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Case Report

A Parietal Wall Desmoid Tumour with Primary Mesh Repair: A Case Report

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Abstract

Desmoid tumours are rare benign tumors which are locally aggressive. They develop from muscular aponeurosis. They are mostly sporadic, usually developing from a previous surgical scar. We report a case of 24 year old lady who presented with an abdominal mass of 2 years duration, following her caesarean section 2 years back. The tumor was excised in toto with parietal wall and was reconstructed subsequently with mesh. Patient recovered well with no complications. Desmoid tumours are rare mesenchymal neoplasms which are occasionally associated with FAP/ Gardeners syndrome. Treatment includes complete resection. If adjacent structures/ parietal wall are involved, then en bloc resection with apt reconstruction with biological and/or synthetic mesh should be performed. Regular follow up is of importance as these tumors are notorious for local recurrence.

Key words: Abdominal wall, Desmoid Tumour, Prolene mesh.

Introduction

Desmoid tumours, also called aggressive fibromatosis, are rare but locally invasive and non-metastasizing mesenchymal neoplasms. They arise from different types of connective tissue including muscles, fascia and aponeuroses. Based on their location, they are categorized as abdominal, extra-abdominal or mesenteric.[1] Most of the tumors are sporadic, but 10-15% of them are associated with APC mutation associated with FAP, Gardeners and Turcot's syndrome. They present as long-standing masses with no or minimal local symptoms, like pain due to compression due to surrounding structures. Diagnosis of these tumours is by through clinical examination and radiological investigation. Different modalities have been used, including surgical resection, radiotherapy, anti-inflammatory agents, hormonal therapy, and chemotherapy.[2] Here we discuss about the rare presentation of desmoid tumour post surgery and progression of tumour.

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Case report

A 24-year-old lady presented in our OPD with a mass over the abdomen for a duration of 2 years which she noticed following a caesarean section. The mass gradually increased in size, and was associated with occasional non-specific pain. There was no history of altered bowel habits, loss of weight or appetite or other constitutional symptoms. Her past history and family history were unremarkable. On examination, a 7x5 cm, firm mass was palpable in the left iliac fossa at the edge of previous surgical scar. It was non tender and showed restricted mobility. Regular haematological tests were within normal limits. Fig 1 shows mass noted in left iliac fossa.

MRI of the abdomen revealed a well-defined, oval T1 isointense & T2 /STR heterogeneously hypertense mass lesion in intramuscular plane of left later anterior abdominal wall measuring 4.6x 5.9x4.5cm. The mass was seen anterior to rectus abdominus. The external oblique muscle was seen stretched over the mass anteriorly. Internal oblique & transverse abdominus muscle were not visualised separately from the mass. Preoperative diagnosis was a giant abdominal cystic tumour. Differential diagnosis was gastrointestinal stromal tumour (GIST), DT and lymphoma.Surgery was performed for definitive diagnosis and improvement of the symptoms.

Operative findings

Lower pfannenstiel incision was preferred to observe large tumour. Intraoperatively, the tumour was found invading the external oblique, rectus sheath and transverse abdominus muscle. Fig 2 shows intraoperative appearance of tumor. It was excised with a margin of 1 cm all around. The resultant defect in the abdominal wall was closed with a prolene mesh of 15x8 cm, which was placed posteriorly. Fig 3 Gross appearance revealed grey white well circumscribed mass of 10x 5cms. All margins were negative for tumour. Specimen consists of single grey white to grey brown globular mass measuring 11x7x6cms. On external surface capsule is intact. On multiple cut sections pearly white solid areas are seen. Section studied from the tumour tissue shows uniform cellularity with bipolar fibroblasts having bland nucleus. Stroma is collagenous with focal areas of myxoid change. The blood vessels were thin walled, non-branching and compressed. Margins were negative. Histopathological features were suggestive of Desmoid tumour. Postoperatively no wound infection was noted. Patient was discharged on 12th day and was advised for regular follow up.

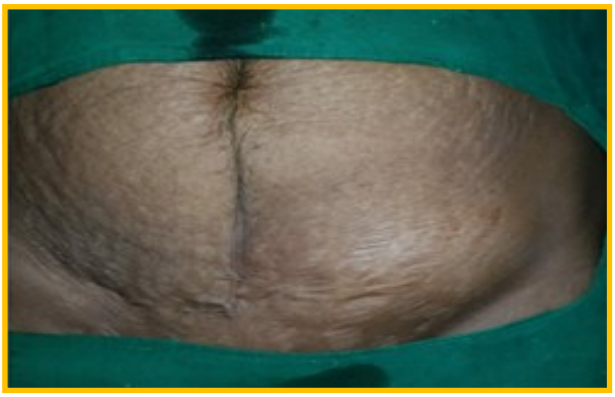

Fig 1. Mass noted in left iliac fossa

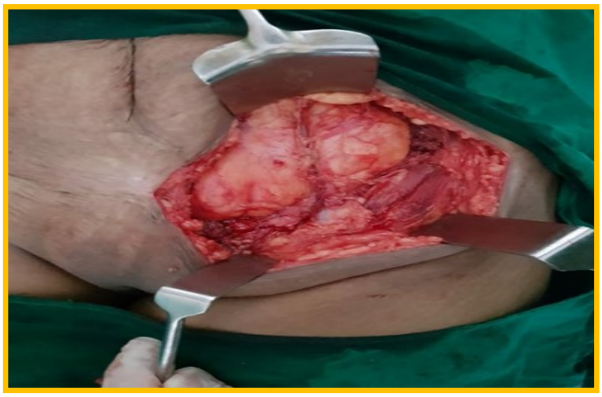

Fig 2. Intraoperative findings of the desmoid tumour

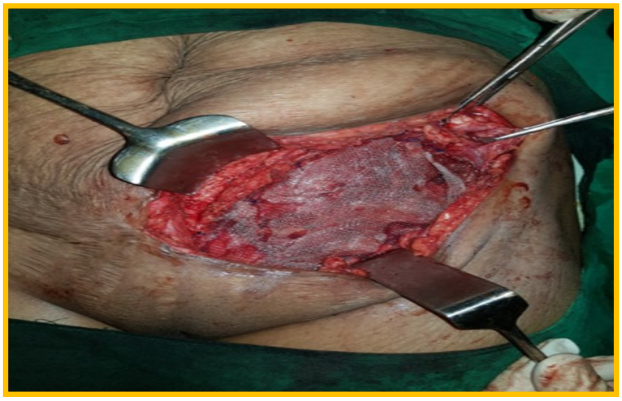

Fig 3. Intraoperative placement of mesh

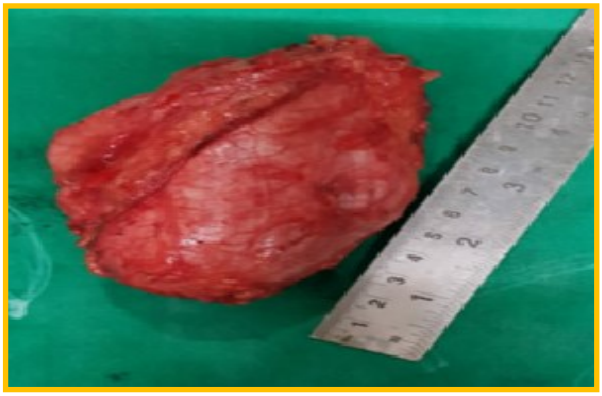

Fig 4. Macroscopic view of excised desmoid tumour

Discussion

Desmoid Tumors are benign, deep-seated monoclonal myofibroblast neoplasms that slowly grow, infiltrate, and arise from musculoaponeurotic stromal elements. They account for only 0.03% of all neoplasms and less than 3% of all soft tissue tumours.[1] They arise from any part of the body extra-abdominal (trunk and extremities), along the abdominal wall, and least commonly, intra-abdominally. Females appear more prone to develop this condition, with a female-to-male ratio as high as 3:1. [2] Desmoid tumours have a tendency for local recurrence after incomplete excision and for accelerated growth after trauma.[3] Etiological factors leading to desmoid tumours are surgical trauma, pregnancy, oral contraceptives, and a family history of desmoid tumours.[4] They have also been associated with 10% to 15% of patients with FAP, gardeners syndrome. [5]

FAP increases the risk from 825 to 1000 times to develop a desmoid tumor compared to the general population.[6] 10–15% of desmoid tumors are associated with germline mutations of APC.[6] Gurbuz et al. noted that out of 825 patients with FAP, 83 patients (10%) were found to have desmoid tumours, many of which were found to be in close proximity to previous surgical trauma. Patients are usually asymptomatic, but patients with huge Intra-abdominal large DT can cause severe pain, intestinal obstruction, and compression around the adjacent structures. [6, 7] Imaging studies, such as CT and magnetic resonance imaging (MRI), are used for preoperative diagnosis and for the planning of the surgery. Following surgery, CT and MRI are used for detecting recurrence.

Regarding treatment, the optimal therapy for DTs is also difficult to ascertain. Surgical resection, radiotherapy, and pharmacologic treatments have all been used, and no one treatment is currently considered a gold standard.[6] Widely used treatment of abdominal wall DT consists in a wide and radical resection with negative margins.[6]The surgical approach may require different techniques to repair the parietal defect including prosthetic material such as synthetic or biologic meshes.[6] Our patient presented with mass initially and later the mass had compressive features around the structures surrounding structures. Complete resection of the tumour was done with blunt dissection. The defect corrected by placement of prolene mesh. The addition of a mesh diminishes intestinal protrusion and generates a long-term strength. Gomes et al. described a parietal reconstruction after surgical resection of a desmoid tumour in a 41-year-old female patient, localized at the upper and medium third of the left recti abdominis muscle. They resected the entire muscle segment and inserted a polypropylene mesh. [7] Other modalities include radiotherapy and chemotherapy. Radiotherapy is indicated for inoperable patients, multiple recurrences or in case of incomplete resection. Chemotherapy (vinblastine, methotrexate, anthracycline) is hardly ever used. [8,9]

Conclusion

Desmoid tumours are common in young female , mostly associated with previous surgical scar. Different modalities are available for treatment of desmoid tumours but the most preferred one is the complete resection of the tumour.

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