Mesenteric desmoid tumor developing on the site of an excised gastrointestinal stromal tumor

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Abstract

We present a case of a rare and unusual occurrence of a desmoid tumor at the site of a resected gastrointestinal stromal tumor and mimicking a recurrence, with a brief discussion of the management of desmoid tumors.

Introduction

Desmoid tumors, also known as deep or aggressive fibromatoses, are rare with an annual incidence of two to four cases per million.1 The term desmoid, coined by Muller in 1838, is derived from the Greek word desmos, which means tendon-like.2 They represent low-grade mesenchymal neoplasms, originating from musculo-aponeurotic stromal elements. They do not metastasize but tend to exhibit a high degree of local infiltration and invasion, thus becoming lethal in some cases, depending on their anatomical localization.3 Desmoid tumor development has been associated with genetic predisposition in patients with familial adenomatous polyposis (FAP)4 and previous trauma of the area.5 Dysregulation of the Wnt signaling pathway, which is involved in the pathogenesis of FAP and also in the process of wound healing, may be a critical underlying molecular mechanism in FAP-associated cases and possibly in sporadic ones as well.6 In one molecular mechanism in FAP-associated cases adenomatous polyposis (FAP)4 and previous pathogenesis of FAP and also in the process of genetic predisposition in patients with familial estrogenic influence; nevertheless, relative data are observational.

We report here a rare and unusual case of desmoid tumor developing at the site of a gastrointestinal stromal tumor (GIST) resection. This created the impression of GIST recurrence. Surgical excision of the lesion was a difficult decision owing to the suspicion of metastatic disease. We present a discussion on the basis of this case for the management of both desmoid tumors and GIST.

Case Report

A 37-year-old IT engineer with an otherwise unremarkable medical history was admitted to the acute assessment unit with acute upper gastrointestinal bleeding. An ultrasound scan of the abdomen performed as part of the work-up for this condition revealed a large gastric mass and liver lesions consistent with metastases. Contrast-enhanced computed tomography (CT) of the abdomen showed a 15 cm exophytic mass originating from the gastric wall (Figure 1A) and four enhancing liver lesions of 19 mm maximum diameter (Figure 1C). Endoscopy revealed a single gastric fundal mass with a large ulcer that was biopsied. Histological diagnosis was of a GIST, staining positively for C-KIT and CD34 and with a high mitotic rate. PET imaging showed a markedly increased uptake in the gastric mass (SUV 15). The larger of the imaged liver lesions did not show any significant FDG uptake while the smaller lesions were not assessable.

Treatment with imatinib mesylate (400 mg daily) was initiated, and the first assessment CT scan performed three months later showed a partial response of the gastric tumor (Figure 1B), according to the RECIST (Response Evaluation Criteria in Solid Tumors) criteria.7 In addition, the liver lesions were reduced in size uniformly. This treatment was continued and further scans over the next five months showed stable disease. Subsequent scans were done as magnetic resonance imaging (MRI) to assess the extent of the hepatic lesions, from which it was concluded that they were not metastatic in nature. In light of a good response to treatment the patient underwent a total abdominal gastrectomy. The procedure achieved excision of the tumor together with an intraoperatively detected omental secondary lesion (Figures 1C). Pathological findings reported positive margins (microscopic-R1 resection) at the splenic and pancreatic aspects. Imatinib was continued postoperatively. Plans for surgical removal of the liver lesions were abandoned when the MRI scan of the liver revealed multiple nonspecific tiny lesions widespread throughout the liver parenchyma.

The patient remained asymptomatic and well on imatinib treatment for another 11 months, when a repeat MRI scan of the abdomen revealed a new mesenteric mass 3 cm in diameter (Figure 1D), while the appearance of the liver lesions remained unchanged. A multidisciplinary team meeting decision was made to resect the lesion surgically. The excised lesion represented a firm nodular mass with a thin connective tissue capsule, measuring 6.0 x 4.0 x 4.5 cm (Figure 2). A mesenteric and an anterior abdominal wall nodule were also removed. Microscopically the larger mass appeared as a circumscribed unencapsulated tumor of varying cellularity. Toward the periphery it was composed of spindle cells with elongated nuclei and a small amount of pale eosinophilic cytoplasm arranged between broad sweeping fascicles (Figure 3A). No nuclear atypia was noted. Toward the center of the lesion there was pronounced keloidal collagen deposition (Figure 3B). The tumor showed weak staining for desmin and was negative for CD34 and C-KIT. The pattern was typical of an intra-abdominal desmoid tumor. The other two nodules turned out to be a mesenteric lymph node with sinus histiocytosis and a pseudocyst with no evidence of malignancy. The patient continued on imatinib (400 mg OD) treatment and follow-up MRI scans did not show any recurrence of the desmoid tumor over the next eight months.

Discussion

Desmoid tumors usually manifest as slow-growing, deep-seated, painless or slightly painful masses and can develop at virtually any anatomical site.8 Typically three localizations are described: trunk/leg/extremity, abdominal wall, and intra-abdominal region. Usually FAP-associated cases occur in the abdomen, while non-FAP-associated cases present in the shoulder or hip regions and in the extremities.9 They can be multifocal on an extremity, but different anatomical regions rarely are affected in the same patient. Inside the abdomen they can...
cause changes in bowel habits, pain, obstruction, ischemia, rectal bleeding, or a dysfunctional anastomosis, and are a significant cause of mortality for FAP patients. Non-intra-abdominal desmoids have a better prognosis.7,12 Complete surgical resection remains the cornerstone of management of desmoid tumors, while unresectable or residual disease can be treated with radical radiotherapy.11 Depending on tumor size, type of treatment, and negative margins post-resection, recurrences occur in up to 45% of treated adult cases, usually within three years from diagnosis.13 Systemic treatments with NSAIDs, anti-estrogens or androgens, chemotherapy (doxorubicin-based, methotrexate, vinblastine, vinorelbine), and lately imatinib have been used for unresectable or relapsed desmoids, often resulting in long-lasting responses.14 In selected asymptomatic patients, a period of watchful waiting is recommended.15 Prior trauma is identified in 30% of patients who develop desmoid tumors, and is typically abdominal surgery for FAP.4 In addition, sporadic cases of intra-abdominal desmoid tumors have been observed in sites of previous abdominal surgery.2,3,4,5,6,7,8,9,11 Our literature search yielded one previous report of a desmoid tumor arising in the site of a previously excised GIST.18 GISTs and desmoid tumors share a common stromal origin but are diverse histologically, genetically, and biologically. Nevertheless, surgical trauma at the GIST excision site may predispose to the development of the desmoid tumor. An accurate diagnosis is possible only after surgical removal and pathological examination, as there are no typical imaging findings to suggest a desmoid tumor. Excluding recurrence of the GIST was crucial for the further management of our patient; he remained on imatinib for the metastatic GIST; therefore he continued to benefit from first-line treatment and remains progression-free after eight months.

References


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