Case Report

Desmoid-type Fibromatosis of the Splenic Hilum: A Rare Tumor at an Unusual Location

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Abstract

Desmoid-type fibromatoses (DTFs), also known as desmoid tumors, are benign but infiltrative neoplasms that often appear next to previous surgical site. Intra-abdominal tumors usually involve the mesentery, but splenic hilum is an unusual localization. We present a case of a desmoid tumor of the splenic hilum laparoscopically resected in a 70-year-old male with a previous history of chromophobe renal cell carcinoma and ocular spindle melanoma. Although benign, desmoid tumors might be infiltrative and produce serious complications. Treatment remains controversial, ranging from surgery and medical therapies to observation. Management of DTF must be individualized, considering the risk of complications and recurrence.

Keywords: *Desmoid tumor, desmoid-type fibromatosis, splenic hilum tumor*

Introduction

Desmoid-type fibromatoses (DTFs) or desmoid tumors are benign but infiltrative neoplasms arising from the fibroblast of muscular aponeurotic structures.[1] Although most DTFs are sporadic, 5%–10% are associated with familial adenomatous polyposis (FAP).[1,2] Sporadic DTF has a young female predominance, whereas PAF-associated DTF has no gender predilection.[2-4]

DTF clinical course is unpredictable, with a high incidence of recurrence despite R0 resections. No evidence-based approach for treatment is available, so therapeutic strategy should be individualized.[3]

We report the first case of sporadic DTF located at the splenic hilum treated by laparoscopic approach in a patient with a history of melanoma and renal cell carcinoma.

Case Report

A 70-year-old male, with a history of a chromophobe renal cell carcinoma and an ocular spindle melanoma, was referred to our consultation. Both neoplasms were surgically excised, staging as T2N0M0 and T2bN0M0, respectively. He did not receive any chemotherapy treatment.

During the follow-up, an enhanced computed tomography (CT) revealed a mass at the splenic hilum, which was progressively growing [Figure 1]. A fine-needle aspiration was performed to dismiss malignancy, but histopathological analysis was not determinant. However, neither DTF nor FAP was initially considered in differential diagnosis because renal and ocular tumors are not the typical neoplasms associated with FAP. A laparoscopy showed the presence of a well-defined tumor, adjacent to the splenic hilum but respecting splenic vessels, which was resected. The patient was discharged 24 h after surgery.

Histopathologic examination revealed a DTF arising from the perisplenic fatty tissue. Macroscopically, it was a 3.5 cm × 2.5 cm × 2 cm, well-defining, and nonencapsulated tumor [Figure 2]. Histological analysis showed a spindle cell tumor with moderate cellularity

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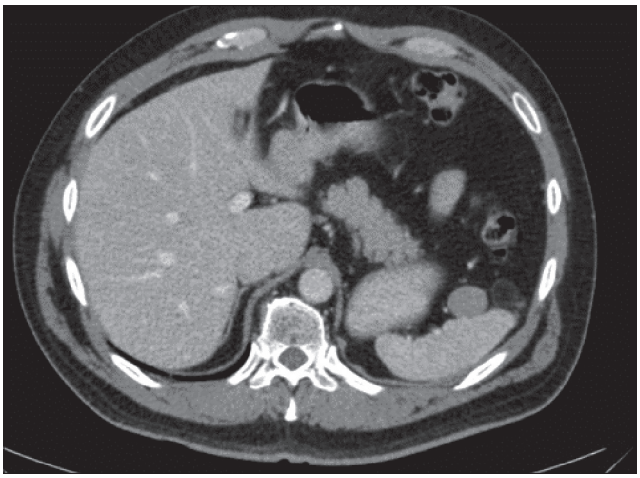

Figure 1: Enhanced computed tomography showing the tumor adjacent to splenic hilum

Figure 2: Macroscopic view of the tumor

and no atypical mitoses. Immunohistochemistry results were positive for β-catenin and CD10, whereas melanoma and chromophobe renal cell carcinoma markers were negative.

In view of this diagnosis and the patient's oncological history, endoscopic examinations were performed postoperatively. Colonoscopy findings ruled out the existence of FAP. He did not receive any postoperative medical treatment because the resection was complete. In fact, he is still followed up by surgeons and oncologists without evidence or recurrence.

Discussion

Fibromatosis can be categorized into superficial and DTF (also known as deep fibromatosis, aggressive fibromatosis, or desmoid tumor).[4] DTF is a rare fibroblastic neoplasm with an annual incidence of 3–4 cases/million.[2,3,5] It accounts for approximately 3% of soft-tissue tumors.[1,2] Most DTFs are sporadic, but 5%–10% are associated with polyposis syndromes.[1,3]

DTF can involve any part of the body. Depending on the localization, it can be categorized into extra-abdominal (50%–60%), abdominal wall (25%), and intra-abdominal fibromatosis (12%–15%).[4] Reported cases of DTF are usually into the abdominal cavity, arising from the mesentery or pancreatic tissue. To our knowledge, this is the second case at the splenic hilum and first described in a patient with oncological history.[5]

Different risk factors have been implicated in DTF etiology, including genetic mutations, female gender, or previous surgery next to the place where DTF occurs.[5] Estrogens are maybe implicated in the pathogenesis because DTF is more frequent in women, and spontaneous regression has been described after menopause or oophorectomy.[1,2,5] Genetically, sporadic desmoid tumors show a somatic mutation in β-catenin gene, leading to high intracellular levels and intranuclear localization of β-catenin.[3]

Clinically, DTF is characterized by infiltrative growing and a tendency toward local recurrence and inability to metastasize.[2,3,5,6] However, clinical behavior is uncertain, ranging from stabilization or regression to complications such as neurological compression and bowel or urinary obstruction.[2,5] Differential diagnosis is difficult because DTFs are not usually suspected unless there is a previous history of polyposis syndrome.[5] However, DTF should be suspected in patients with surgical history who develop a tumor next to previous surgical site.

Radiologically, DTF appears as a well-defined, solid, or solid-cystic soft-tissue tumor.[5] CT and magnetic resonance imaging (MRI) define the relationship between the tumor and the surrounding structures, but MRI gives more information about its behavior.[1]

Histologically, desmoid tumor are composed of spindle cells which infiltrate the surrounding tissues.[1] Immunohistochemical stains are positive for vimentin and nuclear β-catenin and negative for S100, CD117, and CD34. Immunostaining for β-catenin with nuclear positivity is helpful in establishing the diagnosis, but not pathognomonic [Figure 3].[6]

Nowadays, management of DTF is controversial and it should be individualized. Conventionally, complete surgical resection has been the aim in the treatment of DTF and it remains the ideal treatment when R0 resection is possible.[4] However, wide resections may lead to large tissue defects and functional impotence or even short-bowel syndrome. In that sense, many nonsurgical treatments have been tried to minimize

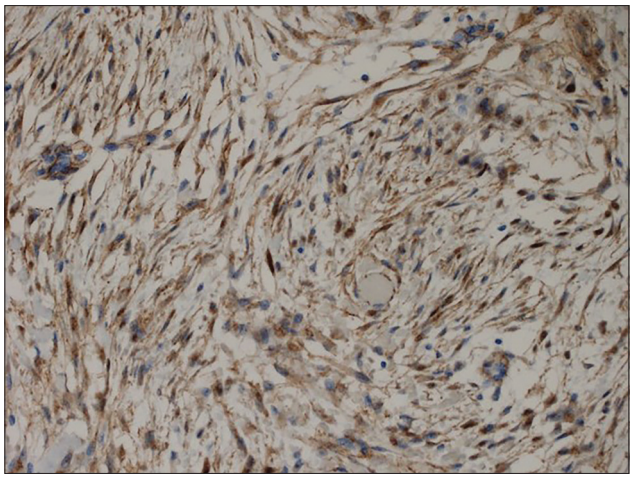

Figure 3: Immunohistochemical stain showing nuclear beta-catenin

surgical consequences, but the results are still not conclusive.

Among medical therapies, nonsteroidal anti-inflammatory drugs have been used as first-line treatment for early stages or asymptomatic advanced disease. They act blocking COX-2 protein which leads to the inactivation of *Wnt* oncogenic pathway, involved in DTF pathogenesis.[2] Antiestrogen drugs have also been tried in the management of DTF showing tumor regression. Clinical trials with imatinib and nilotinib may stabilize tumor progression.[6]

Conventionally, surgical treatment has been the gold standard of care for DTF, and tumor-free resection margin has classically been the aim of surgery.[1,3,7] However, large tumors require wide resections to achieve negative margins, leading to functional and cosmetic consequences with high rates of recurrence. The relationship between positive surgical margin and tumor recurrence has been widely analyzed. A meta-analysis of 16 reports including 1295 patients conclude that the risk of local recurrence was almost twofold higher for patients with microscopically positive resection margins.[7] In contrast, Lev *et al*. reported the outcomes of 189 surgical patients, concluding that margin status was not a significant risk factor for recurrence.[8,9] Due to the frequency of recurrence in DTF, several studies have analyzed the outcomes of wait-and-see strategy, showing that conservative management is safe and effective in selected cases. For example, Park *et al*. managed 20 patients with wait-and-see policy, having a 65% of tumor stabilization and 10% of regression.[10]

Radiotherapy (RT) has been tried as an adjuvant therapy in patients with positive resection margins and as single treatment in unresectable tumors. Gluck *et al*. analyzed the recurrence of 94 patients treated with surgery alone, surgery and RT, or RT alone, concluding that there were no differences.[11] RT alone is recommended for unresectable lesions.[8] The benefit of adjuvant RT is controversial, although several studies suggest that it might reduce the risk of recurrence after an incomplete surgical resection.[7,8,10]

Conclusion

DTF may be suspected in patients with polyposis syndromes and in those with tumors next to previous surgical site. Although radiological images can suggest the diagnosis, it will be confirmed by histological analysis. Management of DFT remains controversial due to the lack of conclusive results in clinical trials. Complete surgical excision is still the mainstay in treatment for resectable tumors; however, wait-and-see strategy should be considered in asymptomatic and stable lesions. RT can be an alternative for unresectable disease and as an adjuvant therapy after incomplete surgical resections.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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