Abdominal Wall Desmoid: A Case Report


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Case Report
Subject: Radio Diagnosis

Abstract:
Desmoid tumors are slow growing benign musculoaponeurotic tumors. They are commonly seen in women of reproductive age group, during or after pregnancy. We report a case of a young female who presented with painless swelling in the right lower quadrant of abdomen following child birth. Abdominal ultrasound, computed tomography and magnetic resonance imaging were performed which revealed desmoid tumor. Surgical resection was done successfully and the abdominal defect reconstructed.

Key Words: Benign, Desmoid Tumors, Extra-Abdominal, Fibromatosis, Recurrence.

Introduction:

Desmoid tumors, also called deep or aggressive fibromatosis, are rare benign musculoaponeurotic tumors. They are slow growing, locally aggressive tumors without any malignant potential [1,2]. Local recurrence rates of these tumors are as high as 87% [1]. The term desmoid was derived from the Greek word desmos, which means tendon like. The term desmoid was coined by Muller in 1938 [2]. Desmoid tumors constitute 0.03% of all neoplasms and 3% of all soft tissue tumors [2]. Incidence in general population is 2-4 cases per million per year with a peak incidence in 3rd and 4th decades with slight female preponderance [1,5,6]. They can be classified into extra-abdominal, abdominal wall and intra-abdominal types [2]. There is a well known association between extra-abdominal desmoids and Familial adenomatous polyposis and Gardner syndrome [1,2]. Pregnancy, trauma, prior surgery and oral contraceptive use are some of the risk factors for abdominal wall desmoids [1]. Estrogen is implicated as a stimulating growth factor. The most frequently affected muscles are rectus abdominis and internal oblique muscles [3].

Case Report

A 22 year old female presented to the department of surgery with a swelling in the right lumbar region since six months. The swelling was noticed after her last child birth which was six months back. It was a slow growing, painless swelling. There was no history of trauma or surgery. On examination, a solitary, firm, non tender mass with smooth margins measuring 12x8cm was found fixed to the right anterolateral abdominal wall. The analysed blood parameters were within the normal range. On ultrasonography, a large solid heterogeneously hypoechoic mass showing internal vascularity within the muscle planes of right lower quadrant of abdomen was found. The mass showed few areas of necrosis. Plain radiograph of abdomen showed a homogeneously radio opaque soft tissue density mass in right lower quadrant overhanging the right iliac crest. CT scan showed a large, well defined heterogeneously enhancing mass in the parietal wall layers of right lumbar region [figure 1]. There was no evidence of lymphadenopathy. MRI scan showed mixed intensity lesion seen on all sequences with focal areas of necrosis measuring 105x94x135mm in subcutaneous planes arising from right external oblique muscle compressing transversalis and internal oblique muscle. Small foci of hemorrhages were noted [figure 2]. MRI features were consistent with neoplastic anterior abdominal wall mass and was considered as desmoid. After preoperative work up, patient was planned for surgery and a wide local excision was done. The defect was closed with monomax and subcutaneous tissue closed after placing drain. Macroscopically the lesion was firm with coarsely trabeculated margins [figure 3]. The post operative course was uneventful. Histopathological examination revealed fascicles of fibroblastic spindle cells with increased nuclear cytoplasmic ratio, elongated nucleus and occasional mitotic activity suggestive of desmoid tumor with negative surgical margins.

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Figure 1: CT plain and contrast show heterogeneously enhancing mass in the soft tissues of parietal layers.

Figure 2: MRI coronal sections showing mixed intensity lesion arising from right external oblique muscle showing fascial tail sign and few areas of focal hemorrhages

Figure 3: Intraoperative picture of tumor showing excised specimen with coarsely trabaculated firm mass.

Discussion:
Musculoskeletal fibromatoses are a group of tumors constituted by fibroblastic and myofibroblastic proliferation which have similar pathological appearances. Usually they show infiltrative growth with frequent local recurrences but no malignant potential. World Health Organization committee for classification of soft tissue tumors in 2002 classified these tumors into superficial and deep types.

Superficial fibromatoses are palmar and plantar fibromatosis which are small and slow growing. Deep fibromatoses are usually larger and often enlarge rapidly. They include desmoid type and abdominal wall [4,5].

The disease has female predilection, however this tendency is observed among younger patients. In older patients they occur with equal distribution in both men and women [4]. The fibroblasts show proliferative response to estrogen and regression of...
lesions noted after menopause [6]. There is an association of desmoid tumors with prior abdominal or pelvic surgery, trauma, estrogen therapy, Familial adenomatous polyposis (FAP) and Gardner syndrome. These tumors are 1.8 times more common in females than in males. There are cases reported in pediatric patients, but they are rare [5]. The most common site is the anterior abdominal wall with an incidence of 50% [6,7]. Most cases are noted in the infraumbilical location [7]. The rectus abdominis and internal oblique muscles are commonly affected followed by external oblique and transversalis fascia [3] and occasionally cross the midline [2,6]. Most common locations of extra abdominal fibromatosis are shoulder, upper arm, chest wall, para spinal region, thigh and head and neck [1,3]. In 15% of cases multiple desmoids have been noted [1]. Intra abdominal desmoids are seen in mesentery or retroperitoneum. These are associated with Familial adenomatous polyposis or Gardner syndrome and are infiltrative and may cause intestinal or ureteric obstruction or encase mesenteric vessels. Prevalence of desmoid tumors in FAP is 10-25%. These desmoids are associated with adenomatous polyposis coli (APC) germline mutation which was first noted by Caspari et al, and Davies et al in 1995 and supported by many other studies [7]. Pelvic desmoids can infiltrate the urinary bladder or cause hydrosalpinx. Compression of adjacent structures like vessels or nerves are seen in extra-abdominal type. Rarely abscess formation is seen. Mucoid or cystic degeneration is noted in large desmoid tumors [1]. Radiographs are not routinely useful in imaging desmoid tumors [5]. Ultrasonography is helpful in the initial evaluation of these soft tissue masses. They appear as well defined lesions with variable echogenicity and have variable vascularity [1,2,5,6]. Large lesions may show posterior acoustic shadowing [4]. CT reveals a soft tissue mass of variable attenuation, usually similar in attenuation to that of skeletal muscle. More collagen containing lesions may show mildly higher attenuation compared to that of muscle. On CT, lower attenuation is least commonly seen which is associated with myxoid components. Indistinct margins may be seen because of infiltration of adjacent structures. Post contrast, the lesions demonstrate variable degree of enhancement. This enhancement reflects in capillary network that is frequently seen in desmoid type fibromatosis [1, 2, 4, 5,6]. Although CT and MRI aid in determining the extent of local invasion, MRI is superior to CT in defining the pattern and extent of involvement and in determining recurrence [2]. The signal intensity is variable and dependent on the extent of collagen and the degree of cellularity of lesion. There is poor margination, low signal intensity on T1, heterogeneity on T2 and variable contrast enhancement [2,6]. Three stage s of desmoid tumors are described. In the first stage, more cellular lesions with large extracellular spaces and relatively less collagen content are noted which appear as low signal intensity on T1 and increased signal intensity on T2. Second stage shows increased amount of collagen in central and peripheral areas of tumor which leads to increased heterogeneity of T2 signal. In the third stage, increased fibrous deposition, decreased cellularity, decreased extracellular spaces and water content causes decreased signal intensity on T1 and T2 MR images [3,4,5,6]. These lesions are usually located intermuscularly with a rim of fat (split fat sign). Linear extension along fascial planes (fascial tail sign) is a common manifestation seen in 80-83% cases [3].

Definitive diagnosis must be established with histopathological analysis [6]. Histologically, these tumors are composed of elongated fibroblasts and myofibroblasts with elongated, tapered cytoplasm, elongated, vesicular appearing nuclei and multiple small nucleoli. Cells are linearly arranged and separated by collagen [2,5,6]. The differential diagnosis of desmoid tumors are lymphoma, pleomorphic sarcoma, fiбро sarcoma and giant cell tumor of tendon sheath [1]. Wide local excision followed by reconstruction of the defect is the treatment of choice. Incomplete tumor removal or involved excision margins may lead to local recurrence [6]. Recurrences are more commonly noted in lesions more than 5cm in size [7]. Non surgical treatment includes radiation and systemic therapy. It is considered when resection is not possible because of close association with vital structures [1,2,5].

**Conclusion:**
Desmoid tumors are locally aggressive with a strong tendency for recurrence. MRI Features along with age, sex, clinical history and location of the tumor aid in diagnosis. However definitive diagnosis is based on histopathology. Wide local excision is the definitive treatment.

**References:**


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