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Unusual and rare cause of abdominal pain: abdominal desmoid fibromatosis masquerading as a postoperative haematoma

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SUMMARY

Desmoid fibromatosis is a rare connective tissue malignancy. It can occur in a variety of locations, including the abdominal wall, extremities and abdominal cavity. There has been an association with development in a prior surgical scar. Common symptoms can vary depending on the location and can include being painless to having pain at the site, functional impairment and bowel obstruction from intra-abdominal masses. In the following report, we discuss a case in which a patient's abdominal pain was attributed to a postoperative haematoma based on CT radiographic features; however, further work-up and biopsy yielded desmoid fibromatosis, a rare locally aggressive malignancy.

BACKGROUND

Desmoid fibromatosis (DF) is a rare connective tissue malignancy with a reported incidence of 2–5 cases per million per year.^{1,2} DF accounts for 0.3% of all neoplasms, and 3% of all soft tissue tumours.^{1,3} DF can occur in a variety of locations, including the abdominal wall, extremities and abdominal cavity.^{1,4} The WHO classifies DF as an intermediate/locally invasive neoplasm.^{1,4} DF lacks distant malignant potential, though it can be associated with pain or organ dysfunction due to its locally invasive nature.^{1,5} Pain occurs due to direct pressure of local anatomical structures or pain arising due to the swelling.

Patients with familial adenomatous polyposis (FAP) are at increased risk of DF, with DF occurring in 10%–30% of patients with FAP and 10%–20% of patients with DF having FAP.^{1,2,5} This association is related to the implication of the dysregulation of the Wnt/ β -catenin pathway in DF. The APC gene defective in FAP is a regulator of β -catenin.^{1,2,4,5}

DF has also been shown to have a proclivity for development in locations with prior surgical scar. Specifically for abdominal wall DF, most affected patients are female with childbearing history, as was our patient in this case.^{1,3}

CASE PRESENTATION

A woman in her 60s presented to her primary care physician for further evaluation of abdominal pain. She had undergone elective surgical repair of idiopathic scoliosis 3 weeks prior. She had a history of breast cancer remotely requiring bilateral mastectomy, gastro-oesophageal reflux disease, hypertension and hypothyroidism. There was no family

history of gastrointestinal or musculoskeletal malignancy. She had reported to have been recovering well from spinal surgery other than having abdominal discomfort that waxed and waned in intensity, which was apparent postoperatively. She did not require analgesic medications for her spine but was requiring analgesic relief for the ongoing abdominal discomfort. Her abdominal wall pain was deemed related to a small haematoma based on initial CT radiological findings. She was informed it would ultimately resolve over time. Her abdominal pain had escalated seeking her to see care at a local emergency department three times prior to presentation to her primary care physician. She denied fever, chills or sweats. Her main symptom was midline abdominal discomfort. On our initial examination, she had midline abdominal pain with a lump of the midline. Her pain was out of proportion to examination. There was no induration or fluctuance. Her postoperative spinal scar was healing well, and her examination was otherwise unremarkable. Additional pathology such as neoplastic disease was considered in our differential diagnosis and further work-up ensued.

INVESTIGATIONS

Laboratory investigations yielded normal blood counts, normal chemistry panels and urine studies. There was a mildly abnormal alkaline phosphatase deemed related to recent spinal surgery.

CT of the abdomen and pelvis with contrast revealed no acute abnormality in the abdomen or pelvis and revealed postoperative changes of the spine without acute abnormalities. She had a hyperdense lesion of the midline abdominal wall measuring 2.2×1.7 cm, which was felt to be a small haematoma (figure 1).

Repeat CT was performed through the emergency department at a return visit as a result of increased pain and failure of outpatient supportive therapy with analgesics within a few weeks of initial imaging. This showed small 3.2×1.5 cm subcutaneous hyperdense lesion in the upper midline abdominal wall, increased in size when compared with the prior CT.

An ultrasound was significant for 2.9×2.1×1.5 cm irregular lobulated hypoechoic mass in the left paramedian abdominal wall with internal vascularity in which biopsy was advised (figure 2).

Soft tissue pathology demonstrated spindle cell proliferation, consistent with DF.



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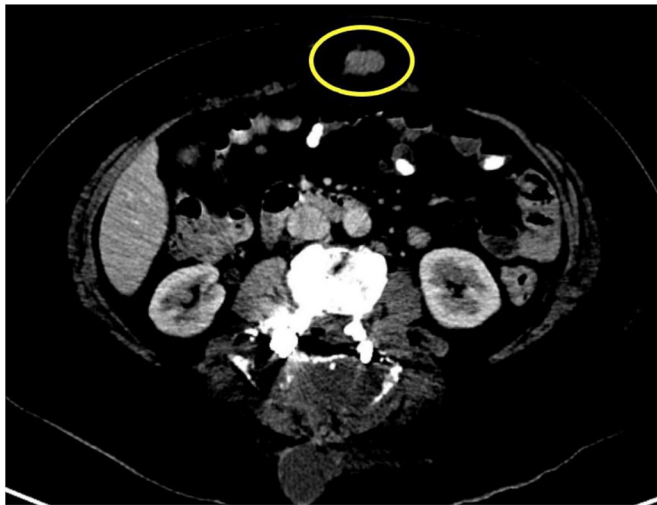


Figure 1 CT scan.

The patient was referred to surgical oncology. Given the size of the mass, superficial location and associated pain, surgical resection was deemed appropriate. She underwent surgical resection of the mass with abdominal wall repair with mesh. Postoperatively, she reported that her pain had resolved and recovered uneventfully.

Final pathology was consistent with preoperative diagnosis of DF, and margins were negative (figure 3).

DIFFERENTIAL DIAGNOSIS

The differential diagnoses for abdominal wall DF include haematoma, abscess, infections, arteriovenous malformations, lipomatous tumours, neurogenic tumours, fibromas and other mesenchymal malignancies such as fibromyxoid or spindle cell carcinomas.^{1 6} Our patient did not have fever, chills or other systemic symptoms suggestive of infection or abscess. Clinically, symptomatic postoperative spinal haematomas can be identified

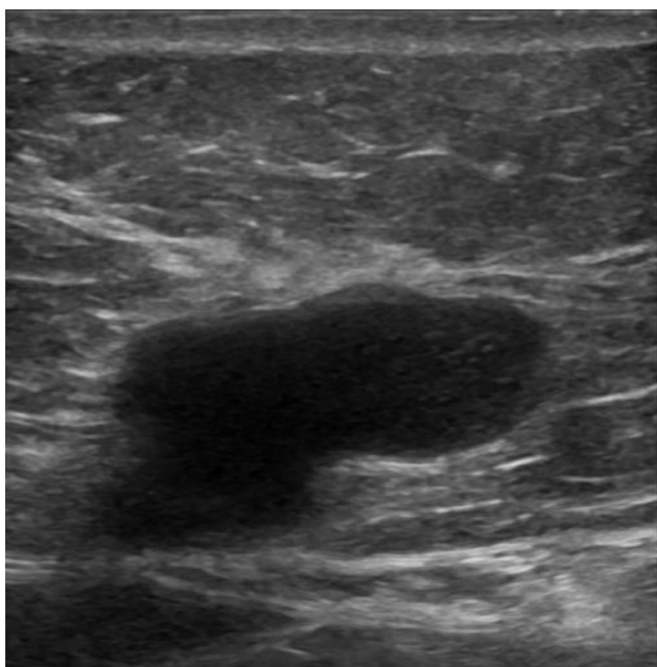


Figure 2 Ultrasound.

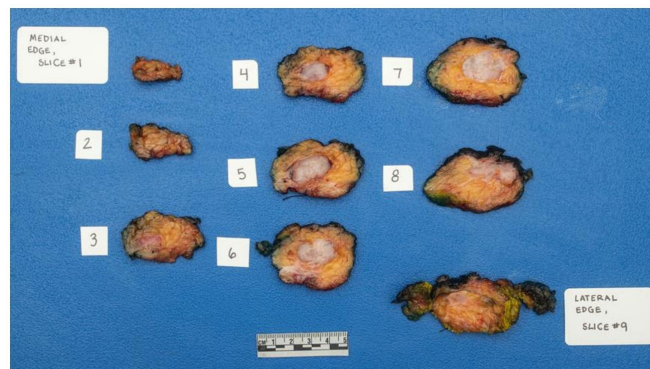


Figure 3 Pathology.

in 33%–100% of surgical cases but are identified of the epidural region.⁷ The features of her imaging did not have features of a fistula or arteriovenous malformation.

TREATMENT

Surgical excision alleviated our patient's symptoms and was chosen due to her refractory pain and interval increased size with repeat CT. Surgery had historically been deemed the mainstay of treatment with wide resection being standard practice in order to lower recurrence rates. Surgical resection may be associated with increased mortality and morbidity and thus a more conservative approach with watchful waiting is reasonable. Additional treatment options include radiotherapy and systemic therapy. Therapeutic agents that include anti-oestrogenic drugs, non-steroidal anti-inflammatory drugs, cytotoxic chemotherapy and tyrosine kinase inhibitors have been reported in achieving disease stability.

Multiple publications reveal that despite wide margin excision, local neoplastic disease recurs in up to 28% of cases.^{4 5} There is also evidence that growth arrest or spontaneous resolution may occur, and surveillance may be a reasonable alternative with intervention after watchful waiting.² Medical management in cases of progression during active surveillance may include tamoxifen, non-steroidal anti-inflammatories, methotrexate plus vinblastine, tyrosine kinase inhibitors and full-dose chemotherapy such as doxorubicin and other anthracycline uses for sarcomas.^{2 8}

OUTCOME AND FOLLOW-UP

In this case, surgical resection led to full recovery and resolution of symptoms. She was placed on non-steroidal anti-inflammatories daily. She has been actively followed with MRI at 6-month intervals and has had no recurrence of mass. She will continue surveillance over the next 5 years based on specialty recommendations.

DISCUSSION

DF is a rare but important consideration in the evaluation of a new mass. Patients with FAP are at increased risk of DF, with DF occurring in 10%–30% of patients with FAP and 10%–20% of patients with DF having FAP.^{1 2 5} This association is related to the implication of the dysregulation of the Wnt/ β -catenin pathway in DF. The APC gene defective in FAP is a regulator of β -catenin.^{1 2 4 5}

DF has also been shown to have a proclivity for development in locations with prior surgical scar. Specifically for abdominal

wall DF, most affected patients are female with childbearing history, as was our patient in this case.¹³

With regard to imaging choices during consideration of DF, different modalities may be considered but are suboptimal compared with MRI. On ultrasound, DF presents as variably defined infiltrative heterogeneous mass, with variable echogenicity.¹ On CT, DF is often isodense to skeletal muscle with areas of variable attenuation attributed to areas of high collagen content or myxoid components.^{4,5} MRI is frequently useful and is considered the gold standard for imaging. Biopsy is needed for diagnosis. MRI is considered the gold standard for staging and surveillance of DF as it provides detailed information on relationship of the mass to adjacent structures.^{2,3}

Treatment of DF was historically surgery, but local recurrence rate is around 28%.^{4,5} More contemporary approaches to treating DF include watchful waiting, radiation therapy, cryoablation and systemic agents such as chemotherapy or tyrosine kinase inhibitors.^{1,5} Regarding our patient, her DF was symptomatic and had shown interval enlargement, so surgery was the best approach.

This particular patient had recently undergone extensive spinal surgery with a posterior approach, which is associated with retroperitoneal or epidural haematoma.^{9,10} Abdominal wall haematoma resulting from spinal surgery appears to be rare, as we were not able to identify any such association in the literature.

This case demonstrates the importance of keeping a wide differential for abdominal wall mass postoperatively, especially in the case of interval enlargement or non-timely resolution of the mass as well as persistent discomfort. A clinician must

consider neoplastic disease in the differential diagnosis in cases where a haematoma seems atypical in nature.

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Case reports provide a valuable learning resource for the scientific community and can indicate areas of interest for future research. They should not be used in isolation to guide treatment choices or public health policy.

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Learning points

- ▶ Clinicians should keep a broad differential diagnosis and a soft tissue neoplasm should be considered when clinical symptoms do not resemble radiological findings.
- ▶ Desmoid fibromatosis is a rare locally invasive neoplasm, and some studies suggest observation rather than surgical excision based on an otherwise asymptomatic clinical presentation.
- ▶ Imaging with additional modalities such as ultrasound or MRI may be warranted for further differentiation if initial imaging is inconsistent with clinical findings.

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