Sporadic Desmoid-Type Fibromatosis Occurring at Laparoscopic Trocar Site: An Unusual Entity

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Abstract

Desmoid-type fibromatosis (DF) or desmoid tumours are rare benign soft tissue tumours of musculo-aponeurotic origin, which have no malignant potential but are locally aggressive and can result in significant morbidity. They may be hereditary (in association with FAP, i.e. familial adenomatous polyposis syndrome resulting from APC gene mutation) or sporadic, with hereditary cases having a much higher risk of developing DF compared to sporadic ones. Desmoids are known to occur in previous surgical scars. However, previous laparoscopic port site/trocar site is an extremely uncommon site for sporadic desmoids, with only two cases of sporadic laparoscopic trocar site desmoids (occurring in absence of FAP) reported in world literature. We thus describe a case of sporadic DF, occurring at the site of umbilical port 8 months following laparoscopic cholecystectomy for cholelithiasis, treated successfully by margin-negative resection. To the best of our knowledge, this appears to be the first such case in Indian literature and the first being reported after laparoscopic cholecystectomy. In the era of laparoscopic surgery, one must be aware of the existence of such an entity after laparoscopy as it may be confused with conditions such as metastatic deposit, port site tumour recurrence, port site hernia, scar site endometriosis, soft tissue sarcoma etc. and may result in diagnostic dilemma.

Keywords Aggressive fibromatosis · Desmoid tumour · Laparoscopy · Port site · Trocar site

Introduction

Desmoid-type fibromatosis (DF) are benign, locally invasive soft tissue tumours arising from musculo-aponeurotic tissues. DF may be sporadic or hereditary (associated with familial adenomatous polyposis, i.e. FAP). In absence of FAP, DF occurring at laparoscopic trocar sites is extremely unusual. We report a case of sporadic DF occurring at umbilical port site following laparoscopic cholecystectomy. To the best of our knowledge, this is the first such case in Indian literature and the first described after laparoscopic cholecystectomy in world literature. Port site swellings

may result in diagnostic dilemma and the possibility of DF must be considered.

Case Presentation

A 53-year-old female presented with pain and swelling in periumbilical region since 2 months. She had undergone four-port laparoscopic cholecystectomy for cholelithiasis 8 months back. Post-operative course was uneventful and histopathology revealed chronic cholecystitis. She had no significant medical or family history. On examination, 4 × 4 cm lump was palpable at previous umbilical port site, possibly arising from parietal wall muscles. The overlying skin and umbilical port site scar were healthy with no cough impulse.

Ultrasound suggested focal omental and soft tissue thickening at surgical site. Contrast-enhanced computed tomography (CECT) revealed an ill-defined 5 × 1.5 cm enhancing lesion posterior to rectus at the level of umbilicus (Fig. 1a–c). Greater curvature of the stomach and mid-transverse colon were abutting the lesion (Fig. 1a). There was no scar site

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Fig. 1 **a–c** CECT abdomen: an ill-defined enhancing lesion posterior to the rectus muscle at previous umbilical trocar site. Lesion is seen abutting the stomach and transverse colon. Note is made of post-cholecystectomy

status (empty gallbladder fossa). **d** Barium meal: extrinsic compression at the greater curvature of stomach

herniation or any gallbladder fossa mass. Ultrasound-guided percutaneous fine-needle aspiration cytology (FNAC) showed benign mesenchymal cells with possibility of fibromatosis. Endoscopy and barium studies revealed extrinsic compression of pylorus along the greater curvature (Fig. 1d). Colonoscopy was normal.

At laparotomy, the mass was seen arising from posterior rectus sheath, behind the rectus muscle and adherent to the previous trocar site scar tissue. There was no extension into rectus muscle; however, it was extending intra-peritoneally, adherent to greater curvature of stomach, mid-transverse colon and greater omentum. We performed a wide local excision of the lump along with the adherent posterior rectus sheath. A gastric sleeve was removed en bloc, with segmental transverse colectomy with stapled colo-colic anastomosis and omentectomy (Fig. 2a). Umbilical trocar site scar tissue was excised completely and abdominal wall reinforced with polypropylene mesh. Post-operative course was uneventful.

Cut section showed areas of fat, haemorrhage and calcification (Fig. 2b). Gastric and colonic mucosal surfaces were normal (Fig. 2c, d). Histopathology confirmed desmoid-type

fibromatosis extending up to muscularis propria of the colon and stomach; however, cut ends were uninvolved. All margins were negative. The patient is on follow-up with no clinical signs of recurrence after 1 year.

Discussion

Desmoid tumours are rare fibroblastic mesenchymal tumours. They are histologically benign but are locally aggressive and tend to invade adjacent structures. Majority of DF are sporadic; only 10–15% being hereditary, associated with FAP with APC gene mutations [1]. The most significant risk factor for both types is surgical trauma. Several cases have been described following surgical or traumatic scars. However, sporadic desmoids arising in laparoscopic port sites are extremely uncommon with only two cases in world literature [2, 3]. Kaplan described the first such case in an infra-umbilical scar of laparoscopic tubal ligation [2]. Hayashi et al. reported another case masquerading as port site recurrence after laparoscopic retroperitoneal lymph node dissection [3]. Our case

Fig. 2 **a** Gross specimen showing en bloc wide excision of lump along with the adherent posterior rectus sheath, gastric sleeve (solid arrow), transverse colonic segment (dashed arrow) and omentum. **b** Cut section of mass showing areas of fat, calcification and haemorrhage. **c** Gastric sleeve opened up to reveal normal gastric mucosa. **d** Transverse colonic segment opened up to reveal normal colonic mucosa

represents the first report of sporadic laparoscopic port site desmoid in Indian literature and the third case worldwide. Also, it is the first case reported after laparoscopic cholecystectomy. Cases of port site desmoids have been described following laparoscopy in FAP patients [4, 5]. These patients, however, have a much higher risk of developing DF (around 850 times) compared to sporadic cases [1]. Such desmoids represent an extra-intestinal manifestation of FAP itself.

Port site desmoids present with palpable lump mimicking other conditions like port site tumour recurrence/metastasis, soft tissue sarcomas, endometriosis, incarcerated hernia or organised hematoma. Imaging may provide clue; however, definitive diagnosis is based on biopsy and immunohistochemistry. Colonoscopy must be performed in young patients to rule out FAP.

The treatment involves margin-negative resection, but extent of resection remains controversial. Although R0 resection seems appropriate, recent literature indicates that microscopic close margins (< 1 mm) or even R1 resection may have similar outcomes [1, 6]. Few asymptomatic patients may be followed up, if there is no tumour progression on serial imaging ('Wait and Watch' policy) [7]. Tumours which are unresectable, recurrent, have positive margins or resulting in substantial abdominal wall defect, are treated with radiotherapy, non-steroidal anti-inflammatory drugs, anti-oestrogens (tamoxifen), systemic chemotherapy (anthracyclines, vinblastine, methotrexate) or targeted therapy (Tyrosine kinase inhibitors) [1, 6]. However, treatment strategy must be individualised. Post-resection follow-up is mandatory as recurrence rates are high (23–25%) even after R0 resection [6, 8].

Conclusion

Sporadic DF can arise at laparoscopic trocar sites and must be considered whenever patients present with port site swelling months after laparoscopy. With increasing number of laparoscopic procedures, more number of cases may be encountered in the future. Early diagnosis with high index of suspicion leads to definitive treatment when the tumours are small and avoids morbidity of extensive abdominal wall resection and reconstruction.

Compliance with Ethical Standards

Conflict of Interest The authors declare that they have no conflict of interest.

Informed Consent The authors confirm that an informed consent was obtained from the patient to be the subject of this case report. None of the images used disclose the identity of the subject in any form and patient anonymity has been maintained at all places in the text.

Ethical Statement An ethical approval was not necessary as this was a retrospective review of a clinical case and not experimental research data.

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