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CASE REPORT

DESMOID TUMOR OF ANTERIOR ABDOMINAL WALL: A CASE REPORT

***[1]Dr. Fazal Ameen, [2]Dr. Maria Ahmad, [3]Dr. Khurshid Samo, [3]Dr. Sumayya Saeed and [3]Dr. Amjed Siraj Memon**

[1]General Surgery, Civil Hospital Karachi/Dow University of Health Sciences
[2]Dow University of Health Sciences
[3]General Surgery, Dow University of Health Sciences

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ABSTRACT

Desmoid tumors are non metastatic, rare tumors with a tendency of local invasion and recurrence. These lesions are associated with female gender in their fertile age typically appearing during or after pregnancy. The case of a desmoid tumor of anterior abdominal wall in a 22 year old lady is reported, that presented during pregnancy, describing its CT and MRI appearance. The lesion was treated surgically, reconstructing the defect with polypropylene mesh and was referred to oncology department for further management. The goal of treatment for these lesions is complete surgical excision, and in case where negative margins are not achieved, radiotherapy is the next modality.

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INTRODUCTION

Desmoid tumors are locally aggressive but non-metastatic, typically well differentiated benign tumor of myofibroblastic cell origin, arising from muscular aponeuroses that are also called as deep fibromatoses. The reported incidence for desmoid tumors are about 0.03% of all neoplasms, whereas with familial polyposis of the colon, the prevalence of these tumors can be as high as 25%. The most common site for origin of this tumor is anterior abdominal wall, with incidence of up to 50%. These tumors usually occur in the women of childbearing age and less common after menopause supporting the hypothesis of estrogen stimulated tumor growth. Surgical excision has been the primary modality for treatment of these lesions but with a high rate of recurrence. Post-operative radiotherapy reduces the local recurrence rate if surgical margins are involved. We report a case of a female who developed the lesion during pregnancy, and surgical excision was performed after she delivered her baby.

CASE

A 22 years old female consulted surgical outpatient department for a swelling in right lower abdomen for 1 year.

**Coresponding author: Dr. Fazal Ameen*,
General Surgery, Civil Hospital Karachi/Dow University of Health Sciences.

She noticed the swelling about 1 year back when she was pregnant with her second child. It was a painless swelling that progressively increased in size. She had delivered her baby through a LSCS 4 months before presenting to us and had a smooth recovery. There was no history of OCP use or weight loss. On examination, a diffuse swelling of about 13x10 cm was palpable in right lower abdomen extending up to inguinal region, which was hard, immobile, not attached to the overlying skin. No restriction of movements was noted below the hip and distal pulses were palpable. CT guided FNAC was performed which reported a benign spindle cell lesion with features favoring desmoid fibromatosis. Color doppler imaging of the lesion showed intralesional vascularity with RI 0.59, and right external iliac vessels adjacent to lesion were intact, and showed normal flow, waveform and velocity. CT scan showed a well-defined minimally enhancing mass in right lower abdominal wall measuring 8.7x11.2x7.8cm involving anterior abdominal wall muscles, invading external oblique, internal oblique and transverse abdominis muscles and rectus sheath. It extended into the peritoneal cavity, indenting urinary bladder and abutting the anterior wall of uterus and posterosuperiorly displacing the bowel loops. The mass partially abutted the external iliac vessels with partial loss of intervening fat planes. MRI showed a large well defined abnormal signal intensity mass involving rectus abdominis, external and internal oblique muscles on right side and extending anteriorly into the

subcutaneous soft tissues. It appeared isointense on T1W1 and intermediate on T2W1. Laterally it closely abutted the right external iliac vessels and iliacus muscles with loss of intervening fat planes. The patient was operated and, after safeguarding external iliac vessels, mass was completely excised. The defect was reconstructed with Polypropylene mesh. Microscopic description of the tissue revealed spindle cells showing fascicular and random arrangement, having elongated partly wavy nuclei with inconspicuous nucleoli, accompanied with perivascular myxoid change. The cells were positive for ASMA and beta-catenin immunostain. No evidence of malignancy was seen. The excision margins were positive, so the patient was referred to oncology department for further management.

DISCUSSION

Fibromatosis are of two types based on their location in the body: Superficial and deep. Desmoid tumor, also known as aggressive fibromatosis or deep fibromatosis, is a rare benign tumor with no metastatic potential of myofibroblast cells with an annual incidence of about 3.7 new cases per million and accounts for 0.03% of all malignancies (Kasper *et al*., 2011; Economou *et al*., 2011; Overhaus *et al*., 2003). It commonly occurs in women and in age group of 25 – 40 years, supporting estrogen stimulated growth (Economou *et al*., 2011). The cause of the desmoid tumors is unknown in most of the cases, however an association with estrogen, pregnancy, FAP (Gardner's syndrome) and surgical trauma has been documented. 10 - 25% of patients with FAP develop desmoid tumor and mostly in intra-abdominal location as compared to extra-abdominal location in general population (Sinha *et al*., 2011; Kumar *et al*., 2009). Family history, female sex, previous abdominal surgery and an APC mutation have been implicated as risk factors in FAP (Overhaus *et al*., 2003; Kumar *et al*., 2009; Escobar *et al*., 2012). These tumors may be extra-abdominal (shoulder girdle, trunk and lower extremities), intra-abdominal (in the abdominal wall especially the rectus and internal oblique muscles and their fascial coverings, and occasionally cross the midline, mesentery or retroperitoneum), multiple and as a part of Gardner's syndrome. The most common site of the occurrence of desmoid tumor is anterior abdominal wall with an incidence of up to 50%, often presenting as a firm mass with ill-defined margins (Economou *et al*., 2011). These tumors arise from the fascia or aponeurosis of muscles. Histologically, the tumor is composed of linearly arranged stellate to spindle shaped cells with keloid-like collagen separating them, in a poorly circumscribed pattern, (Economou *et al*., 2011; Escobar *et al*., 2012). Various imaging techniques are used to establish the diagnosis of these tumors. On ultrasonography these tumors have variable echogenicity with well-defined margins. (Economou *et al*., 2011; Atul *et al*., 2011). CT and MRI are needed to define the extent and invasion of the tumor and its relationship with the surrounding structures, (Escobar *et al*., 2012). A CT scoring system has been developed, characterizing specimens according to the presence of desmoid precursor lesions ('mesenteric fibrosis') and true Desmoid Tumors, providing an evidence for a stepwise progression in desmoid development.[8] They may appear homogenous or heterogenous on CT scan, depending upon their composition, with variable enhancement after administration of contrast. MRI findings of these tumors are also variable which include poor margination, low-signal intensity on T1-weighted images and heterogeneity on T2-weighted images, and variable contrast enhancement (Economou *et al*., 2011). MRI is superior to CT scan in defining the pattern and the extent of involvement as well as in determining a recurrent disease after surgery. Though both the modalities aid in diagnosing and determining the extent of local invasion but histopathology is required to confirm the diagnosis. (Escobar *et al*., 2012) Surgery (wide local excision) with negative surgical margins followed by reconstruction of the defect is the most successful primary treatment modality for desmoid tumors (Escobar *et al*., 2012; Latchford *et al*., 2006). As these tumors are locally infiltrative, adjacent structures involved by the tumor must be resected as well, which may include bony structures or abdominal viscera. Surgical excision of intra-abdominal tumor is much more difficult and may need massive small bowl resection (Sturt *et al*., 2006). Other treatment options available are radiotherapy (pre-op, post-op or as a sole treatment option), anti-estrogen, prostaglandin inhibitors and chemotherapy, with variable response. Radiotherapy, chemotherapy and hormonal therapy can be used for unresectable, widely infiltrative, partially removed and recurrent tumors. These tumors are associated with high recurrence rates which, depending upon the location, local invasion and completeness of initial resection, is about 24% to 77%. Lamboley *et al*., 2012) Abdominal wall desmoid tumor has lower recurrence rate of about 20% to 30% usually recurring within six months after excision or in relation with subsequent pregnancy. (Kumar *et al*., 2009) Abdominal wall reconstruction can be done either by direct repair with sutures or by using prosthetic material (mesh) or by using musculocutaneous flap if the defect is extensive (Brenner and Rammelt, 2002).

Conclusion

Desmoid tumor is a rare locally aggressive, but non metastatic, benign tumor often presenting as painless anterior abdominal mass more commonly in young females during or after pregnancy. CT and MRI are used to establish a diagnosis but definite diagnosis requires histopathologic confirmation. Aggressive wide local excision with clear margin is the primary therapeutic modality, but with high recurrence rate. The reconstruction of abdominal wall defects may be achieved with prosthetic mesh. Unresectable or recurrent tumors should be treated with radiotherapy or chemotherapy.

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