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Intestinal obstruction caused by desmoid tumours: a review of the literature

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Lesson

Intraabdominal desmoid tumours are rare and can cause intestinal obstruction. Based on the review of the literature, surgical resection with negative margins and adjuvant chemotherapy is the optimal strategy for treatment of this pathology.

Keywords

Desmoid tumours, caecum, colon

Introduction

Desmoid tumours are extremely rare, causing 0.03% of all neoplasms and less than 3% of all soft tissue tumours.[1] They are locally aggressive neoplasias, which do not show metastatic tendency. While most cases have an asymptomatic course, urgent surgical interventions have been reported due to reasons such as intestinal obstruction, perforation and abscess formation.[2] We present the case of a 47-year-old male who developed an intestinal obstruction from a desmoid tumour of the caecum.

Case report

A 47-year-old male with a past medical history of coronary artery disease, hypertension, end-stage renal failure and exploratory laparotomy for a gunshot wound with subsequent ventral hernia repair presented with one month of abdominal pain. The patient characterised the pain as intermittent and localised to the right lower quadrant. A computed tomography scan Computed tomography scan of the abdomen and pelvis showed a 7.7×10 cm caecal mass along with partially obstructed loops of small bowel (Figure 1). Intraoperatively he was found to have a large mass in the caecum extending up into the right colon to which the distal ileum and appendix were adherent. He also had palpable enlarged lymph nodes extending down to the root of the right mesocolon. He was also found to have thickened and dilated distal small bowel loops consistent with long-standing obstruction. An ileocolic anastomosis was successfully created and the patient recovered uneventfully. Pathology revealed a $10 \times 9 \times 5$ cm desmoid tumour which stained positive for beta-catenin (Figures 2 and 3). The margins were negative as were the seven resected lymph nodes. He was discharged home on postoperative day 7 and continues to do well on outpatient follow-up.

Discussion

Desmoid tumours are rare well-differentiated and aggressive musculoaponeurotic fibromatosis tumours, considered as grade 1 fibrosarcoma.[3] These tumours are characterised by their propensity for slow, incessant growth and invasion of contiguous structures. Although locally aggressive, these tumours do not metastasise.[4] On computed tomography scan, most desmoid tumours appear as well-circumscribed homogeneous masses that may be isodense or hyperdense relative to muscle.[5] Histological assessment is mandatory for differentiating desmoid tumours from other neoplasms such as gastrointestinal stromal tumours, lymphoma, pleomorphic sarcoma and fibrosarcoma. Microscopically they are composed of spindle- or stellate-shaped fibroblastic cells embedded in a collagenous stroma. The spindle cells usually stain for vimentin and smooth muscle actin and nuclear beta-catenin.[6,7]

Depending upon the location, most desmoids present as slow-growing, painless masses.[8] Mesenteric fibromatosis commonly arises from the mesentery of the small bowel but can also originate from the ileocolic mesentery, gastrocolic ligament and omentum.[9] Intraabdominal desmoids are usually asymptomatic until their size causes compression of surrounding viscera. This compression can lead to intestinal obstruction, ischaemic bowel secondary to vascular compression and hydronephrosis due to ureteric compression.[10] Although intestinal obstruction has been described as a potential complication of intraabdominal desmoid tumours, only seven prior cases have been reported in the literature (Table 1).

Figure 1. Abdominal tomography image demonstrating the large mass originating in the caecum.

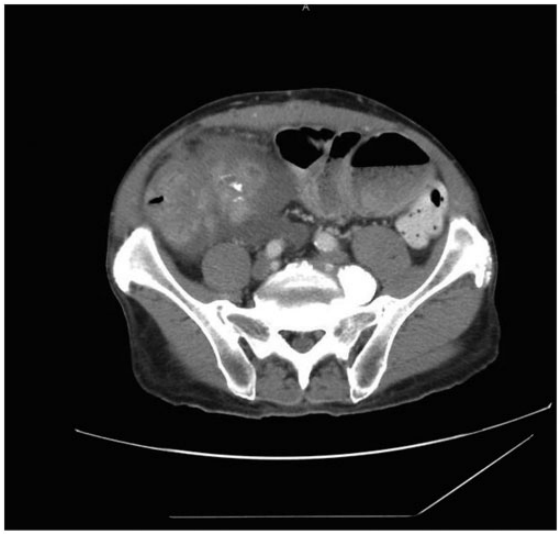
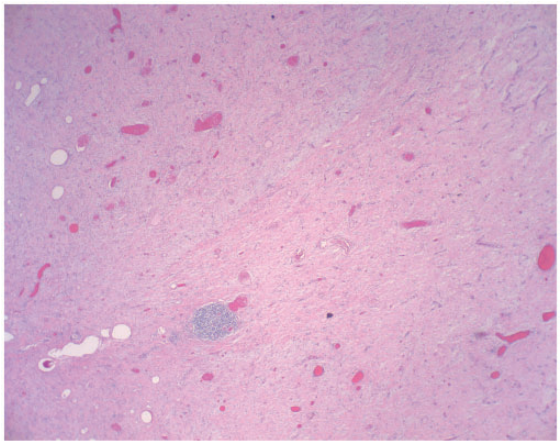
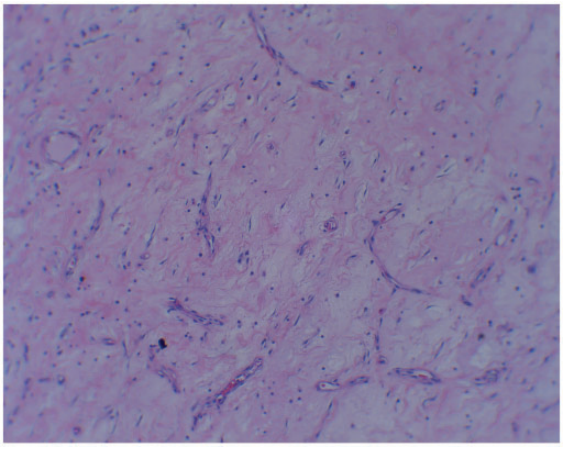


Figure 2. Histological slide of specimen – different staining specimen.



Most of these tumours occur sporadically; however, patients with Gardner's syndrome are at higher risk than others. The incidence of abdominal wall and mesenteric desmoids in patients with Gardner's syndrome ranges between 4 and 29%, and the tumours typically occur after abdominal surgery.[14] In the presence of polyposis syndromes, patients should be managed at a specialist colorectal unit with surgery reserved only when absolutely necessary. In these patients the high recurrence rate mandates medical therapies such as non-steroidal anti-inflammatory drugs, tamoxifen and chemotherapy to be used as first- and second-line therapy.

Figure 3. Histological slide of specimen – collagen staining.



This contrasts with the management of sporadic intraabdominal desmoid tumours, which should be managed in a specialist sarcoma unit by a multidisciplinary team.[6] Early referral to a centre that specialises in multimodality care of sarcomas is warranted. In asymptomatic patients, close observation is often the preferred strategy. Patients with symptoms are typically treated, given the inevitably progressive growth of desmoid tumours. Surgery with a wide margin of resection is the preferred treatment whenever feasible.[15] Patients with sporadic desmoid tumours tend to have low recurrence rates after resection.[6] All of the patients who presented with obstruction underwent oncologic resections of the involved intestinal segments with no reported recurrences.[3,6,8,9,11–13]

Complete resection of the tumour with negative microscopic margins is the standard surgical goal but is often constrained by anatomic boundaries.[10] One recent review of multimodal therapy for desmoid tumours in all anatomic locations showed that surgery supplemented with radiotherapy had reduced recurrence rates for patients with both positive and negative surgical margins. Radiotherapy alone also had promising outcomes for local control.[16] For patients with recurrence despite local therapy, systemic medical therapy is often prescribed. Options include tamoxifen, which is thought to suppress desmoid growth due to the presence of estrogen receptor beta on tumour cells, non-steroidal anti-inflammatory drugs such as Sulindac, and doxorubicin and methotrexate with vinca alkaloid-based chemotherapy. Post-treatment surveillance includes clinical examination and radiographic assessment every six months for at least three years and then yearly thereafter.[8]

Table 1. Cases of intraabdominal desmoid tumours causing intestinal obstruction.

Author	Year	Age	Presentation	Operation	Pathology
Abdalla et al.[6]	2016	54M	Altered mental status with abdominal pain	Laparotomy with ileal and ileocaecal resections Planned second look with jejunoileal and ileocolic anastamoses Laparotomy and drain placement for jejuno-ileal anastomotic leak	7 cm mass at ileum + 6 cm mesoappendix mass +Beta-catenin +Actin +Vimentin
Venkat et al.[8]	2010	46F	Abdominal pain	Laparotomy with right hemicolectomy	6.2 cm mass of the right colon +Beta-catenin +Desmin
Lasseur et al.[3]	2016	32F	Incidentally discovered abdominal mass	Laparotomy with en bloc resection of the distal duodenum and proximal jejunum and superior mesenteric artery and vein Laparotomy for hemiperitoneum	7.5 cm mass of the small bowel at the ligament of Treitz +Beta-catenin
Aggarwal et al.[11]	2015	28F	Early satiety	Laparotomy with resection of left internal oblique muscle with mesh repair, resection of right external iliac vein	Large mass adherent to anterior rectus sheath and parietal peritoneum
Mazeh et al.[9]	2006	55M	Early satiety	Laparotomy with gastrojejunostomy	20 cm duodenal mass
Ozmen et al.[12]	2004	57M	Abdominal pain	Laparotomy with partial duodenojejunectomy and right hemicolectomy	10 cm tumour at jejunum, proximal duodenum and ascending colon +Actin +Vimentin
Holubar et al.[13]	2006	52M	Abdominal pain	Laparotomy with en bloc resection of the terminal ileum and superior mesenteric artery and vein	22 cm mass at terminal ileum +Beta-catenin

Conclusion

Intraabdominal desmoid tumours are rare and may result in intestinal obstruction. Based on the review of the literature, resection with negative margins with consideration towards adjuvant radiotherapy is the optimal treatment. In patients with obstruction, oncologic bowel resection with primary anastomosis appears to be an acceptable surgical strategy.

Declarations

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Ethical approval: Given that this was a retrospective chart review, no formal IRB approval was required. Informed consent was obtained from the patient (which is available upon request).

Guarantor: SR

Contributorship: AJ wrote the manuscript. SR performed the literature review. PZ and JR edited the manuscript. PG was the

primary surgeon who spearheaded the submission of the manuscript.

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