare which occurs in absence of any predisposing conditions. Here, we report a case of a 66 year old male who presented with an abdominal mass. Abdominal examination revealed a firm mobile non-tender mass palpable in the lower portion of epigastrium. Ultrasonography showed a solid lesion of mesenteric origin. He underwent surgical excision of the mass with resection of involved small bowel and end to end anastomosis. Histopathology showed fascicles of bland spindle shaped cells with dilated blood vessels and keloidal collagen fibres. Immunohistochemistry showed β-catenin +ve, CD 117-ve and CD 34 –ve, which is confirmative of fibromatosis. Postoperative period was uneventful. Complementary therapies were not suggested in our patient as the tumour was a primary desmoid-type fibromatosis with complete resection and tumour free resection margins.

Keywords: Mesentery; Myofibroblasts; Fibromatosis, Abdominal; Immunohistochemistry; Beta catenin

Introduction

Mesenteric fibromatosis (MF) or intra-abdominal desmoid tumour is a rare neoplastic, monoclonal myofibroblastic proliferative disease affecting the mesentery accounting for 0.73% among all abdominal tumours with incidence rate of 2-5/ million/ year. MF is prone to aggressive local recurrences, but lacks metastatic potential. Despite its rarity, mesenteric fibromatosis is the most common mesenteric tumour. Most reported cases have been in association with Gardner's syndrome, previous trauma and prolonged intake of estrogen, but primary or spontaneous mesenteric fibromatosis is rare which occurs in absence of any predisposing conditions.[1,2,3,4]

Case Report

A 66 year old male presented with complaints of vague abdominal pain and discomfort since 3 months. He had no history of abdominal trauma or surgical therapy for any other disease. Abdominal examination revealed a firm mobile non-tender mass palpable in the lower portion of epigastrium. All his baseline blood investigations were normal. An abdominal ultrasound showed a large solid lesion measuring 10cm at its greatest diameter arising from mesentry with minimal vascularity. With a provisional diagnosis of neoplastic solid lesion probably of mesenteric origin, the patient was posted for laparotomy. Elective laparotomy revealed a firm mass measuring 10x9cm that appeared to originate from jejunal mesentry. Resection of small bowel along with the mass was done with end to end anastomosis. The post operative course of the patient was uneventful and patient was discharged healthy on the seventh postoperative day.

Pathological findings: Tumour mass measuring 10x9x6cms was extramural in location and exhibited well circumscribed borders. The specimen weighed 500grams. Cut section revealed a tan-gray glistening surface with trabeculations. Surgical specimen was subjected to histopathological examination. [Fig. 1]

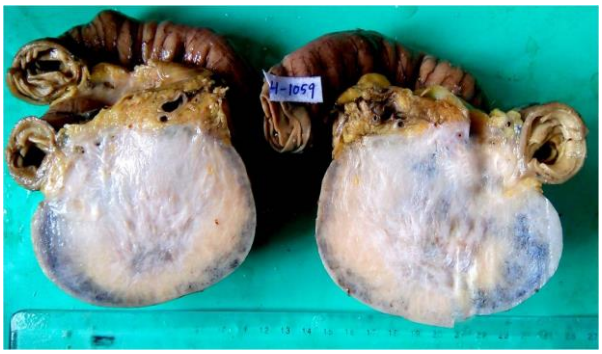

Fig. 1: Gross specimen of mesenteric fibromatosis along with attached segment of small bowel

The histopathology of the specimen showed a tumour composed of spindle shaped cells arranged in long fascicles. The cells were cytologically bland with areas of keloidal collagen admixed with dilated blood vessels. The mitotic count was relatively low. Tumour margins were of infiltrative type with nodular lymphoid aggregates at the advancing edge of the tumour. [Fig. 2] Resected margins were free from tumour.

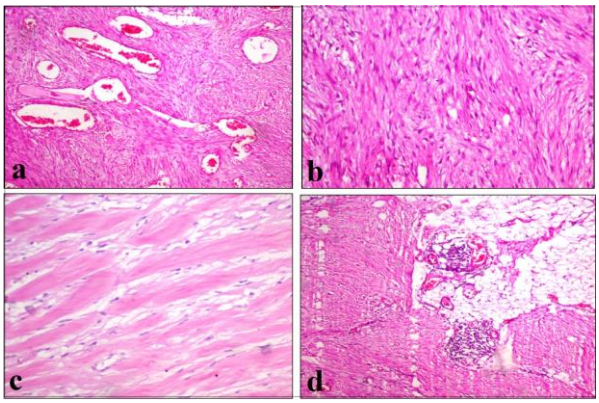

Fig. 2: (a) Microscopy showing intersecting fascicles composed of bland looking spindle shaped cells admixed with thin walled dilated blood vessels (H and E, x10). (b) Cytological characteristics of MF with stellate fibroblasts (H and E, x20). (c) Prominent keloidal collagen fibers (H and E, x20). (d) Infiltrative margins into mesenteric fat with lymphoid aggregates (H and E, x10)

Immunohistochemically, nuclear positivity for β - catenin was observed and the tumour was negative for CD117 and CD34, which is confirmative of Fibromatosis. [Fig. 3]

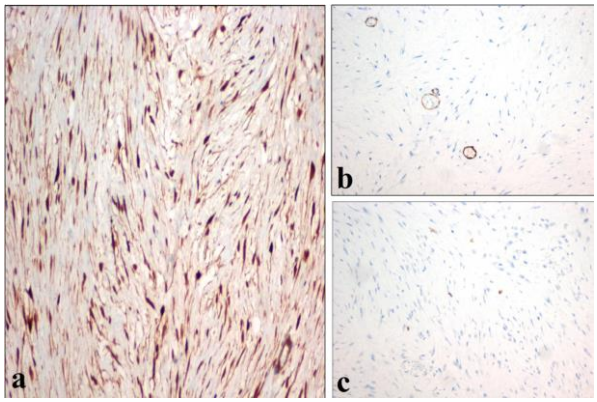

Fig. 3: (a) Immunochemistry positive for beta-catenin. (b) Negative for CD34. (c) Tumor is negative for CD117

Discussion

Fibromatoses comprise a broad group of benign fibroblastic proliferations of similar microscopic appearance characterised by infiltrative growth and a tendency toward recurrence, but they never metastasize.[5]

Mesenteric fibromatosis is characterized by an infiltrative pattern of growth and a tendency to local recurrence when excised incompletely and/or associated with Gardner syndrome.[6]

Majority of patients with mesenteric fibromatosis remain clinically asymptomatic, with little or no focal symptoms until later in their course, at which stage they complain of abdominal pain and discomfort, constipation, vomiting, weight loss and organ compression symptoms. The differential diagnosis for mesenteric fibromatosis includes Gastro Intestinal Stromal Tumour (GIST), lymphomas, carcinoids, fibrosarcomas and inflammatory fibroid polyps. GIST being the most common differential diagnosis can be differentiated by a positive CD 34 staining and absent nuclear β-catenin.[4]

The etiopathogenesis of mesenteric fibromatosis has been unclear for many years. The predisposing factors known to play a role in the onset of this tumour are Gardner syndrome, pregnancy, abdominal surgery or trauma, estrogen as growth factor whereas in our case there was no history of abdominal trauma or surgical therapy for any other disease. Currently, these tumours are regarded as clonal proliferation of myofibroblasts that show adenomatous polyposis coli (APC) gene mutations. The resultant beta-catenin over expression and accumulation is known to trigger fibroblastic proliferation through a nuclear mechanism.[7,8]

The treatment modality in mesenteric fibromatosis is still controversial, but wide local surgical resection with tumour-free margins remains the ideal treatment. The Standard Task force of the American Society of Colon and Rectal Surgeons suggest that surgery should be reserved for small tumours with a well-defined and clearly resectable margin. Surgery is not recommended if the disease has advanced locally or if vital structures or major nerve trunks are involved. The alternative approaches known are radiation therapy, chemotherapy and tamoxifen. Treatment modalities other than surgical excision are controversial.

In our case, complementary therapies were not suggested as the tumour was a primary desmoid-type fibromatosis with complete resection and tumour free resection margins. As noted in our case, most of these lesions require resection of attached segment of bowel. The patient has shown no signs of recurrence at present 14 months after surgery.

Conclusion

Mesenteric fibromatosis is a rare benign myofibroblastic tumour arising is small bowel mesentry posing a diagnostic difficulty in differentiating from GIST. Immunohistochemistry with β-catenin, CD117 and CD34 helps in diagnostic differentiation.

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