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, 6 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. 7 In one third of sporadic cases, chromosomal changes such as trisomy-20 or -8 have been identified. 8 An association with the female gender and pregnancy 9 suggests a pathogenic role for 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. 9, 10 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 through 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 appear-

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Discussion

Desmoid tumors usually manifest as slow-growing, deep-seated, painless or slightly painful masses and can develop at virtually any anatomic site. 11 Typically three localizations are described: trunk/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. 12 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.\textsuperscript{7,12} Complete surgical resection remains the cornerstone of management of desmoid tumors, while unresectable or residual disease can be treated with radical radiotherapy.\textsuperscript{13} 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.\textsuperscript{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.\textsuperscript{14} In selected asymptomatic patients, a period of watchful waiting is recommended.\textsuperscript{15}

Prior trauma is identified in 30% of patients who develop desmoid tumors, and is typically abdominal surgery for FAP.\textsuperscript{4} In addition, sporadic cases of intra-abdominal desmoid tumors have been observed in sites of previous abdominal surgery.\textsuperscript{2,11,16} Our literature search yielded one previous report of a desmoid tumor arising in the site of a previously excised GIST.\textsuperscript{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

1. Reitamo JJ, Häyry P, Nykyri E, Saxén E. The desmoid tumor. Incidence, sex, age and anatomical distribution in the Finnish population. Am J Clin Pathol 1982;77:665-73.
2. Müller J. Veber den Feinern Bau und die Formen der Krankhaften Geschwulste. Berlin: G Reimer, p 80, 1838.
3. Schlemmer M. Desmoid tumors and deep fibromatoses. Hematol Oncol Clin North Am 2005;19:565-71.
4. Soravia C, Berk T, McLeod RS, Cohen Z. Desmoid disease in patients with familial adenomatous polyposis. Dis Colon Rectum 2000;43:363-9.
5. Dei Tos A P, Dal Cin P. The role of cytogenetics in the management of desmoid tumors. Hematol Oncol Clin North Am 2002;16:1623-43.
6. De Cian F, Delay E, Rudigox RC, et al. Desmoid tumor arising in a Cesarean section scar during pregnancy: monitoring and management. Gynecol Oncol 1999;75:145-8.
7. Penna C, Tiret E, Parc R, et al. Operation and abdominal desmoid tumors in familial adenomatous polyposis. Surg Gynecol Obstet 1995;177:263-8.
8. Dei Tos AP, Dal Cin P. The role of cytogenetics in the management of desmoid tumors. Hematol Oncol Clin North Am 2002;16:1623-43.
9. Müller J. Veber den Feinern Bau und die Formen der Krankhaften Geschwulste. Berlin: G Reimer, p 80, 1838.
10. Schlemmer M. Desmoid tumors and deep fibromatoses. Hematol Oncol Clin North Am 2005;19:565-71.

Case Report

Figure 1. A contrast-enhanced computed tomography scan of the abdomen showing: a 15-cm exophytic mass originating from the gastric wall (arrow) (A); and four enhancing liver lesions of maximal diameter of 19 mm (C). A three-month follow-up abdominal CT scan (B) demonstrated a partial response to treatment of the gastric tumor (arrow). A repeat MRI scan of the abdomen after 11 months revealed a secondary mesenteric omental mass, 3 cm in diameter (D).

Figure 2. Macroscopic view of the secondary mesenteric lesion, showing it as a firm nodular mass with a thin connective tissue capsule, measuring 6.0×4.0×4.5 cm.

Figure 3. A histological section of the larger mass illustrating: (A) toward the periphery of the tumor, spindle cells with elongated nuclei and a small amount of pale eosinophilic cytoplasm, and (B) pronounced keloidal collagen deposition toward the center of the lesion. No nuclear atypia was noted. (Hematoxylin and cosin stain; magnification 100X).
9. Gansar GF, Markowitz IP, Cerise EJ. Thirty years of experience with desmoid tumors at Charity Hospital. Am Surg 1987;53:318-9.

10. Therasse P, Arbuck SG, Eisenhauer EA, et al. New guidelines to evaluate the response to treatment in solid tumors. J Natl Cancer Inst 2000;92:205-16.

11. Ballo MT, Zagars GZ, Pollack A, et al. Desmoid Tumor: Prognostic Factors and Outcome After Surgery, Radiation Therapy, or Combined Surgery and Radiation Therapy. J Clin Oncol 1999;17:158-67.

12. Enzinger FM, Weiss SW. Soft Tissue Tumors. 3rd ed. St Louis: Mosby-Yearbook Inc., p 201, 1995.

13. Nuyttens JJ, Rust PF, Thomas CR Jr, et al. Surgery versus radiation therapy for patients with aggressive fibromatosis or desmoid tumors: a comparative review of 22 articles. Cancer 2000;88:1517-23.

14. Patel SR, Benjamin RS. Desmoid Tumors Respond to Chemotherapy: Defying the Dogma in Oncology. J Clin Oncol 2006;24:11-2.

15. De Bree E, Keus R, Melissas J, et al. Desmoid tumours: Need for an individualized approach. Expert Rev Anticancer Ther 2009;9:525-35.

16. Kersting S, Herbst H, Senninger N, Mittelkötter U. Intra-abdominal fibromatosis after appendectomy as cause for ileus. Zentralbl Chir 2004;129:317-20.

17. Moudouni SM, Tazi Mokha K, Nouri M, et al. Desmoid tumor of the mesentery secondary to colectomy. An exceptional cause of ureteric obstruction. Ann Urol 1999;33:424-7.

18. Vendrell J-F, Mazars R, Funakoshi N, et al. Desmoid tumor subsequent to resection of a gastrointestinal stromal tumor. Eur J Radiol 2008;65:9-11.