CLINCAL CASE

UNUSUAL CASE OF MESENTERIC FIBROMATOSIS MASQUERADING AS AN APPENDICULAR ABSCESS

Kawari Sowbhagyalaxmi[1], Rajat Choudhari[1], Rahul Bhat[1], Shivananda Prabhu[1], Poornachandra Thejeswi[1]

[1]Department of General Surgery, Kasturba Medical College, Mangalore, Manipal Academy of Higher Education (MAHE), Manipal, Karnataka, India

Corresponding author: Rajat Choudhari
E-mail: rajatc10@gmail.com

Abstract

Desmoids are rare tumors of fibroblast origin and comprise about 0.03% of all neoplasms. These are benign tumors arising from soft tissues and classified based on location - abdominal wall, intra and extra abdominal. In the abdomen, mesentery is the commonest location, and they are termed as mesenteric fibromatosis (MF). A 38-year-old male presented with fever, abdominal pain with tenderness and rebound tenderness localized to the right iliac fossa and was diagnosed as appendicular abscess. He was initially treated with percutaneous drainage and antibiotics and underwent exploratory laparotomy which revealed a mass at the ileocecal junction. Histopathology showed spindle cells and immunohistochemistry confirmed a diagnosis of mesenteric fibromatosis. These are slow growing tumors and present with nonspecific symptoms such as pain abdomen, constipation, appetite and weight loss, fever, abdominal distension and ureteral obstruction. Diagnosis is confirmed only after immunohistochemistry. Multimodal approach for treatment is followed with surgery being the primary treatment and Radiation therapy (RT), chemotherapy, anti-estrogenic therapy, NSAIDs and tyrosine kinase inhibitors (TKIs) used adjunctively. Mesenteric fibromatosis is a rare condition with varied presentation and if diagnosed early, has a good prognosis with few complications due to variety of available treatment options.

Keywords*: desmoid tumors, mesenteric fibromatosis, beta catenin, appendicular abscess*

Introduction

Desmoids are uncommon tumors of fibroblast origin and comprise about 0.03% of all neoplasms [1]. These are benign tumors which can arise from various soft tissues of the body and are segregated based on location - abdominal wall, intra and extra abdominal. In the abdomen, mesentery is the commonest location and they are termed as mesenteric fibromatosis (MF) [2], [3]. They are usually asymptomatic when small in size and are detected incidentally or with symptoms on increase to a large size. The presentation of MF is that of a silently growing mass with pain, loss of appetite and weight, fever, urinary retention or bowel symptoms [4]–[6].

In this report, we outline an unusual presentation with symptoms, clinical findings and imaging suggestive of an appendicular abscess and finally diagnosed to have a tumor at laparotomy, which was confirmed to be mesenteric fibromatosis on histopathology.

Case presentation

A 38-year-old male reported to our hospital with fever, pain in right lower abdomen and constipation of 3 days duration with tenderness and rebound tenderness localized to the right iliac fossa. He underwent initial evaluation and was found to have an elevated total white cell count of 18,300 cells/mm^3. Ultrasonogram of the abdomen showed a collection in the right iliac fossa and he underwent a contrast enhanced computerized tomogram (CT) scan which revealed a 12 cm x 13 cm x 19 cm peripherally enhancing collection, with air fluid levels and air foci within, in the right iliac and lumbar area abutting the terminal ileum and ascending colon with non-visualization of appendix (Figure 1).

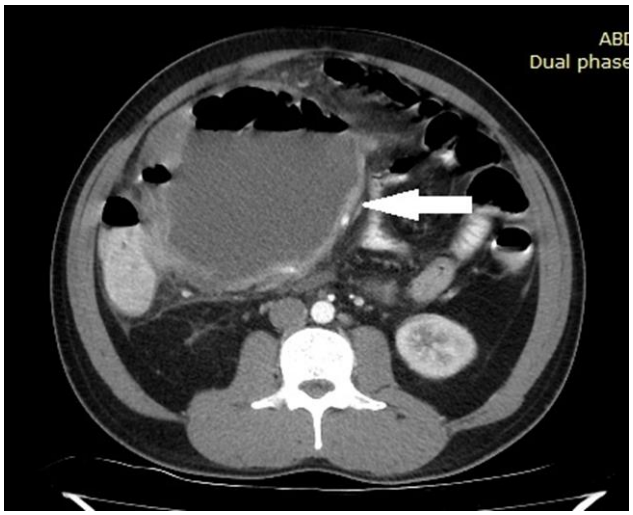

Figure 1 – Venous phase axial section showing large peripherally enhancing collection abutting the ascending colon

He was suspected to have an appendicular abscess and was managed with percutaneous drainage and antibiotics. The fluid was sent for culture which grew Escherichia coli, and he was treated with appropriate antibiotics. Due to suspicion of underlying malignancy, the drain fluid was evaluated for carcino-embryonic antigen (CEA) level which was 0.7 ng/dL and cytopathological examination which turned out to be negative for malignant cells. Due to persistent symptoms and non-resolution of the collection on repeat scans, he was taken up for exploratory laparotomy and was found to have a7 cm x 5 cm tumor (Figure 2) at the medial mesenteric border of the caecum with normal appendix, ileum and ascending colon and 15 cm x 15 cm abscess cavity adjacent to the mass with hemorrhagic content (infected hematoma). He underwent a right hemicolectomy and drainage of the abscess. Post operatively, his recovery was uneventful, and he was discharged on normal oral diet on post-operative day 7.

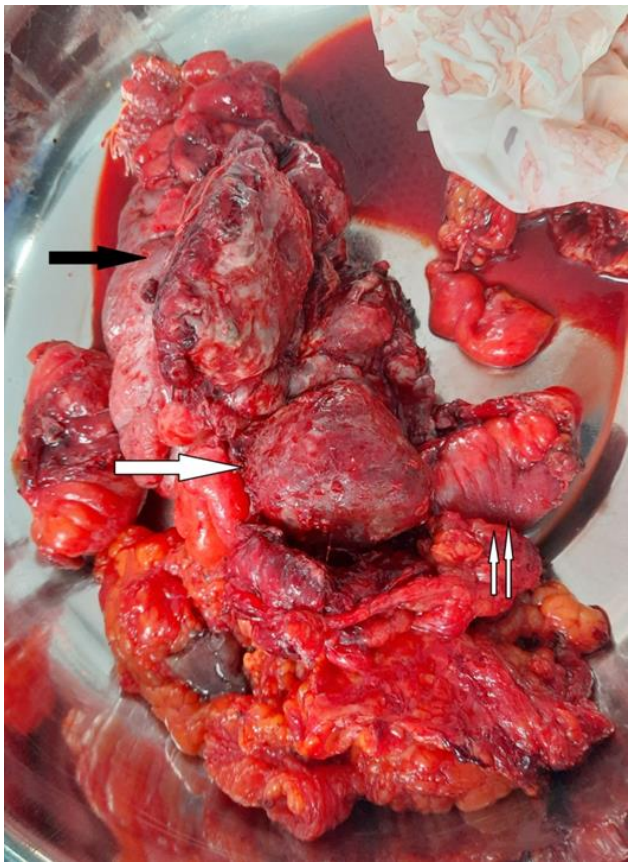

Figure 2 – Surgical specimen showing ascending colon (black arrow), ileum (double white arrow) and tumor at the IC junction (single white arrow)

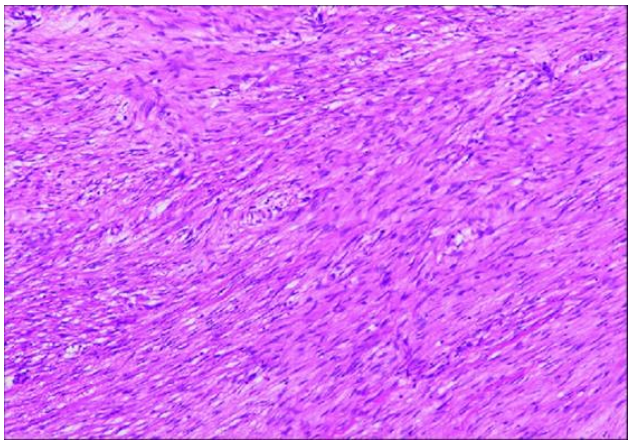

Figure 3 – Histopathology image showing spindle shaped cells containing Eosinophilic cytoplasm, bland nuclei, low mitotic activity and abundant inflammatory infiltrates

On Histopathological examination, grossly the tumor was found to be involving only the mesenteric border of the bowel and mesentery

with normal lumen inside. The mass was solid and firm with whorled appearance on cut section. On microscopy, the tumor was composed of spindle shaped cells containing Eosinophilic cytoplasm, bland nuclei, low mitotic activity and abundant inflammatory infiltrates. On Immunohistochemistry, the cells showed positivity only for beta catenin whereas DOG-1, CD34, CD117, S100, SMA, ALK-1 and Bcl-2 were all negative. Hence, a final diagnosis of mesenteric fibromatosis was arrived at.

Discussions

Desmoids are benign tumors arising from clonal myofibroblast proliferation and divided into extra and intra-abdominal. In the abdomen, these can arise from various organs with mesenteric origin being the most common. Although these are rare tumors, mesenteric fibromatosis is the commonest mesenteric tumor [7]. These tumors can arise de novo, secondary to trauma or in association with FAP, where they appear at the site of bowel resection [7]. The pathogenesis involves CTNNB1 and APC gene mutation which are involved in signaling for cellular proliferation [8].

Koh et al. identified various courses that these tumors can present with in patients with FAP and categorized them into - spontaneous regression, static, variable growth, progressive growth and aggressive growth[9].These are slow growing tumors and present with nonspecific symptoms such as pain abdomen, constipation, appetite and weight loss, fever, abdominal distension and ureteral obstruction[6].However, gastrointestinal bleeding or bowel obstruction is rarely seen even in large sized tumors[8].They can present as large abdominal mass with erosion into bowel leading to fistulization and intra-abdominal abscess formation[3].

Mesenteric fibromatosis does not have any distinct radiological features and appear as solid soft tissue masses which resemble soft tissue sarcoma, GIST or lymphomas. They can have homogenous or heterogenous enhancement, solid or solid-cystic areas and with or without involvement of bowel loops. Although both CT and MRI are routinely accepted, MRI is the investigation of choice due to more accurate characterization of soft tissue lesion with treatment planning and follow up [10]. Mesenteric desmoid tumors have also been reported to present as a fat containing cystic mass by tan et al [11].

Differential diagnoses of these tumors include inflammatory myofibroblastic tumors, retroperitoneal tumors or GIST. GIST is the most common differential and should be ruled out as the treatment differs significantly. Grossly, these tumors appear as large masses with irregular margins and may involve or infiltrate multiple bowel loops. However, lack of hemorrhagic foci or cystic degeneration areas helps differentiate them from gastrointestinal stromal tumors (GIST) [7].

Microscopically, they appear to be composed of spindle or stellate cells with abundant collagen and inflammatory cells with low mitotic index [7]. The absence of atypical mitoses, abundant inflammatory cells and immunohistochemistry rules out GIST [10]. On immunohistochemistry, these tumors are negative for DOG-1, S100, CD117 and CD34 which differentiates them from GIST. They show positivity forβ –catenin and actin but less commonly for desmin. They are also negative for estrogen and progesterone receptors but can show occasional positivity in around 10-15% cases [7], [10].

The Collaborative Group of the Americas for Inherited Colorectal Cancers (CGA-ICC) has established a staging system for intra-abdominal desmoids in patients with FAP (Table 1) [12]. Although the study was done among FAP associated desmoids, it seems to be applicable to sporadic tumors also as the management for these is similar.

There are no defined guidelines for management of fibromatosis and various forms of treatment modalities have been tried and compared. Surgery in form of wide local excision is the cornerstone of treatment for operable cases as these are locally aggressive tumors. Radiation therapy (RT), chemotherapy, anti-estrogenic therapy, NSAIDs and tyrosine kinase inhibitors (TKIs) have all been evaluated [2], [7], [13]. RT has been shown to be effective in reducing recurrence rates when used in adjuvant setting after resection of the tumor. Also, in inoperable cases due to involvement of mesenteric root or entire bowel length where surgery is not feasible, neoadjuvant RT can be used to downsize the tumor before surgical resection [2], [14].

Stage	Characteristics of tumor	Management
I	Asymptomatic, not growing, incidentally detected	Observation
II	Mild Symptoms, ≤10 cm, not growing	Surgical resection
III	Moderate Symptoms, 11-20 cm or asymptomatic and growing	NSAIDS, tamoxifen, chemotherapy, Surgery
IV	Severely symptomatic, >20cm, rapidly growing or complications such as perforation, abscess	Emergent surgery, adjuvant radiation

Table 1 – Desmoid tumors Staging system (CGA-ICC)

Among the chemotherapeutic agents, pegylated liposomal doxorubicin is the least toxic but has a limited role in management and is not widely used. NSAIDs like Sulindac have been shown to cause gradual reduction in tumor size used in combination with tamoxifen [2], [15]. Tyrosine kinase inhibitors Imatinib, sunitinib, pazopanib and sorafenib have been studied and shown use in inoperable tumors for downsizing the tumors and to hinder progression[2]. Overall, the management of operable tumors entails wide local excision followed by adjuvant RT for local control, neoadjuvant RT for downsizing borderline tumors and lifelong hormone and NSAID therapy for inoperable tumors.

Our case is peculiar in the sense that although the tumor itself was relatively small in size, possible hemorrhage from it and subsequent secondary infection of the hematoma brought the patient to the hospital with symptoms and signs resembling an intra-abdominal abscess. However, high index of suspicion for underlying malignancy lead to early surgery and detection of tumor. We also observed intra-operatively that the transverse colon did not contain any polyps, and none were found in the surgical specimen at pathology. Considering these findings and age of the patient, FAP was clinically ruled out and we did not advise routine colonoscopy for follow up.

Conclusion

Mesenteric fibromatosis is a rare condition with varied presentation and can be often misdiagnosed and mistreated. Keen observation regarding presentation and origin of tumor intra-operatively can guide the clinician to suspicion and pathological confirmation with immunohistochemistry can clinch the diagnosis. If diagnosed early, it has a good prognosis with few complications due to variety of available treatment options.

References

[1] K. Hayashi et al., “A mesenteric desmoid tumor with rapid progression,” Intern. Med., vol. 56, no. 5, pp. 505–508, 2017.

[2] T. Desmoid Tumor Working Group et al., “The management of desmoid tumours: A joint global consensus-based guideline approach for adult and paediatric patients The Desmoid Tumor Working Group 1,” Eur. J. Cancer, vol. 127, pp. 96–107, 2020.

[3] K. Huang, H. Stuart, K. Lyapichev, A. E. Rosenberg, and A. S. Livingstone, “Mesenteric desmoid tumour presenting with recurrent abdominal abscess and duodenal fistula: A case report and review of literature,” Int. J. Surg. Case Rep., vol. 37, pp. 119–123, Jan. 2017.

[4] M. D, “Mesenteric Fibromatosis (Desmoid Tumour) - A Rare Case Report,” J. Clin. DIAGNOSTIC Res., vol. 8, no. 11, p. ND01, 2014.

[5] H. Ji et al., “A giant mesenteric fibromatosis involving the muscular layer of the colon wall,” Medicine (Baltimore)., vol. 98, no. 1, p. e14015, Jan. 2019

[6] P. Li, Z. Zhang, S. Qin, and J. Li, “The diagnosis of mesenteric fibromatosis: A 90-month five patients case report,” J. Cancer Res. Ther., vol. 12, no. 4, p. 1318, Oct. 2016.

[7] P. Chaudhary, “Mesenteric fibromatosis,” International Journal of Colorectal Disease, vol. 29, no. 12. Springer Verlag, pp. 1445–1451, Aug. 21, 2014.

[8] M. Abate and A. Pigazzi, “Mesenteric fibromatosis in a patient with a history of neuroblastoma: a case report,” J. Surg. Case Reports, vol. 2018, no. 9, pp. 1–3, Sep. 2018

[9] P. K. Koh et al., “Mesenteric desmoid tumors in singapore familial adenomatous polyposis patients: Clinical course and genetic profile in a predominantly chinese population,” Diseases of the Colon and Rectum, vol. 50, no. 1. Springer New York, pp. 75–82, Jan. 01, 2007.

[10] M. Wronski, B. Ziarkiewicz-Wroblewska, M. Slodkowski, W. Cebulski, B. Gornicka, and I. W. Krasnodebski, "Mesenteric fibromatosis with intestinal involvement mimicking a gastrointestinal stromal tumour," Radiol. Oncol., vol. 45, no. 1, pp. 59–63, Mar. 2011.

[11] C. H. Tan, U. Pua, K. H. Liau, and H. Y. Lee, "Mesenteric desmoid tumour masquerading as a fat-containing cystic mass," Br. J. Radiol., vol. 83, no. 994, p. e200, Oct. 2010

[12] J. Church et al., "Staging Intra-Abdominal Desmoid Tumors in Familial Adenomatous Polyposis: A Search for a Uniform Approach to a Troubling Disease," Dis. Colon Rectum, vol. 48, no. 8, pp. 1528–1534, Aug. 2005

[13] G. H. Sakorafas, C. Nissotakis, and G. Peros, "Abdominal desmoid tumors," Surgical Oncology, vol. 16, no. 2. Elsevier, pp. 131–142, Aug. 01, 2007

[14] S. Tolan, J. H. Shanks, M. Y. Loh, B. Taylor, and J. P. Wylie, "Fibromatosis: Benign by Name but not Necessarily by Nature," Clin. Oncol., vol. 19, no. 5, pp. 319–326, Jun. 2007

[15] A. Hansmann, C. Adolph, T. Vogel, A. Unger, and G. Moeslein, "High-Dose Tamoxifen and Sulindac as First-Line Treatment for Desmoid Tumors," Cancer, vol. 100, no. 3, pp. 612–620, Feb. 2004