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Case Report

Pelvic aggressive fibromatosis masquerading as a uterine fibroid: a diagnostic challenge

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ABSTRACT

An invasive pelvic mass in a young female presents a diagnostic challenge, as a variety of benign and malignant conditions can contribute to such findings. One rare but significant cause of an invasive pelvic mass are desmoid tumours (also called aggressive fibromatosis) benign, slowly growing fibroblastic neoplasms with no metastatic potential but a propensity for local recurrence, even after complete surgical resection. We report a case of pelvic aggressive fibromatosis - a young woman who presented with gynaecological symptoms and was found to have a large invasive pelvic mass that closely mimicked a uterine fibroid or gynaecological malignancy on clinical examination and imaging. Surgical management was undertaken due to the extent of the lesion and diagnostic uncertainty. Definitive diagnosis was established only on histopathological examination, supported by immunohistochemistry, confirming desmoid fibromatosis. This case underscores the importance of including desmoid fibromatosis in the differential diagnosis of invasive pelvic masses in young females. It also highlights the limitations of imaging in distinguishing benign from malignant pelvic tumours and emphasises the pivotal role of histopathology and immunohistochemistry in establishing the diagnosis and guiding appropriate multidisciplinary management.

Keywords: Desmoid fibromatosis, Pelvic mass, Immunohistochemistry, Histopathology

INTRODUCTION

Desmoid fibromatosis (DF), also referred to as aggressive fibromatosis or musculoaponeurotic fibromatosis, is a rare mesenchymal tumour characterized by clonal proliferation of fibroblasts in deep soft tissues.[1] The estimated incidence is approximately 2.4–5 per one million population per year.[2] DF occurs more commonly in females, often in the third to fourth decades of life.[2]

Although histologically benign and lacking metastatic potential, DF demonstrates a locally aggressive and infiltrative behaviour with a high propensity for local recurrence. Because of this, it is classified by the World Health Organization (WHO) among tumours of "intermediate (locally aggressive)" behaviour.[1]

Tumour location influences clinical presentation and management. DF may occur in the abdominal wall, intra-abdominal (mesentery, retroperitoneum), or extra-abdominal soft tissues (limbs, trunk) — intra-abdominal or pelvic lesions being relatively rare.[2] In women of reproductive age, pelvic desmoid tumours are especially rare, and due to their size and infiltrative growth, they may mimic gynaecologic or adnexal malignancies, making preoperative diagnosis difficult.[3]

Historically, wide surgical excision was the standard of care. However, surgical morbidity, functional impairment, and high recurrence rates (ranging from ~19% to 77% in various series) have led to a paradigm shift towards conservative management, including active surveillance, and systemic therapies in many cases.[4]

We present here a rare and challenging case of a large invasive pelvic DF in a young woman, with tumour infiltration through sciatic and obturator foramina — a presentation that closely mimicked a uterine fibroid or a gynaecologic tumour — managed by surgical excision and adjuvant tyrosine-kinase inhibitor therapy.

CASE REPORT

A 30-year-old woman, P2L2, previous LSCS, not sterilised with no known comorbidities referred from a local hospital in view of mass in the vagina. Patient complaints of scanty menstruation, vaginal discharge & dyspareunia. No other relevant gynaecological complaints or pressure symptoms. Per speculum and per vaginal examination revealed a large irregular mass seen occupying right vaginal wall extending up to the right fornix around 6×8 cm with ill-defined border, mass seems to be adherent to right pelvic wall. On per rectal examination, a large mass felt about 2 cm from anorectal junction. Vaginal excision of the mass was tried at the local hospital and the excised HPE report came as low grade spindle cell neoplasm.

Imaging

MRI & CT was done and it showed a large heterogeneous appearing lobulated mass lesion, measuring 10.9×12.4×7.6 cm in the right side of pelvic cavity, inferiorly extending to the ischio-rectal region, superior extension to the S1-S2 level. Medially- displacing rectum, cervix, vagina, uterus urinary bladder, proximal anal canal to the left. Anteriorly-abutment on external iliac muscle. Antero laterally- extra pelvic extension of the lesion through the obturator foramen. The obturator muscles are not properly delineated - s/o invasion. Laterally-the lesion is abutting the ischium, medial wall of acetabulum.

Postero-laterally- lesion is extending through the greater sciatic foramen abutting the pyriformis muscles and gluteus minimus muscle. Antero-inferiorly the lesion extends above the pubic symphysis Right distal ureter is closely related to the anteromedial aspect of the right pelvic lesion. Both ovaries normal. Uterus appears otherwise grossly normal.

The margins were poorly defined, with infiltrative characteristics. The appearances favoured a deep soft tissue mesenchymal proliferation rather than a simple adnexal mass.

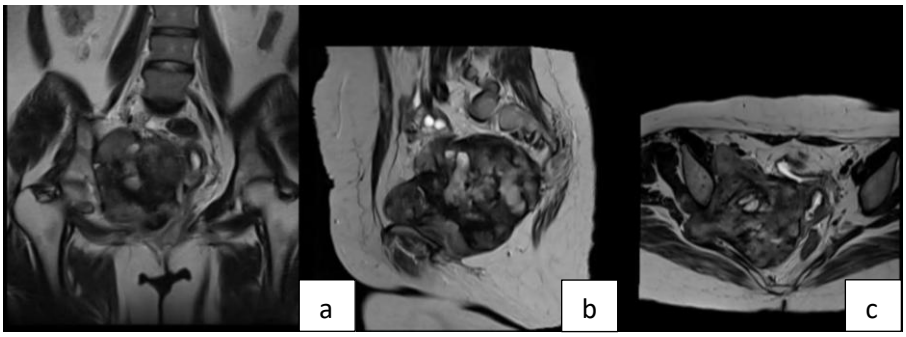

Figure 1 (a-c): MRI Pelvis demonstrating large infiltratuve right sided mass lesion with displacement of pelvic viscera.

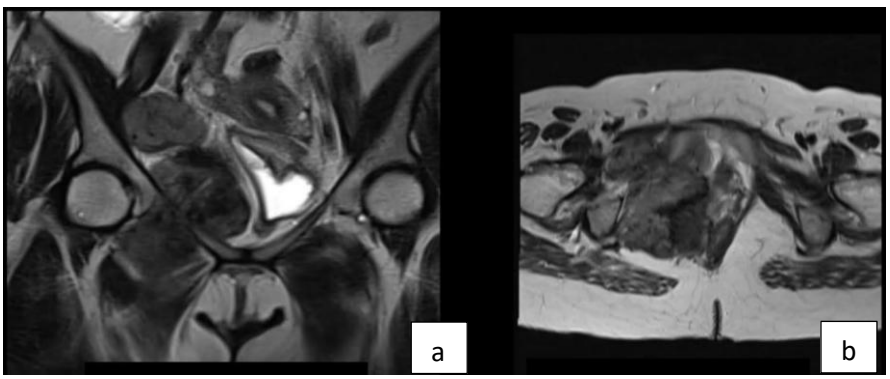

Figure 2 (a and b): Tumour extension through obturator and greater sciatic foramina with lateral pelvic wall infiltration.

Procedure

An exploratory laparotomy was performed. Extraperitoneal approach by a supra pubic transverse incision.

Intra-operative finding

Intraoperatively large hard masses in the pelvis occupying right half of pelvis, right iliac fossa around iliac vessels reaching up to femoral canal was seen. The deep part of the tumour was extending to the right obturator foramen, greater sciatic notch shifting the uterus, bladder, urethra, bladder neck. Per vaginally the mass is also felt as hard nodular mass with induration to vaginal wall. There was another mass along the posterior aspect of right iliac vessels. Tried to mobilize the mass in toto. As it was fixed everywhere it was decided to excise the mass in pieces. Most of the tumour were removed. Some residual mass at iliac vessels near the inguinal ligament, lateral pelvic wall. Because of the high risk of injury to neurovascular structures, complete en bloc resection was not feasible. Resected tissue was sent for histopathological examination.

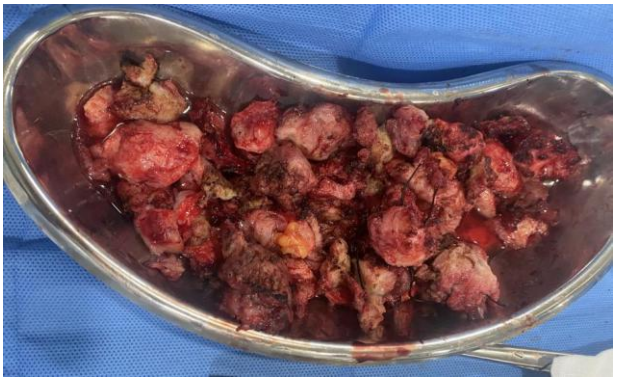

Figure 3: Intraoperative image showing piecemeal surgical excision of the infiltrative pelvic mass.

Histopathological report

Macroscopy- resected tumour - multiple fragments -cut section – grey-white, firm with whorling and focal glistening areas.

Microscopy

Microscopically, the tumour consisted of bland spindle-shaped fibroblasts arranged in long fascicles embedded in a collagenous stroma. There was no significant nuclear atypia, mitotic activity, or necrosis. These features are characteristic of desmoid fibromatosis.

Immunohistochemistry results

Smooth muscle actin (SMA): positive, desmin & beta catenin: negative, CD117 (c-KIT): negative, other markers (e.g., S-100, cytokeratins) were negative — excluding sarcoma, GIST, or other soft-tissue tumours.

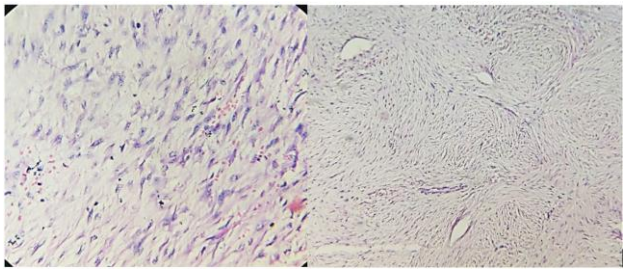

Figure 4: Histopathological examination showing bland spindle-shaped fibroblasts arranged in long fascicles embedded in a collagenous stroma.

Final diagnosis

Based on morphology and IHC, a diagnosis of desmoid fibromatosis was made.

Adjuvant treatment

Case was discussed in multidisciplinary tumor board and plan was taken for adjuvant therapy. Given the residual tumour and risk of progression, the patient was started on Pazopanib, a tyrosine-kinase inhibitor (TKI). She was kept under regular follow up.

DISCUSSION

Desmoid fibromatosis remains a clinical enigma: benign histological appearance, but aggressive, infiltrative behaviour. The modern understanding and management of DF have evolved substantially over recent decades.[5]

Epidemiology and pathogenesis

DF incidence is low: ~2.4–5 per million per year.[2] There is a clear female predominance; many cases occur in women of childbearing age (20s–40s).[4] Risk factors include hormonal influences (estrogen), prior surgery or trauma, and in some cases, association with Familial Adenomatous Polyposis (FAP) or prior radiation therapy.[2] However, the majority of desmoids are sporadic, with no inherited syndrome.[3]

Clinical behaviour

DF does not metastasize, but is locally aggressive, infiltrates surrounding structures, and can cause major morbidity if near neurovascular or visceral structures.[1] The natural history of DF is unpredictable: some may remain stable, some regress, while others progress rapidly.[5]

Recurrence after surgery is common, especially when margins are positive or when the tumour infiltrates major vessels, nerves, or other structures.[6]

Diagnostic challenges

Pelvic or intra-abdominal DF is rare; reports in women are few.[3] When arising adjacent to pelvic organs (vagina, uterus, adnexa, rectum), DF may mimic gynaecologic tumours (ovarian, adnexal, fibroid), gastrointestinal stromal tumours (GIST), sarcomas, or deep infiltrating endometriosis.[3] Imaging (MRI) is useful to define extent and relation to surrounding structures; DF on MRI is often iso- or hypointense to muscle on T1 and hyperintense on T2, with variable contrast enhancement and low-intensity bands corresponding to collagen.[3] However, imaging is rarely diagnostic — histopathology with IHC is required to distinguish DF from GIST, sarcoma, or other soft tissue tumours.[3]

Management: evolving paradigm

Historically, the standard management of DF was wide surgical excision (R0 resection) with negative margins.[7] But Recurrence rates have varied widely: many series report 19–77%.(4) Risks include functional impairment, morbidity, and difficulty when tumours are near vital structures (vessels, nerves).[7] Because of these limitations, a paradigm shift has occurred: initial active surveillance ("watch-and-wait") is now often recommended, especially for asymptomatic or stable disease.[8] In cases of symptomatic, progressive, or life-threatening DF (due to size, compression, pain or functional impairment), intervention may be necessary. Options include surgery, radiotherapy, systemic therapy (e.g., TKIs), ablation techniques, etc.[9]

Surgery

Best suited for small, resectable tumours in favourable anatomical locations (e.g., abdominal wall).[7] For deep pelvic, intra-abdominal, or neurovascular-involving tumours, surgery may be partial or even unfeasible. In such cases, piecemeal excision or debulking may be considered — with acceptance of residual tumour and plan for adjuvant therapy.[3]

Systemic therapy

Recent advances include use of tyrosine-kinase inhibitors (TKIs) such as Imatinib, Sorafenib, Pazopanib, and newer agents like Nirogacestat (a gamma-secretase inhibitor – recently FDA-approved). These agents offer non-surgical management, especially useful for unresectable, residual, or recurrent tumours.[4]

Radiotherapy / ablation

Radiotherapy may be considered when surgery is incomplete (positive margins) or not feasible, but potential long-term toxicity is a concern (especially in younger patients).[7]

Ablative techniques (e.g., cryoablation, radiofrequency) are sometimes considered in selected cases.[9]

CONCLUSION

Desmoid fibromatosis — though benign under the microscope — can behave aggressively, infiltrating deep pelvic structures, mimicking uterine fibroids or malignant tumours. However, rare soft-tissue tumors such as desmoid fibromatosis may closely mimic these conditions, leading to diagnostic delay and inappropriate initial management. This case highlights the importance of a multidisciplinary approach in managing desmoid fibromatosis. Early diagnosis, individualized surgical intervention, and targeted adjuvant therapies, including tyrosine kinase inhibitors, are essential for optimizing outcomes in this rare and challenging condition. These rare mesenchymal tumours in spite of the benign histopathology have an aggressive and invasive growth pattern which mainly leads to the misdiagnosis. Hence, it is essential for gynaecologic oncologists to be aware of its presentation and treatment.

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