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DESMOID TUMOR OF THE ANTERIOR ABDOMINAL WALL IN A YOUNG FEMALE: A RARE CASE REPORT

Pathology

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ABSTRACT

Desmoid tumours are rare lesions which has a strong tendency to invade locally and to recur. They constitute 3% of all soft tissue tumours and 0.03% of all neoplasms. These tumours are commonly seen in women of fertile age, especially during and after pregnancy. We report a case of thirty year old female patient presenting with a painless mass in the anterior abdominal wall with past history of abdominal surgery 8 years back. Ultrasonography revealed a solid, heterogenous hypoechoic lesion within abdominal wall. On Fine needle aspiration cytology of the mass, spindle cell tumour suggestive of desmoid tumour was diagnosed. Wide local excision of the affected abdominal wall mass was performed and the defect was replaced with a polypropylene mesh. The specimen was sent for histopathological examination where cytological diagnosis was confirmed as desmoid tumour of anterior abdominal wall.

KEYWORDS

desmoid tumour, spindle cell tumour, anterior abdominal

INTRODUCTION

Desmoid tumours (also called desmoids fibromatosis) are rare clonal fibroblastic proliferations that arise in the deep soft tissues and are characterised by infiltrative growth and a tendency towards local recurrence but an inability to metastatize. [1]The term desmoids was coined by Muller in 1838 and is derived from the Greek word *desmos,* which means tendon like. These tumours often appear as infiltrative, usually well differentiated and locally aggressive in nature. It is thought that the lesions may develop in relation to estrogen levels or trauma or operations. They constitute 3% of all soft tissue tumours and 0.03% of all neoplasms. [2] Abdominal wall lesions typically arise in young, gravid or parous women during gestation or more frequently during the first year following childbirth[1] and are uncommon during the menopause. During pregnancy, an increase in volume occasionally occurs in already existing tumours. This corroborates the estrogen-stimulated tumor growth hypothesis. [3]The most common site of occurrence of desmoids is the anterior abdominal wall with an incidence of 50%.[4]

CASE REPORT

A thirty year old female patient presented with a painless mass in the anterior abdomen since ten months which was gradually increasing in size. The patient had past history of abdominal surgery 8 years back. There was no other significant Family history and personal history.The general examination and all the systemic examination were normal. On local examination ,a single lower abdominal lump involving left umbilical region was present (Figure 1).

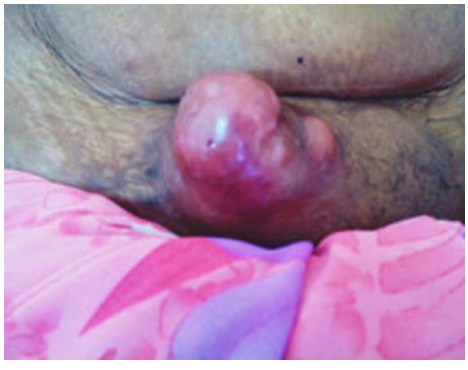

Figure 1: A Single Lower Abdominal Lump Involving Left Umbilical Region

Lump was fixed to the anterior abdominal wall, nontender, globular in shape and approximately 5 × 4.5 × 4.2 cm of size with smooth surface, and firm in consistency. The routine haematological and biochemical investigations were within normal limits. Ultrasonography demonstrated a large solid heterogeneous hypo echoic lesion showing internal vascularity with few anechoic areas of necrosis. With written informed consent Fine needle aspiration cytology was done. The slides were fixed in methanol and stained with haematoxylin and eosin stain .The cytological study shows clusters and short fascicles of oval to spindle shaped cells with pleomorphic hyperchromatic nuclei and scanty eosinophilic cytoplasm against stromal fragments and haemorrhagic background (figure 2).

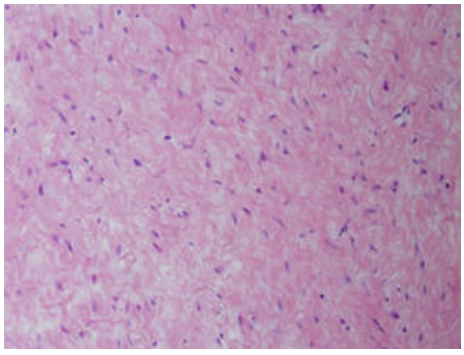

Figure 2: A Cytological Presentation Of FNAC Sample

The diagnosis was given as spindle cell soft tissue lesion suggestive of desmoid tumour. After preoperative workup, patient was planned for surgical management and complete excision of the tumor with wide surgical margins of 3 cm healthy tissue. The defect was reconstructed with polypropylene mesh. The excised specimen of wide local excision was sent to pathology department for histopathological examination. On gross examination, we received specimen totally measuring of size 9x4x4 cm. (figure3).

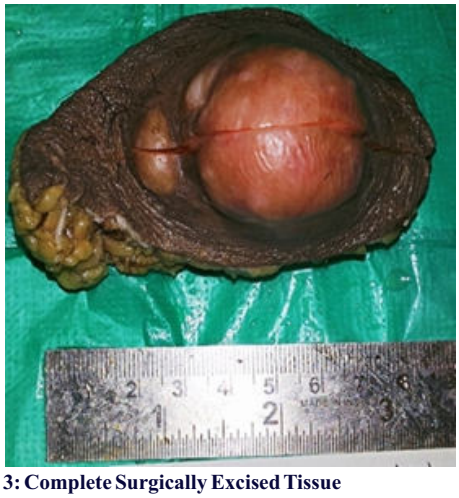

Figure 3: Complete Surgically Excised Tissue

Skin tag of size 9x4 cm.On external surface it was single large globular mass of total size 4x4.5 cm.On the cut surface, it was glistening white and coarsely trabeculated, resembling scar tissue(figure 4).

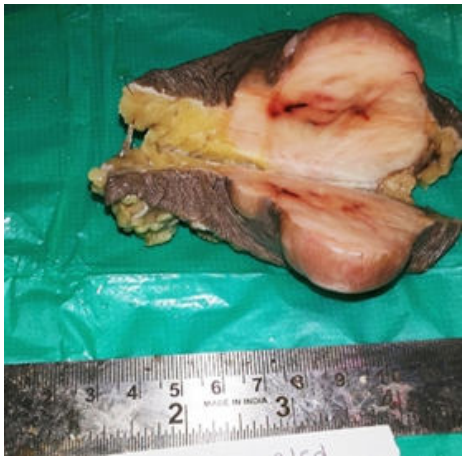

Figure 4: Cut Section Of Sample

On histopathology, sections show lining of stratified squamous epithelium underneath seen tumour tissue comprising of proliferation of fibroblastic cells, spindle shaped cells & at places small round to oval cells with hyperchromatic pleomorphic nuclei & scanty eosinophilic cytoplasm arranged in sweeping collagen bundles, fascicles & in trabecular pattern.(figure5)

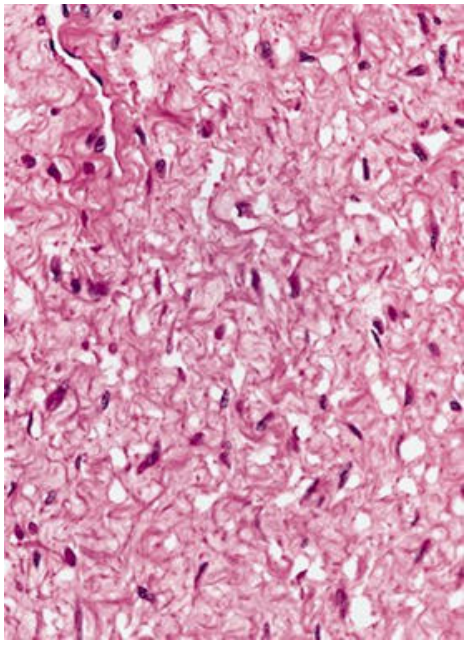

Figure 5: Histoplathological Presentation Of The Lesion

The diagnosis was given as desmoid tumour. Thus cytological findings were confirmed on histopathology.

DISCUSSION:

Desmoid tumours can occur throughout the body in any skeletal muscle, but are most commonly seen to arise from rectus abdominis in postpartum women. Desmoid tumor arises from myofibroblast, lacks a true capsule, and usually infiltrates into adjacent muscle bundles. The telomerase length and activity is normal, [5] nuclei are small and regular, and mitoses are infrequent all of which support its histologically benign nature. Despite the benign histologic character, their biological behaviour is more malignant, since the infiltrative pattern of growth can ultimately lead to life threatening visceral involvement and even cause death. [4]Surgical trauma has been intimated in 68.86% of abdominal and intra-abdominal desmoids. [4]Role of an endocrine etiology in the occurrence of desmoid tumor has also been suggested. The commonest groups associated with these tumours are young women during or after pregnancy. The fibroblast has been shown to exhibit a proliferative response to estrogen. Women with desmoid tumours have regression of their lesions after attaining menopause. [6]Additionally, desmoids regress on tamoxifen and oralestrogen therapy. The prevalence of desmoid tumor in FAP is 10.25%.[9] They can be divided into five subgroups: extra abdominal,intra-abdominal, multiple, multiple familial and as a part of Gardner's syndrome Abdominal desmoids arise from musculoaponeurotic structures of the abdominal wall, especially the rectus and internal oblique muscles and their fascial coverings, and occasionally cross the midline. Less commonly, they originate from the external oblique muscle and the transversalis muscle or fascia. [10] There is a well-known association in patients with a history of abdominal or pelvic surgery. Abdominal desmoid tumour usually presents as a mass that is sometimes associated with pain and weight loss. Most of the abdominal wall desmoids measure 5 cm by 15 cm in dimensions. Our patient presented with a painless mass measuring 4x4cm in dimension. These masses have a firm, gritty texture. On the cut surface, they are glistening white and coarsely trabeculated, resembling scar tissue. These tumours have no distinct capsule, and their margins are ill defined even when they appear well circumscribed on imaging.[10] The differential diagnoses for abdominal desmoid include acute hematoma fibrosarcoma, lymphoma, rhabdomyosarcoma, liposarcoma, leiomyosarcoma, neurofibroma, benign fibrous tumour and primitive neuroectodermal tumor. On histopathological examination these lesions are typically poorly circumscribed with infiltration of the surrounding soft tissue structures. All are characterized by a proliferation of elongated, slender, spindle-shaped cells of uniform appearance, set in a collagenous stroma containing variably prominent vessels, sometimes with perivascular oedema. The cells lack hyperchromasia or atypia and have small, pale-staining nuclei with 1 to 3 minute nucleoli. Cells are usually arranged in sweeping bundles.[1]These tumours show a tendency to evolve over time.On ultrasonography, desmoid tumours appear as well defined lesions with variable echogenicity. The lateral borders may appear ill defined or irregular[10].Definitive diagnosis must be established with histopathologic analysis.Wide local excision followed by reconstruction of the defect is the treatment of choice.

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