Case Report

Lower Abdominal Parietal Tumours in the Puerperal Period — A Rare Etiology

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Parietal tumours developing in the lower abdominal wall in the puerperal period in post lower segment caesarean section women are rare and can be confused with organized hematoma, nodular fasciitis and scar endometriosis. Desmoid fibromatosis,though rare, should be considered in the differential. The high estrogen physiological state during pregnancy, abdominal wall stretching due to foetal growth and previous scar due to lower segment caesarian section, may trigger its development. These tumors are locally aggressive and highly prone to recurrence but rarely metastasize to distant sites.Some physicians choose a wait and watch policy for this tumour. But, wide surgical excision remains the mainstay of treatment, in the absence of any standardized guidelines for non-operative and adjuvant management. A high level of clinical suspicion supported by imaging and core needle tissue biopsy helps in clinching the diagnosis. We report two cases of abdominal wall DF that presented to our institution within a year of childbirth.

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Lower abdominal wall tumours in the puerperal period in post-lower segment caesarean section (LSCS) patients are extremely rare and a cause of great concern to the treating physician. The first differential seems to be a rectus sheath haematoma while a close second is a scar endometriosis. Desmoid fibromatosis (DF) is a rare entity, representing <0.03% of all soft tissue neoplasms, which may affect any musculoaponeurotic area in the abdominal cavity, trunk or extremities[1]. DF as a possible etiology for lower abdominal wall tumour in the puerperal period is rare but has to be kept in consideration. We report two cases of abdominal wall DF that presented to our institution within a year of childbirth.

Case Reports

Case 1 :

A 23-year-old primipara lady presented with a lump in the right iliac fossa, 3 months after undergoing LSCS. The parietal lesion was 8cm x 5cm in size, well circumscribed, elliptical, non-tender and free from overlying skin (Fig 1a). A diagnosis of organized hematoma as a complication of LSCS was considered in view of its proximity to the operative scar and short duration of symptoms. Ultrasonography showed anisoechoic, homogeneous mass from which a core needle biopsy was done. This reported a DF. A MRI scan done subsequently, revealed a solid, homogenous mass arising from the anterior abdominal wall.

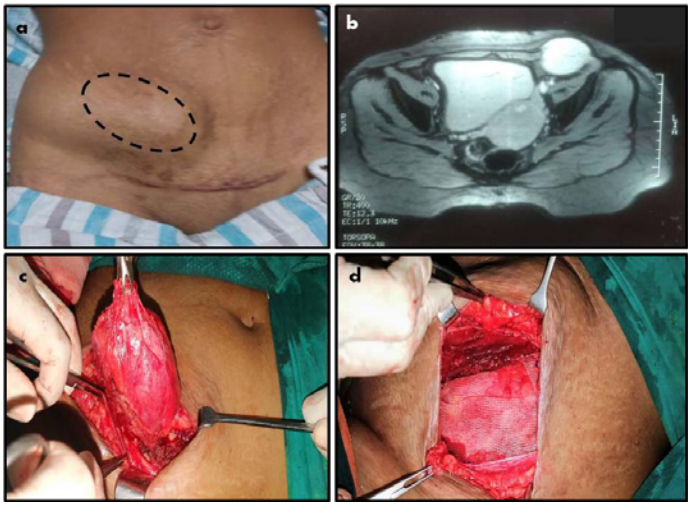

Fig 1 — (a) Case 1 showing lesion in lower abdominal wall close to LUCS scar. (b) MRI showing lesion arising from anterior abdominal wall with no intra-abdominal extension. (c) Intra operative image showing spindle shaped tumor mass arising from anterior abdominal wall. (d) Reconstruction of post excision defect with polypropylene mesh

Case 2 :

A 28-year-old gravida 2 lady presented with a painless, slow growing parietal lump in her left iliac fossa 1 year after her second LSCS. The lesion measured 10cm x 6 cm in size. Endometriosis was initially considered. Ultrasonography and MRI confirmed a homogenous fibrotic mass arising from the abdominal wall musculature without any intra-abdominal extension (Fig 1b). Core needle biopsy confirmed DF.

Both patients underwent wide local excision of the lesion with polypropylene mesh reconstruction of the

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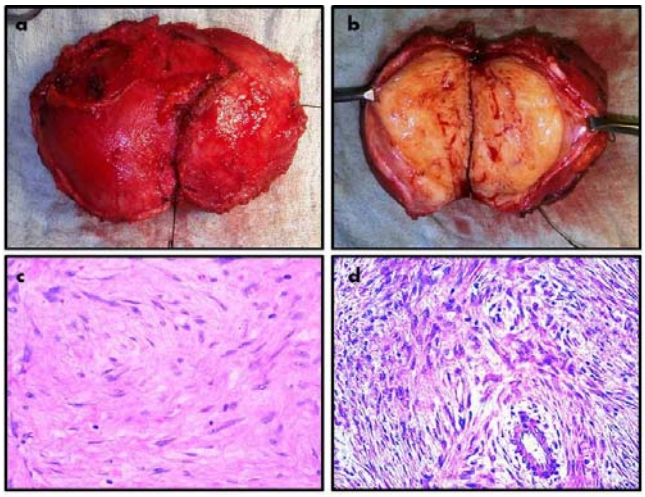

Fig 2 — (a) Specimen of wide local excision of tumour. (b) Cut section of excised tumour. (c) & (d) Haematoxylin &Eosin stained low and high power microscopic view

abdominal wall defect (Fig 1c,d) and had an uneventful recovery. Histopathology showed bland fibroblasts arranged in broad sweeping fascicles, spindle cells with small slender nuclei and dense collagen consistent with DF (Fig 2). Surgical margins were clear. Immuno-histochemistry for CD117, DOG1, CD34, ER, PR were negative. They were followed up for two years and did not show any recurrence.

Discussion

DF is a rare group of slow growing deep fibromatoses characterized by monoclonal fibroblastic proliferation with a variable, unpredictable clinical course[2]. They are benign, locally aggressive and rarely metastasize. Both sexes can be equally affected. Various intra and extra abdominal sites can be affected since it affects musculoaponeurotic structures, though there is a predilection for the abdominal wall[3,4]. Most arise sporadically, though an association with FAP is reported in 5-10% of total cases[5]. Scar endometriosis is a possible differential diagnosis. However, most endometriotic tumours of the abdominal wall are subcutaneous in location, are likely to be painful and fluctuate in size in relation to menstruation[3]. Other differentials include gastrointestinal stromal tumour of abdominal wall, schwannoma, nodular fasciitis[2]. A CT / MRI and tissue diagnosis is mandatory to plan further management.

DF can range in size from small lesions to massive ones[3,6]. Within the abdominal wall there is a predilection towards the infra umbilical rectus sheath.

There is no clear consensus as to why DF may occur in the puerperal period. A relation between endogenous estrogen levels and DF has been reported, with 33% exhibiting estrogen receptors[7]. The initial growth of the tumour may be triggered during pregnancy, with subsequent growth occurring in the puerperal period. Fetal growth leading to stretching of the abdominal wall musculature may be another triggering factor. Presence of a surgical scar in the abdomen also further predisposes to DF development.

Surgical excision is the most effective treatment. A disease free margin of >1 cm has been reported to effectively reduce recurrences. DF is locally aggressive and incidence of local recurrence may range from 20-60%. Age <37years, tumour more than 5-7cm,extra abdominal location has been considered as other predictors of recurrence[3].

Some authors have recommended a conservative, non-operative approach since arrest of growth of the lesion or even regression has been described in postpregnancy, postoophorectomy and postmenopausal period. These authors described tumor regression in patients treated with antiestrogendrugs like Tamoxifen and Toremifene[3].

Unresectable tumours and those at inaccessible sites may be treated with radiotherapy or hormonal therapy[1]. Chemotherapy and tyrosine kinase inhibitors have also been tried in large, aggressive DF, not responding to hormonal therapy[8]. But, there are no definitive guidelines regarding non-surgical management of DF.

Conclusion

DF of the abdominal wall, though rare, should be considered in the differential diagnosis of parietal wall swellings during the puerperal period. As its definitive treatment remains an enigma, surgical excision remains the mainstay of treatment. Being a locally aggressive neoplasm, early diagnosis is essential to be able to achieve clear margins.

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Conflict of interest - Nil

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