Journal of Cancer Therapy, 2014, 5, 1000-1004
Published Online September 2014 in SciRes. http://www.scirp.org/journal/jct
http://dx.doi.org/10.4236/jct.2014.511104

Sporadic Desmoid Tumor of the Small Bowel Mesentery in a Male Patient: A Case Report and Literature Review

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Received 20 July 2014; revised 18 August 2014; accepted 12 September 2014

Abstract

Aggressive fibromatosis arising from the small bowel mesentery is extremely rare. It may occur in association with previous trauma, abdominal surgery, drugs, Gardner's syndrome, or familial adenomatous polyposis. This paper presents a 32-year-old man with no significant medical or surgical history, complaining of diffuse abdominal pain and discomfort. His computed tomography scan revealed a well-defined soft tissue mass in the peritoneal cavity. He underwent surgical excision of the mass with resection and anastomosis of the involved loop of the small intestine. Histological examination confirmed mesenteric fibromatosis without infiltration of the bowel. The patient remained well during the 8 months follow-up.

Keywords

Desmoid Tumor, Mesenteric Fibromatosis, Surgery

1. Introduction

Desmoid tumors (DT) are rare benign, monoclonal, myofibroblastic proliferations that originate from musculoaponeurotic structures. They are slow-growing and locally aggressive tumors with strong tendency to recur after surgical resection, but lack the capacity to metastasize. Mesenteric DT, also known as mesenteric fibromatosis, is a part of the clinical-pathologic spectrum of deep DT and represents the most common primary mesenteric neoplasm.

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How to cite this paper: Amaadour, L., Benbrahim, Z., Benhayoune, K., Chbani, L., Arifi, S. and Mellas, N. (2014) Sporadic Desmoid Tumor of the Small Bowel Mesentery in a Male Patient: A Case Report and Literature Review. *Journal of Cancer Therapy*, **5**, 1000-1004. http://dx.doi.org/10.4236/jct.2014.511104

We present here a case of sporadic mesenteric desmoid tumor, and we review the literature data concerning this extremely rare disease.

2. Case Report

A 32-year-old man presented with history of diffuse abdominal pain and discomfort of four months duration. He had no past history of colonic polyps, abdominal trauma or surgical therapy for any other disease. His general physical examination was unremarkable. Abdominal examination revealed a mobile, non-tender, firm and globular supra-umbilical mass.

A contrast enhanced computed tomography (CT) scan showed a well-defined soft tissue mass in the peritoneal cavity (**Figure 1**).

A laparotomy was performed which found a well vascularised mass measuring 10 cm in diameter arising from the mesentery of the proximal ileum. The mass was completely excised with a part of the adherent bowel. The histological examination of the specimen demonstrated the mesenteric fibromatosis without mitosis or infiltration of the bowel, and with CD117, DOG1, MDM2, AML negativity (**Figure 2**).

The postoperative period was uneventful and patient was free of recurrence during the 8 months follow-up period, as observed by a three monthly CT scan and physical examination.

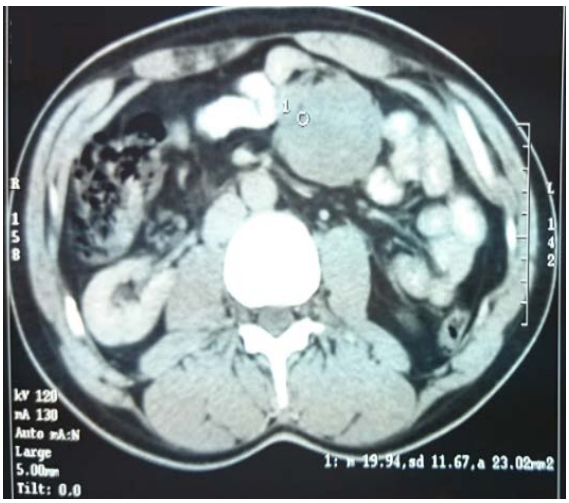
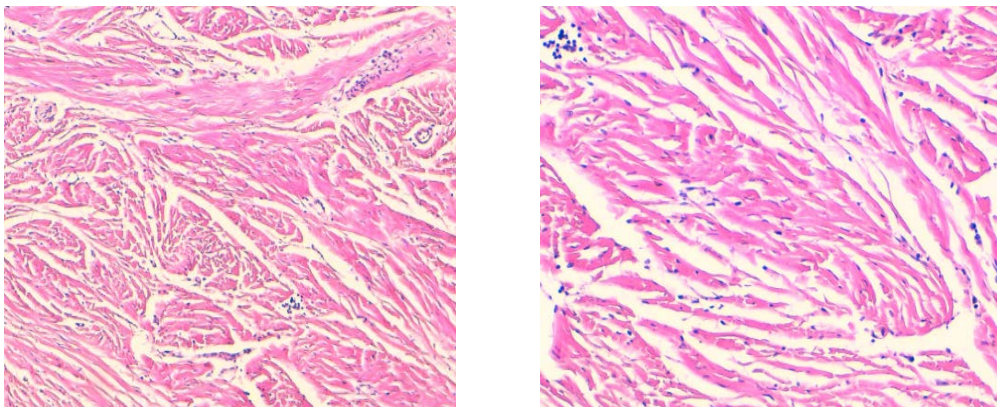


Figure 1. Computed abdominal tomography showing a well defined mass in the mesenteric area, related to the small bowel wall.



(a) (b)

Figure 2. (a, b) Progressive increasing magnification showing tumor composed of fascicles of fibroblastic spindle cells with abundant intercellular collagen without mitosis or necrosis.

3. Discussion

DT are rare; they account for approximatively 0.03% for all neoplasm [1]. They can affect any superficial or deep part of the body, but abdominal wall and extremities are sites of predilection [2]. Very rarely, DT involve the mesentery, omentum and retroperitoneum [3]-[5]. The small bowel mesentery is the most common site of origin of intraabdominal fibromatosis [6]. Mesenteric fibromatosis may occur at any age, but is most frequently found in young adults usually between the ages of 25 and 40 years with a strong prevalence among women of childbearing age [7]. Most mesenteric fibromatosis tumors manifest sporadically. In some cases, they may be secondary to trauma, especially postoperative trauma [8] [9], or hormonal stimulation. Genetic predisposition also plays a role and patients with familial adenomatous polyposis coli, specifically, the Gardner's syndrome variant, have a much greater potential of developing mesenteric fibromatosis compared with the normal population [6] [10]. The majority of patients with mesenteric fibromatosis remain clinically asymptomatic until later in their course, in which stage they complain of painless enlarging mass, discomfort or symptoms caused by compression of the adjacent structures and their vascular supply such as small bowel obstruction and hydronephrosis [6] [11]. The imaging studies are needed for identifying, characterizing and staging fibromatosis. Both CT and MRI are valuable modalities for the diagnosis and differential diagnosis of DT, since features of desmoid-type fibromatosis are characteristic of DT, but MRI is considered the primary modality for the imaging of DT [12] [13]. On CT scan, these tumors appear as masses with soft tissue density, causing compression or dislocation of the adjacent structures [3]. Characteristic MRI findings of DT include poor margination, low signal intensity on T1-weighted images and heterogeneity on T2-weighted images, and variable contrast enhancement. Low T2 signal intensity bands are characteristic and represent foci of high concentrations of collagen deposition [14] [15].

As there is no classical symptomatology or specific radiological features related to mesenteric fibromatosis, the diagnosis is confirmed only after biopsy with histopathological analysis of the tumor. Microscopically, mesenteric fibromatosis is composed of proliferating stellate to spindle cells arranged in long fascicles or whorling patterns with bland nuclear features and dense keloid-like collagen in areas [2] [16]. The cells usually show no nuclear atypia or hyperchromasia. Immunohistochemistry demonstrates tumor cells being strongly positive for vimentin and showing variable reaction to smooth muscle actin. Nuclear beta catenin immunore activity is commonly expressed with 67% - 80% of cases staining positive in some reported series [17]. The histological differential diagnosis usually includes low-grade sarcoma, benign fibroblastic proliferation, and reactive processes [2] [18]. Choosing optimal therapy for DTs is difficult because the diagnosis is rare, the anatomical presentations are varied, and no randomized and prospective trials for different treatment approaches are available. Complete resection with negative microscopic margins, when feasible, is the standard care for most mesenteric fibromatosis [2] [8]. As noted in our case the majority of these lesions require resection of the attached segment of the bowel [19]. Conservative management may be advocated over initial resection for patients with Gardner syndrome or with large slow growing and asymptomatic DT involving the mesentery or encasing vessels and organs [20]. Radiotherapy is an effective primary therapeutic option for patients who are not good surgical candidates, those who decline surgery, and those for whom surgical morbidity would be excessive. Adjuvant radiation therapy can be considered in patients with large tumors and positive margins [21]. In cases of tumors which have progressed and are no longer amenable to surgery or RT, systemic therapy using estrogen receptor antagonist tamoxifen, nonsteroidal anti-inflammatory drugs agent sulindac and chemotherapy with dactinomycin, vincristine and cyclophosphamide singly or in combination, have shown promising results in patients with mesenteric fibromatosis [22]-[24].

4. Conclusion

The diagnosis of mesenteric fibromatosis is difficult to establish preoperatively, especially in patients without a significant medical or surgical history. The optimal therapeutic strategy for these tumors should be evaluated by a multidisciplinary team because no randomized and prospective trials for different treatment approaches are available.

Disclosure

All the authors declare no conflict of interest.

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