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Rare Case of Pregnancy with Desmoid Tumour

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Introduction

Desmoid tumor, referred to as benign deep fibromatoses, originating from fascia and muscle aponeurosis with an aggressive infiltrating growth. It constitutes about 3% of all soft tissue neoplasms.[1] Desmoid tumor is often associated with female gender, familial adenomatous polyposis and occasionally with surgical trauma. It has a higher prevalence in women who experienced pregnancy. The superficial disease is slowly growing, small and rarely involves deeper structures. The deep variant has a relatively rapid growth, attains larger size and has a high local recurrence rate. Most spontaneous desmoids occur in the shoulder girdle and the anterior abdominal wall, whereas intra-abdominal desmoids, especially mesenteric desmoids are more common in patients with familial adenomatous polyposis.[2] Obstetricians encounter a miniscule number of these tumors during their career.

Most of the reported cases of abdominal wall desmoids were associated with previous trauma, prolonged estrogen intake, and pregnancy in young females.[3,4] These associations are correlated with detection of estrogen receptors in the substance of these tumors. Molecular studies demonstrated desmoid tumors in FAP as clonal neoplasms arising from germ line mutation or changes in the APC alleles.[5,6,7] Abdominal desmoid tumor usually presents as a mass that is sometimes associated with pain and weight loss.[8] The treatment of choice is surgical removal with broad 'clean' margins in order to reduce the expected high local recurrence rate of 25–50%.[9,10].

Case Scenario

A 24 years old woman, gravid 2 para1 living 1 with term gestation with previous LSCS came with c/o labour pains since 2 hours. On abdominal examination, Vertical infraumbilical scar healed by primary intention present. Uterus term size, Oblique lie. Head in right iliac fossa. FHS 110/min, regular. No scar Tenderness. An oval mass measuring 20*15 cm in the left iliac fossa, non tender, firm in consistency, superficial to peritoneum, with restricted mobility (? parietal wall tumour). Another cystic swelling of 2*2 cm is present in the healed scar, 7 cm above the pubic symphysis. On per vaginal examination, cervix was 2 cm dilated, 80% effaced, thick meconium stained liquor, presenting part was high up. Pelvis was clinically adequate.

She was taken up for emergency cesarean section in the view of previous

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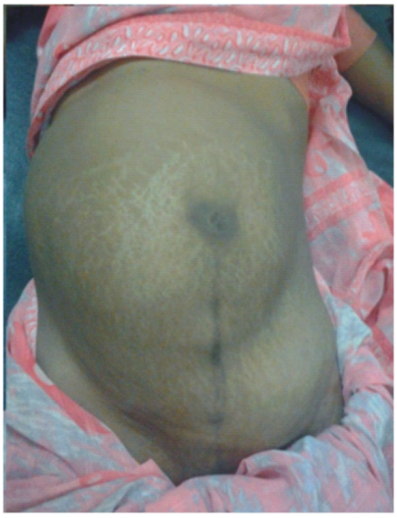
Figure 1: Preoperative



LSCS with fetal distress. Abdomen opened by infra umbilical vertical incision. Exposure to the operative field was restricted as the tumor was occupying left half of abdominal wall. Baby was extracted with difficulty. After

Figure 2: Preoperative

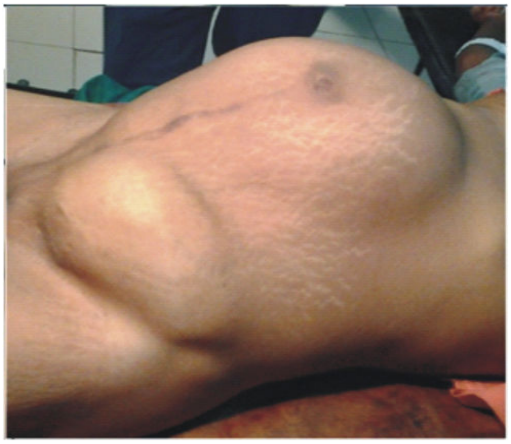


Figure 3: Preoperative

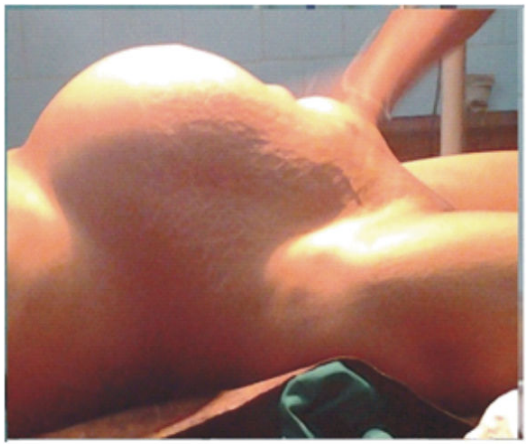


Figure 4: Intraoperative

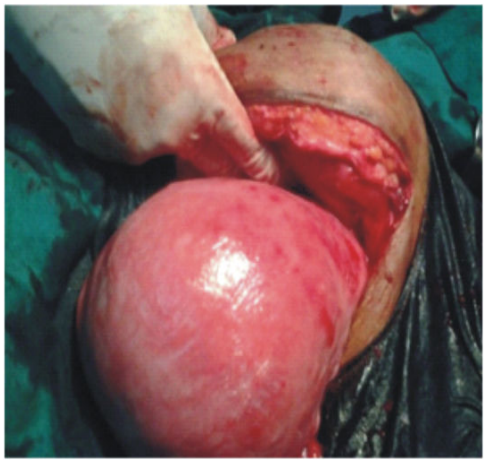
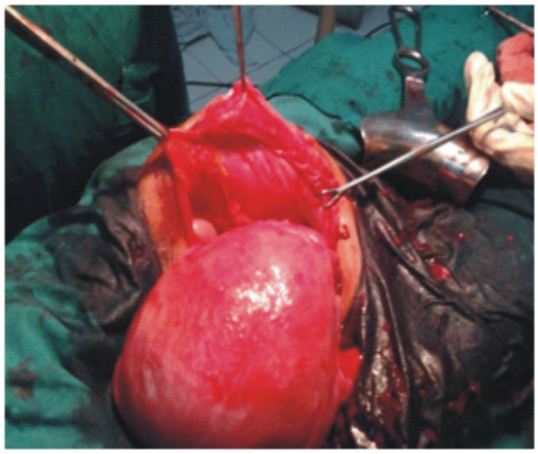


Figure 5: Intraoperative



uterine closure, the cystic mass in midline is excised for biopsy. The mass in left iliac fossa was in the plane of rectus sheath and rectus muscle.

Fine needle aspiration cytology of mass revealed many scattered tiny fragments of fibrillary amorphous material – collagen and

Figure 6: Histopathology

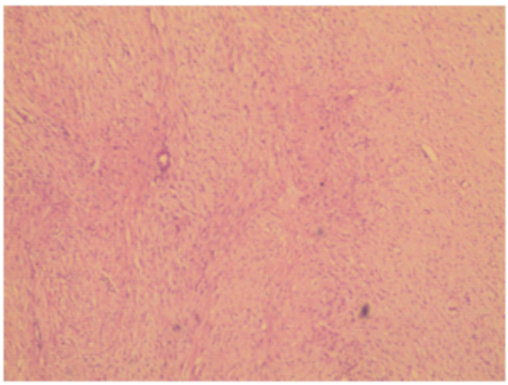


Figure 7: Histopathology

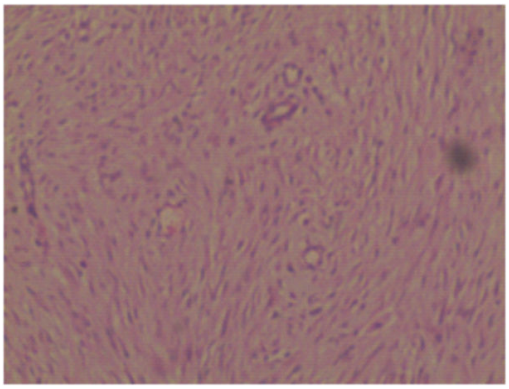
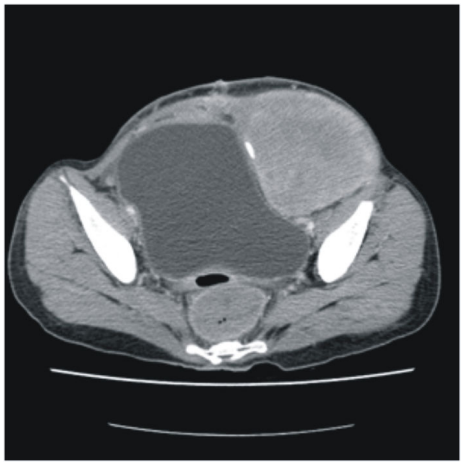


Figure 8: Computed Tomography



few spindle cells in groups. The cells have elongated nucleus, scanty cytoplasm with cytoplasmic processes, suggestive of Fibromatosis. The histopathological examination of excised specimen revealed fibromatosis CT scan of abdomen & pelvis

Figure 9: Computed Tomography

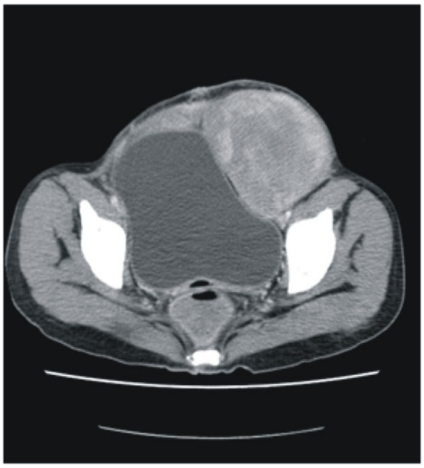


Figure 10: Histopathology

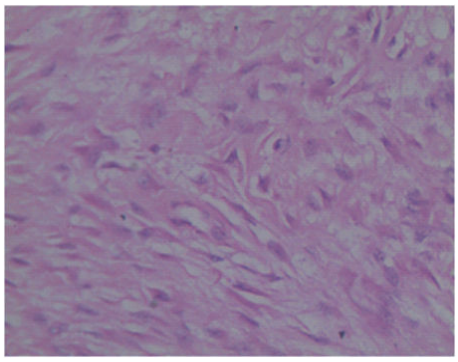
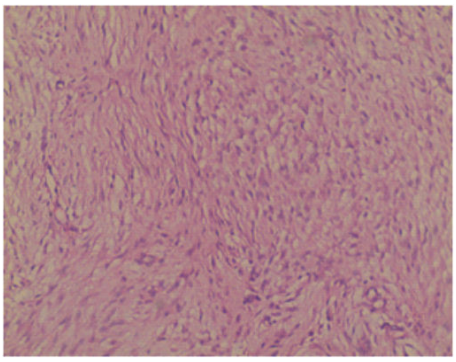


Figure 11: Histopathology



shows a well defined solid enhancing mass lesion in the lower anterior abdominal wall measuring 20*17 cm, in the intramuscular plane, suggestive of desmoid tumour.

Post op period was uneventful. Three months post partum she underwent complete surgical excision of 22*20 cm tumor and immediate reconstruction with Gortex mesh. Post operative period was uneventful. There is no recurrence in 5 months follow up.

Discussion

This case report is interesting and unusual because, the imaging techniques used initially reported the possibility of a subserosal leiomyoma as the primary diagnosis rather than a desmoid tumor, because some sections in imaging had shown very close attachment of the tumor to the uterus. This case emphasizes the importance of clinical examination and a multidisciplinary approach to ensure the best outcome for the patient. The majority of large pelvic masses in female

patients represent commonly encountered entities, such as uterine fibroid tumors, dermoid tumors, ovarian cysts, and ovarian cancer. However, uncommon pelvic masses, such as mesothelioma, adenosarcoma, carcinosarcoma, leiomyosarcoma, and desmoid tumor, may also be encountered. Clinical symptoms are masked by slow growth of tumour.[9]

Surgery is the treatment of choice for sporadic abdominal desmoid tumors. Wide radical local excision with safety margins of at least 2–3 cm is mandatory. Complete resection is often challenging, especially with vascular and neural involvement. Frequently, a sizable defect in the abdominal wall is formed mandating mesh reconstruction.[10] In spite of complete macroscopic resection, desmoid tumors are notoriously known for their high recurrence rate. Thomas[11] found no recurrence. Shao *et al*[12] reported a recurrence rate of 5.5% in a large series in China of 42 abdominal wall desmoids.

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