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Case Report

A Rare Case of Large Desmoid Tumour of anterior Abdominal Wall in a Young Female

B. Ananda RamaRao, M. Siddartha Reddy, Ramachandra Reddy, S. Vinay Kumar

Dept. of General Surgery, SVS Medical College and Hospital, Yenugonda, Mahabubnagar, Telangana

Corresponding Author: B. Ananda RamaRao

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ABSTRACT

Desmoid tumors (also called desmoids fibromatosis) are rare slow growing benign and musculo-aponeurotic tumors originating from fascia and musculoaponeurosis with an infiltrating growth. Although these tumors have a propensity to invade surrounding tissues, they are not malignant. These tumors are associated with women of fertile age, especially during and after pregnancy more common between 25 to 40 yrs. We report a young female patient with a giant desmoid tumor of the anterior abdominal wall who underwent primary resection. The patient has a history of an earlier abdominal surgery. Preoperative evaluation included abdominal ultrasound, computed tomography, and magnetic resonance imaging. The histology revealed a desmoid tumor. Primary surgical resection with immediate reconstruction of abdominal defect is the best management of this rarity.

Keywords: Desmoids, Benign, Locally aggressive, Pregnancy, Anterior abdominal wall.

INTRODUCTION

Desmoid tumors (also called desmoids fibromatosis) are rare slow growing benign fibrous tumors arising from fascia and muscle aponeurosis with a strong tendency to infiltrate locally and to recur. [1] The term desmoids was coined by Muller in 1838 and is derived from the Greek word desmos, which means tendon like. These tumors often appear as infiltrative, usually well differentiated and locally aggressive in nature and also known as deep fibromatoses. [2] Desmoid tumors may be classified as extra-abdominal, abdominal wall, or intra-abdominal (the last is more common in patients with familial adenomatous polyposis (FAP)). [3] It is thought that the lesions may develop in relation to estrogen levels or trauma/operations. They constitute 3% of all soft tissue tumors and 0.03% of all neoplasms. [4] These tumors are associated with women of fertile age, especially during and after pregnancy. The most common site of occurrence of desmoids is the anterior abdominal wall, with an incidence of 50%.

CASE REPORT

A 25 year old female presented with a complaint of painless swelling in the lower abdomen since 10 months which was gradually increasing in size. H/o caesarean section in the past. No history of abdominal trauma in the past.

O/e a single non tender 10*10cms globular, lower abdominal swelling involving the left lumbar, left iliac, hypogastrium, fixed to the anterior abdominal wall, hard in consistency with a smooth surface.

Ultrasound demonstrated a large solid heterogenous, hypoechoic mass showing internal vascularity localized to the anterior abdominal wall below the umbilicus in the hypogastrium and left iliac fossa.

CT scan revealed single large well defined circumscribed isodense mass lesion measuring 10* 9 cms noted arising from the anterior abdominal wall in the pelvis on the left side extending posteriorly in to the abdominal cavity and abutting the anterior wall of urinary bladder showing no involvement of organs.

After preoperative workup, patient was planned for operation, and complete excision of the tumor with wide surgical margins along with the anterior abdominal wall down to the peritoneum was performed, resulting in a large wall defect of about 15 × 15 cms, which was repaired after mobilization and release of rectus abdominis and reconstructed with polypropylene + Monocryl mesh. Skin was closed after keeping vacuum suction drain beneath the subcutaneous space. The postoperative course was uneventful, and patient was discharged on the 9th postoperative day. Histopathological reports were consistent with desmoid tumor with negative surgical margins. At 8 months of follow-up, patient did not have any recurrence or incisional hernia.

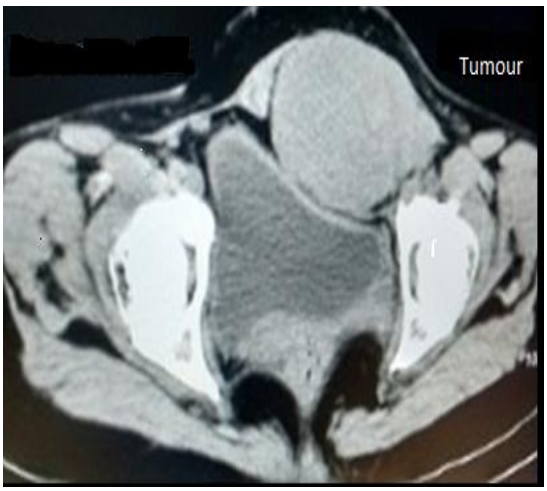

Figure: 1- CT Scan

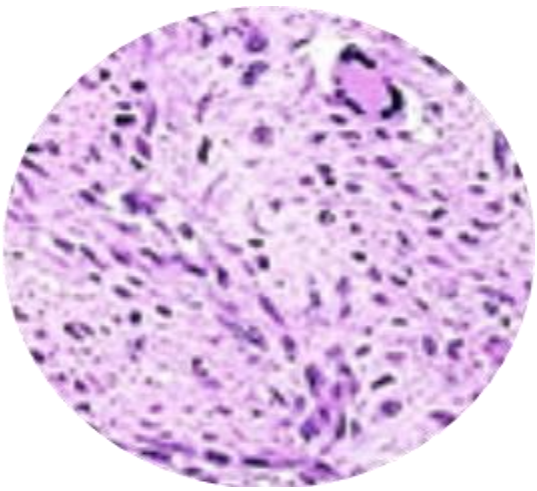

Figure: 2 – Histopathology exam

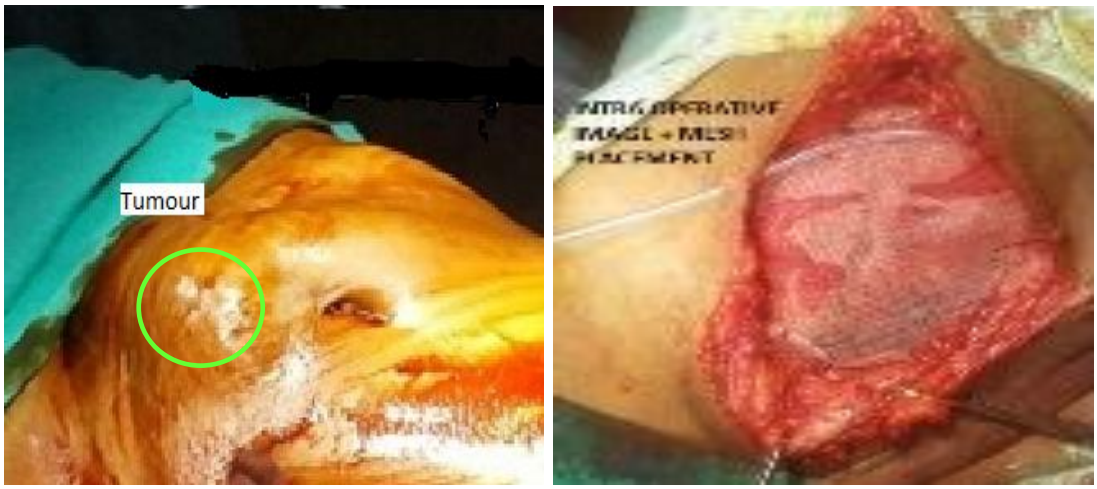

Figure: 3 - Preoperative And Intraoperative Pictures With Insitu Mesh

DISCUSSION

Desmoid tumors are cytologically benign fibrous neoplasm originating from

the musculoaponeurotic structures throughout the body.

Desmoid tumors are benign deep aggressive fibromatoses, originating from fascia and muscle aponeurosis with an infiltrating growth. Approximately, 3.7 new cases occur per one million persons per year and often associated with female gender, FAP, estrogen therapy, and occasionally with surgical trauma (1 in 4 cases). Trauma especially operative trauma may contribute to the formation of desmoid tumors. [5] Estrogen may act as a growth factor. Endogenous estrogen levels have a close correlation with tumor growth factor rate. It has a higher prevalence in young women who experienced pregnancy, [7] and rare in males. They may be extra-abdominal (the shoulder girdle, trunk, and lower extremities), intra-abdominal (in the abdominal wall, especially the rectus and internal oblique muscles with their fascial coverings, and mesentery or retroperitoneum), multiple familial, and as part of Gardner's syndrome.

Abdominal desmoid tumor usually presents as a firm mass with ill-defined margins and no distinct capsule. [7] On cut surface, they are gritty, glistening white and trabeculated resembling scar tissue. Histologically, desmoid tumors consist of elongated fibroblasts and myofibroblasts. [8]

On ultrasonography, desmoid tumors appear as well-defined lesions with variable echogenicity. On CT, they may appear as homogeneous or heterogeneous and hypo-, iso-, or hyperintense compared with the attenuation of muscles. [9] Characteristic MRI findings include poor margination, low-signal intensity on T1-weighted images and heterogeneity on T2-weighted images, and variable contrast enhancement. [10] MRI is superior to CT scanning in defining the pattern and the extent of involvement as well as in determining if recurrence has occurred after surgery, though both the modality CT and MRI aid in determining the extent of local invasion.

Wide local excision with reconstruction of the defect is the treatment of choice. Peritoneum, intraperitoneal organs, or adjacent bony structures involved by tumor must be resected as well. Incomplete tumor removal or positive surgical margins may lead to local recurrence, [11] (20% to 77% depending on the location, extent, and completeness of the initial resection). Abdominal wall desmoid tumors have a significantly lower recurrence rate (20% to 30%) and usually become evident within six months after excision. Radiotherapy, chemotherapy, and endocrine therapy are used in patients with inoperable tumors, local recurrences, or incompletely excised lesions. Metastatic disease has not been reported with desmoid tumor. Only few cases of malignant transformation in desmoid tumor are reported, and all were associated with local irradiation. Sulindac, indomethacin, [12] and tamoxifen along with varieties of antineoplastic medications have been used to treat these patients with variable results.

CONCLUSION

The history of painless abdominal mass, the age and sex of the patient, the location of the mass within the anterior abdominal wall, and the imaging features make desmoid tumor a strong primary diagnostic consideration even if it is a rare entity. Aggressive, wide surgical resection with negative margin is the best surgical option. Complete surgical excision of desmoid tumors is the most effective method of cure, sometimes necessitating removal of most of an involved anterior abdominal wall in such a giant desmoid tumor with immediate repair of resultant huge defect and reconstruction using prosthetic mesh for better functional results.

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