

Pelvic Fibromatosis Following Total Abdominal Hysterectomy with Bilateral Salpingo-oophorectomy: A Case Report

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ABSTRACT

Desmoid tumours, also known as aggressive fibromatosis, are rare, benign, non-inflammatory, fibroblastic tumours. They are characterised by locally aggressive behaviour and a high potential for recurrence, but they do not metastasise. Here, the authors report the case of a 51-year-old female who presented with a lump in the lower abdomen for one month. The patient had a history of exploratory laparotomy with total abdominal hysterectomy and bilateral salpingo-oophorectomy for ovarian cystadenocarcinoma 15 years earlier. Following Ultrasonography (USG) and Contrast-Enhanced Computed Tomography (CECT) of the abdomen and pelvis, an exploratory laparotomy was performed, and a mass that was free from adjacent structures was excised. Histopathological examination revealed benign desmoid-type fibromatosis with osseous and chondroid metaplasia. The patient was discharged uneventfully. The current incidentally detected desmoid-type fibromatosis with osseous and chondroid metaplasia highlights the importance of vigilant follow-up and consideration of diverse pathologies, even in low-risk clinical scenarios.

Keywords: Contrast-enhanced computed tomography, Desmoid tumours, Intra-abdominal fibromatosis

CASE REPORT

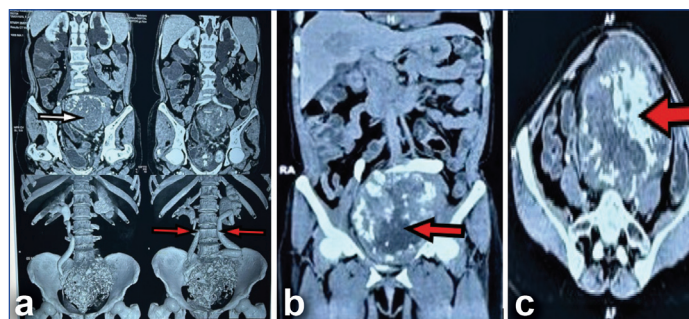
A 51-year-old female presented with the chief complaint of a lump in the lower abdomen for one month. The lump had an insidious onset and was associated with localised, dull-aching pain in the lower abdomen. She also complained of back pain and constipation. The patient had previously undergone exploratory laparotomy with total abdominal hysterectomy and bilateral salpingo-oophorectomy for ovarian cystadenocarcinoma 15 years earlier in the Gynaecology Department, during which she received a one-pint blood transfusion. She had no known co-morbidities and no significant personal or family history. Her obstetric history was G4P4A0L4.

On inspection, a visible lump was noted in the hypogastric region, with the umbilicus centrally placed and a healthy scar from the previous laparotomy. The abdomen was neither warm nor tender. On palpation, a single hard lump measuring approximately 11×10 cm was felt, extending from the hypogastric to the umbilical region. The mass was non-mobile, free from the overlying skin, and had well-defined superior and lateral margins. The inferior margin was not palpable. The surface was smooth, and the mass did not move with respiration. Per rectal examination revealed a mass lesion on the anterior rectal wall; however, the rectal wall itself was free from the mass.

Ultrasonography revealed a heterogeneous mass measuring 10.7×10.8×12 cm in the pelvic cavity, displacing the urinary bladder to the right and causing bilateral hydronephrosis. A 3.8×2.9 cm hyperechoic lesion in the liver was noted, suggestive of either a hemangioma or metastasis. CECT of the abdomen with intravenous pyelography demonstrated a mixed-density pelvic mass measuring 14.8×12.2×12.1 cm, showing mild enhancement, multiple calcified areas, and a non-enhancing central region. The mass compressed both the left and right lower ureters, resulting in moderate bilateral hydroureteronephrosis, and caused severe compression of the urinary bladder and sigmoid colon. The bowel loops were displaced but maintained a clear fat plane.

The liver was normal in size and showed a few well-defined, lobulated hypodense lesions in both lobes, demonstrating peripheral globular

contrast enhancement with gradual centripetal filling and delayed contrast persistence, consistent with hemangiomas. No evidence of metastasis was noted [Table/Fig-1].

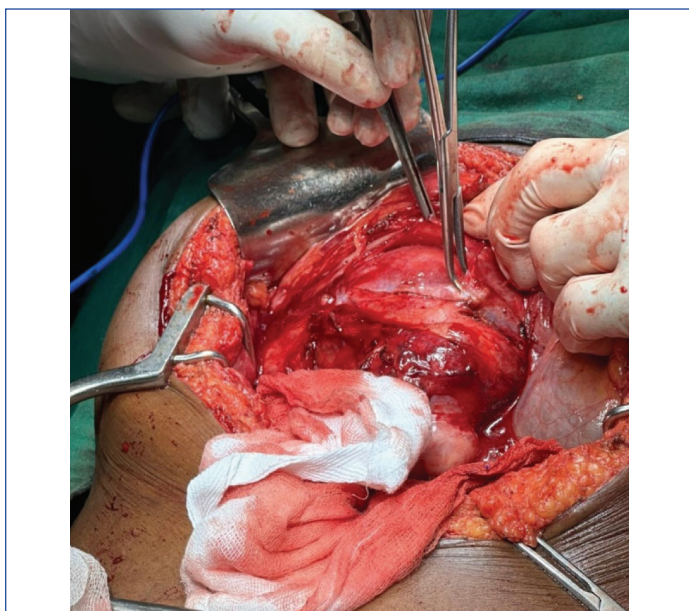


[Table/Fig-1]: CECT abdomen with pelvis with intravenous pyelography: a) Coronal section (white arrow) shows lesion, (Red arrow) shows bilateral hydronephrosis and bilateral hydroureter; b) Coronal section shows a mass lesion; c) Axial section shows mass lesion.

After obtaining a urologist's opinion, surgical intervention was planned. Bilateral ureteric catheters were inserted by the urologist with the patient in the lithotomy position. Subsequently, the patient was placed in the supine position, and an exploratory laparotomy was performed. A mass was identified in the pelvic region [Table/Fig-2]. The small bowel was free from the mass and was retracted cranially. Lateral and posterior dissection was then carried out. The mass was found to be free from the ureters and the common iliac vessels. The encapsulated mass was removed intact.

On gross examination, the excised specimen was a lobulated mass measuring 11×11×9 cm and weighing 1 kg. The outer surface was smooth, glistening, and encapsulated. On cut section, the mass was firm to hard in consistency, with the release of yellowish, jelly-like fluid. The inner surface revealed whitish, homogeneous, and calcified areas [Table/Fig-3].

Microscopic examination showed a hypocellular lesion composed of sheets of slender, elongated spindle cells with pinpoint nuclei. Wavy collagen fibres, myxoid and cartilaginous areas with hyalinisation

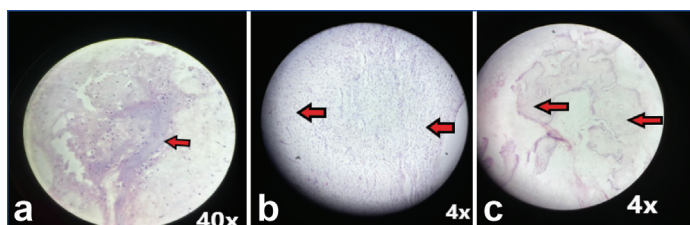


[Table/Fig-2]: Intraoperative picture of mass.



[Table/Fig-3]: Gross specimen.

were noted. Fragmented bony trabeculae were present in certain areas. Scattered chronic inflammatory cells and focal areas of haemorrhage were also observed [Table/Fig-4]. Overall, the histopathological features were suggestive of fibromatosis with osseous and chondroid metaplasia.



[Table/Fig-4]: Histopathology: a) Chondroid component; b) Spindle cells; and c) Osseous component.

Haemostasis was achieved, and a drain was placed in the pelvis. Oral liquids were started on postoperative day 1, followed by a soft diet on postoperative day 2, which was well tolerated. The patient was discharged on postoperative day 7. All sutures were removed on postoperative day 12. The patient was followed up every two months for six months. She remained asymptomatic, and no lesion was detected on follow-up Magnetic Resonance Imaging (MRI).

DISCUSSION

Desmoid tumours (aggressive fibromatosis) are benign, non-metastatic fibroblastic tumours characterised by infiltrative growth

and a high rate of local recurrence. The term “desmoid” was introduced by Müller in 1838 and is derived from the Greek word meaning “tendon-like” [1]. Desmoid tumours are thought to arise from inherited or acquired defects in connective tissue growth regulation. They commonly develop following surgical trauma, such as post-caesarean section, and during periods of increased oestrogen exposure, including pregnancy and oral contraceptive use [2-5].

A strong association exists with Familial Adenomatous Polyposis (FAP), particularly Gardner's syndrome, which involves a germline mutation in the APC gene affecting the Wnt signalling pathway [2-5]. Therefore, assessment of family history is essential, although no such history was present in our patient. Desmoid tumours are rare, accounting for approximately 0.03% of all neoplasms and less than 3% of soft tissue tumours. They predominantly affect young women and are often associated with pregnancy, while in elderly patients, the gender distribution is more equal [6,7].

Pathophysiologically, desmoid tumours represent an infiltrative fibroproliferative disorder involving the fascia and muscle aponeuroses [4,5]. Although they can occur at any anatomical site, they most commonly involve the extremities, trunk, abdominal wall, and intra-abdominal region. Tumours of the abdominal wall typically arise from the rectus abdominis muscle, whereas intra-abdominal lesions commonly involve the mesentery or pelvis [7]. Post-hysterectomy desmoid tumours in postmenopausal women are rare. Interestingly, some tumours have been reported to regress after menopause, oophorectomy, or during male puberty [2].

Desmoid tumours may enlarge and adhere to adjacent structures, resulting in organ damage, functional impairment, or neurological symptoms [4]. Kumar SS et al., reported a case of an intra-abdominal desmoid tumour with pelvic extension involving the abdominal wall, urachus, ureter, and bladder, in which complete excision was achieved; however, long-term follow-up was not described [5]. The clinical course of desmoid tumours is unpredictable, ranging from spontaneous regression to aggressive local growth. Most lesions remain asymptomatic until they cause organ dysfunction, with pain being the most common presenting symptom when present [4].

Our patient presented with mild pelvic and lumbar pain, with hydronephrosis detected only on ultrasonography. Ormonde M et al., (2022) reported a case of pelvic desmoid tumour infiltrating the right ureter, leading to renal failure and necessitating tumour excision, right nephrectomy, hysterectomy, and bilateral salpingo-oophorectomy [4].

According to Kumar SS et al., only approximately 50% of desmoid tumours are correctly diagnosed preoperatively due to their rarity and non-specific clinical and imaging features [5]. Therefore, a high index of clinical suspicion is essential, particularly in patients without a relevant family history or co-morbidities. Differential diagnoses include ovarian neoplasms, sarcoma, metastasis, Gastrointestinal Stromal Tumour (GIST), sclerosing mesenteritis, endometriosis, and haematoma [4,5].

MRI is the preferred imaging modality for desmoid tumours, as it provides superior delineation of tumour extent and its relationship to adjacent structures compared with computed tomography [4,5]. Definitive diagnosis relies on histopathological examination. Although needle biopsy may be attempted, it is technically challenging and carries a risk of tumour cell dissemination, particularly when malignancy is suspected. Consequently, surgical excision is often chosen as both a diagnostic and therapeutic approach [4].

Histologically, desmoid tumours demonstrate infiltrative proliferation of clonal fibroblastic or myofibroblastic cells arranged in long fascicles, without significant cytological atypia or mitotic activity, embedded within a collagen-rich stroma. Perivascular lymphocytes and thick-walled blood vessels may also be present [4]. Immunohistochemically, tumour cells typically show nuclear

β -catenin positivity {associated with Catenin Beta 1 (CTNNB1) mutations} and variably express vimentin, Smooth Muscle Actin (SMA), and calponin, while being negative for cytokeratin, desmin, Cluster of Differentiation protein 34 (CD34) and S100 [4,5].

The management of desmoid tumours remains controversial and depends on tumour location, size, symptomatology, and individual patient factors [4]. Recent guidelines, including those from the National Comprehensive Cancer Network (NCCN), recommend conservative management for asymptomatic, stable, and non-life-threatening tumours [8]. Active surveillance has gained acceptance, with surgery reserved for cases of progressive disease, organ dysfunction, significant symptoms, or when malignancy cannot be ruled out.

In the present case, the patient had bilateral hydronephrosis, progressive pelvic pain, and diagnostic uncertainty. Surgical intervention was therefore undertaken, resulting in complete resection with negative margins. The mass was free from adjacent structures, and postoperative imaging demonstrated resolution of hydronephrosis, with no evidence of recurrence at six months of follow-up.

Although complete excision with negative margins is the preferred goal, it is achieved in only about 50% of cases [9,10]. Positive margins may be acceptable when complete resection poses a high risk of morbidity. A multidisciplinary approach is essential to balance oncological control with preservation of function.

Radiotherapy is generally reserved for unresectable tumours and is not routinely recommended in the postoperative setting. Systemic therapies-including Nonsteroidal Anti-Inflammatory Drugs (NSAIDs), tamoxifen, cytotoxic chemotherapy, and targeted agents such as sorafenib and pazopanib-are emerging treatment options [9]. Chao AS et al., reported long-term remission in a young woman treated with tamoxifen for recurrent pelvic desmoid tumour [11].

Recurrence remains a significant concern, with reported rates ranging from 20% to 60%, most commonly occurring within the first two years following treatment [12]. The NCCN recommends follow-up with clinical assessment and imaging every 3-6 months for the

first 2-3 years, followed by annual surveillance. The patient remains asymptomatic and disease-free at six months, underscoring the importance of individualised, multidisciplinary care.

CONCLUSION(S)

Although desmoid tumours are more commonly seen in premenopausal women and are often associated with pregnancy or childbirth, they should also be considered in the differential diagnosis of postmenopausal women presenting with an abdominal mass, particularly following previous surgical procedures.

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