IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)
e-ISSN: 2279-0853, p-ISSN: 2279-0861.Volume 15, Issue 6 Ver. XIII (June. 2016), PP 98-102
www.iosrjournals.org

Desmoid Tumor of the Anterior Abdominal Wall in A Young Woman: A Case Report

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Abstract: *Desmoid tumors are uncommon benign neoplasms but they aggressively invade the surrounding tissue, making it difficult to remove them surgically. Even after an apparent complete excision they often tend to recur but have no metastatic potential.They arise from fibro aponeurotic tissue and present as a slow growing mass. Most common symptom is pain. Other signs and symptoms are caused by the growth of the tumor in to the surrounding tissue. Intra abdominal desmoids may result in an intestinal obstruction. Extremity desmoids can restrict the movement of affected joints and cause limping or difficulty in arm or leg movements.*

The present case is an young woman who had undergone two caesareans earlier and hysterectomy 4 years back, presented with pain and a mass in lower abdomen. It was diagnosed to be a desmoids tumor arising from right rectus abdominis and was excised successfully, with a meshplasty of the gap created.

Keywords: *Desmoid tumor, aggressive fibromatosis, familial adenomatous polyposis*

I. Case Report

A young female of 34years, reported to us with history of pain lower abdomen since 2months. Burning micturition since last one month. It was a continuous dull aching pain, aggravated with heavy work. Patient had no obstructive or voiding symptoms. She had 2 children, both born by caesareans, last caesarean done 10 years back. She underwent hysterectomy 4 years back for dysfunctional uterine bleeding.

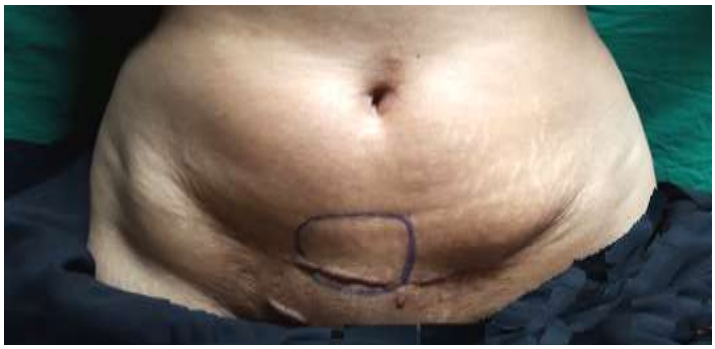

Fig. 1

On examination of abdomen, a pfannenstiel scar of hysterectomy with keloid tendency was found. A firm tender parietal mass 7x5 cm with nodular surface was palpable in supra pubic area. Its upper margin was well defined and lower margin continued behind pubic symphysis and had restricted mobility. Urine examination revealed no signs of urinary tract infection and renal functions tests were within normal limits.

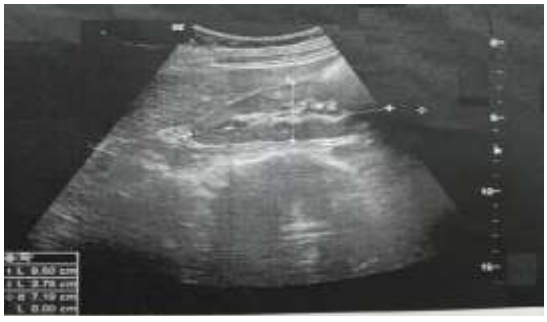

Fig. 2

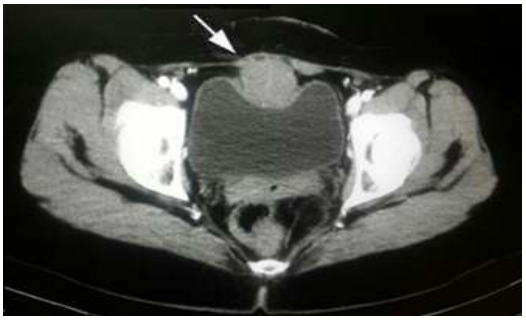

Fig.3

On USG (Fig. 2) a well defined hyper echoic mass 6x4x2.5 cm noted deep to subcutaneous planes in anterior abdominal wall in suprapubic region abutting anterior wall of urinary bladder. No signs of internal vascularity or calcification.

CECT showed evidence of a relatively well defined anterior abdominal wall mass lesion (5.7 x 4.0 x 2.8) cm with nodular outline. It appeared to be arising from right rectus abdominis muscle just above the pubic symphysis, showing inward bulging, causing indentation over the anterior wall of urinary bladder.(Fig. 3)

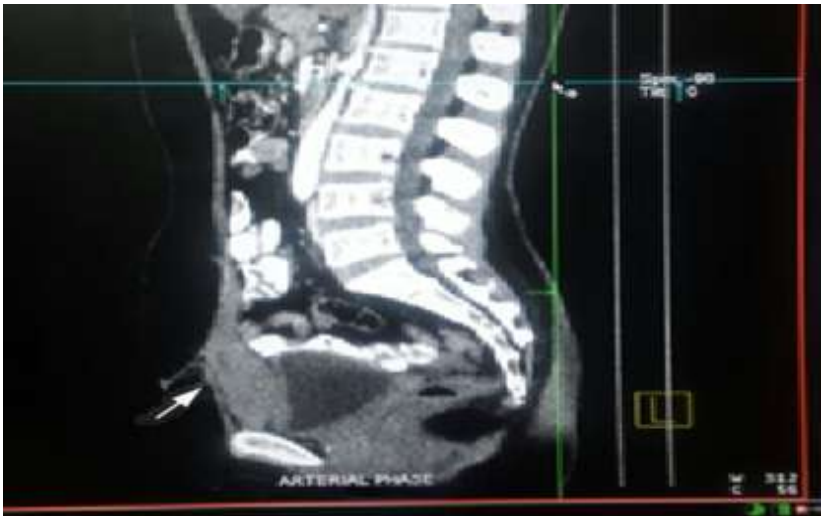

Fig. 4

On post contrast study, the lesion showed mild heterogenous enhancement of contrast.(Fig. 4) Features were in favour of intramuscular mass lesion in the anterior abdominal wall just above the pubic symphysis in the right rectus abdominis muscle at the level of previous incisional site, possibly a desmoid tumor.

A cystoscopy revealed indenation of the tumor over the anterior wall of the bladder, inferiorly few centimetres short of the bladder neck. Bladder mucosa appeared to be normal with no signs of tumor infiltration of entire thickness of bladder wall.

Fig. 5

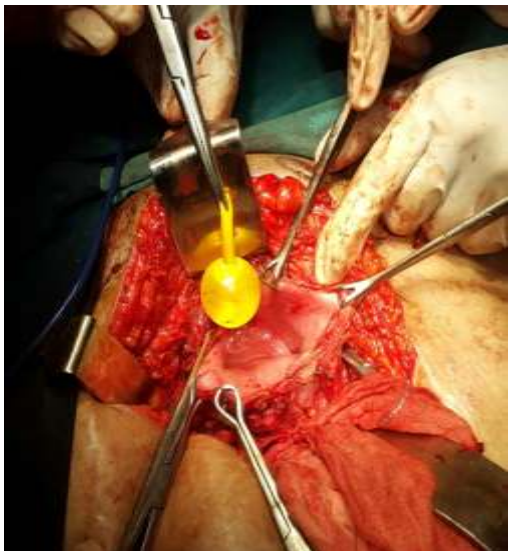

Fig. 6

The tumor was explored by an extra peritoneal approach and found to have infiltrated rectus sheath, lower portion of right rectus abdominis muscle and had crossed the midline to involve part of left rectus.(Fig. 5) It extended behind the pubic bones and badly stuck to the periosteum. It infiltrated in to detrusor in the anterior wall of bladder without involving bladder mucosa. The tumor was excised with safe margin along with a portion of involved bladder wall. (Fig. 6)

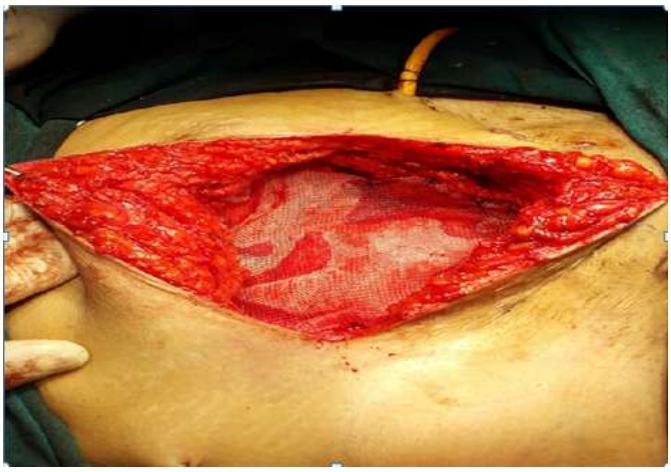

Fig. 7

Bladder was repaired in two layers with a supra pubic and a urethral catheter in situ. The wide defect created in the abdominal wall was bridged satisfactorily by poly propylene mesh reconstruction. (Fig. 7) The excised tumour specimen measured 9 x 4.5 x 2.5 cm. Cut section looked grey white and homogenous. (Fig. 8a,b)

Fig.8 a

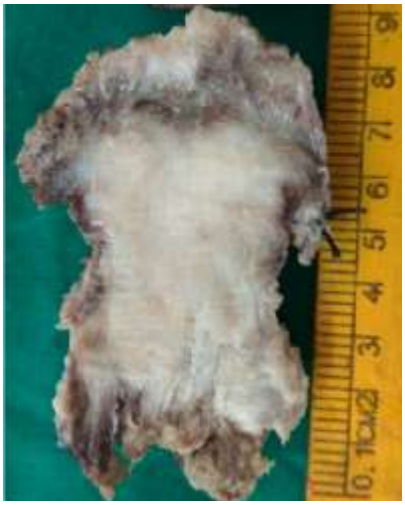

Fig.8 b

Histopathological features

The tumour tissue was composed of elongated spindle cells containing small pale staining nuclei with 1-3 nucleoli. These cells are separated by abundant collagen fibres. Cells and collagen fibres were arranged in sweeping bundles. There were foci showing normal striated muscle fibres along with areas of haemorrhage and lymphoid aggregates.

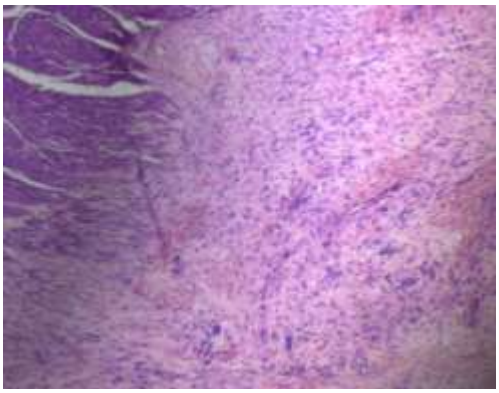

Fig. 9a

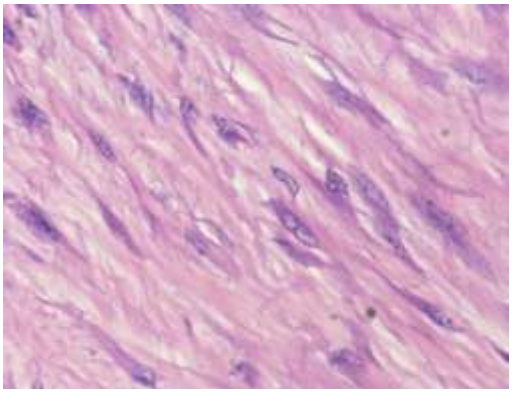

Fig. 9b

II. Discussion

Desmoid tumors account for 0.03% of all neoplasms and 3% of all soft tissue tumors.[1] The frequency of occurrence in the general population is 2.4 to 4.3 cases/ million and the risk is enhanced by 1000-fold in patients with familial adenomatous polyposis, an autosomal dominant syndrome.[2] In patients with familial adenomatous polyposis, 10-25% develop desmoid tumors. [3,4,5] Patients with desmoid tumors are evaluated for associated polyposis syndrome by taking a detailed family history and performing a lower GI endoscopy.[6] The peak of occurrence is between the age of 25 and 40 years.[7]

Most of them are sporadic and about 80% of cases seen in young parous women and often occurs in abdominal operation scars, stretching of the muscle fibres during pregnancy, a small haematoma or trauma to the abdominal wall.[8,9]They arise from the rectus abdominis muscle and infiltrate into the adjacent muscle bundle, often entrap them and result in myxomatous degeneration. Now it grows rapidly but metastasis does not occur.[9] These tumors are associated with a high recurrence rates, even if their microscopic characters indicate a benign disease. [10]

They are classified as per their location, as extra abdominal or extremity desmoids, abdominal wall desmoids, intra-abdominal desmoids involving the mesentery, pelvis, or bowel wall.[2,10] Abdominal wall desmoids originate from musculoaponeurotic structures of the abdominal wall, especially the rectus and internal oblique muscles and their fascial coverings below the umbilicus and often cross the midline. [9,11] Intra-abdominal desmoids are situated within the mesentery of the small bowel. Death occurs as a result of progressive cachexia and complications of small bowel obstruction [5] In CT, the lesions are solitary in 72% and multiple in 28%. Fifty percent are located in the abdominal wall, 41% in the mesentery, and 9% in the retroperitoneum [12]

A modest surgical excision with clear margins is the main determinant of outcome with the risk of local recurrence. Completeness of resection is an important prognostic factor.[3, 13] For high recurrence rate more than one surgery may be needed. A mutilating surgery is avoided and adjuvant radiotherapy is advocated to reduce the recurrence rate, in cases of involved surgical margins. [4,14] For pain and swelling anti inflammatory medications are given. Non-surgical treatment includes irradiation, brachytherapy, endocrine therapy and cytotoxic chemotherapy but outcome is diverse and unpredictable .[15] Radiation alone in form of EBRT is acceptable for unresectable tumors.[3]

Recurrence after surgery was high in the FAP-associated tumor group.[16] Recurrence rates of 38% for surgery alone, 25% for combined surgery and radiation, and 24% for radiation therapy alone. 27% recur even if initial resection is complete with negative margins. Adjuvant radiation therapy tried for patients with positive margins.[2, 11] Surgery should be minimized as much as feasible, while at the same time achieving free margins. Adjuvant therapy should be considered in selected cases only.[10] Abdominal wall desmoids respond to radiotherapy, although the effect is slow. So radiotherapy is a treatment option for unresectable tumors. [2,17] Also various cytotoxic regimens like methotrexate with vinblastine, doxorubicin-based therapy, and ifosfamide-based regimens show positive responses in 20% to 40% cases. Imatinib, a tyrosine kinase inhibitor is another effective treatment option. 2. Percutaneous chemical ablation with acetic acid, radiofrequency ablation, pirfenidone, interferon alpha and glivec (imatinib, 800 mg/d) are still under trail. Gene transfer therapy is currently under intensive research. [10,17]

III. Conclusion

Desmoid tumours have an unpredictable natural history and are challenging.[18] Even if a rare entity, the combination of certain features like history of previous surgery, the age, sex, parity of the patient, the location of the tumor within the anterior abdominal wall and imaging findings lead to a strong possibility to consider it as a desmoids tumor. [19] Biopsy establishes the diagnosis. Immunohistochemistry shows the tumors typically stain positive for β-catenin, actin, and vimentin and stain negative for cytokeratin and S-100. 2.

Estrogen receptors if detected in the tumor, provides support to use antiestrogens. Clinical improvement noticed in 43% of patients receiving tamoxifen but the long-term benefit is minimal. [2,20] In recurrent desmoid tumours with ER positive status, a hormone therapy with tamoxifen or toremifene, and sulindac (NSAID) have a response rates of up to 50%.[15] In spite of controversies, the definitive treatment of abdominal wall desmoids is wide local excision followed by mesh repair. [2,4,7,17,20] Radiotherapy is avoided in cases with negative tumor margins to avert the radiation-related morbidity and sanction a better quality of life.[4]

References

[1]. Economou et al.: Desmoid tumor of the abdominalwall: a case report. Journal of Medical Case Reports 2011 5:326.
[2]. Sabiston Text book of Surgery 19th ed. 2009,ABDOMINAL WALL, UMBILICUS, PERITONEUM, MESENTERIES, OMENTUM. Chapter 45 1095 – 1096
[3]. Chalapathy D. V. et al. Rare case of desmoid tumor of urinary bladder , J of Evidence Based Med & Hlthcare, Vol. 2/Issue 33/Aug. 17, 2015 Page 5078
[4]. Bernd Kasper et al. Desmoid Tumors: Clinical Features and Treatment Options for Advanced Disease. The Oncologist. 2011 May; 16(5): 682–693.
[5]. Jones IT, Jagelman DG, Fazio V et al. Desmoid tumors in familial polyposis coli. Ann Surg 1986; 204:94–97.
[6]. Debi Prasad Mishra Et al. Desmoid tumors: A clear perspective or a persisting enigma? A case report and Review of literature. World journal of Ocology, ISSN 1920-4531 print, 1920-454 x
[7]. Quinn SF, Erickson SJ, Dee PM et al. MR imaging in fibromatosis: results in 26 patients with pathologic correlation. AJR Am J Roentgenol 1991; 156:539- 542.
[8]. Vandevenne JE, De Schepper AM, De Beuckeleer L, et al. Newconcepts in understanding evolution of desmoid tumors: MR imagingof 30 lesions. Eur Radiol 1997; 7:1013–1019
[9]. Bailey & Love's SHORT PRACTICE OF SURGERY 25th edition published in 2008, HERNIAS , UMBI L ICUS AND ABDOMINAL WALL pg 990
[10]. Sakorafas GH et al. Abdominal desmoid tumors. Surg Oncol. 2007 Aug;16(2):131-42.
[11]. Casillas J, Sais GJ, Greve JL, Iparraguirre MC, Morillo G: Imaging of intraand extra abdominal desmoid tumors. Radiographics 1991, 11:959-968.
[12]. Einstein DM et al. Abdominal desmoids: CT findings in 25 patients. AJR Am J Roentgenol. 1991;157:275–279
[13]. M. Overhaus, P. Decker, H. P. Fischer, H. J. Textor, and A. Hirner, "Desmoid tumors of the abdominal wall: a case report," World Journal of Surgical Oncology, vol. 1, article 11, 2003.
[14]. Nuyttens et al. Surgery vs. Radiation for Desmoid Tumors.CANCER April 1, 2000 / Volume 88 / Number 7. 1517-1522
[15]. Hardell L, Breivald M, Hennerdal S, Fernberg JO, Strander H. Shrinkage of desmoid tumor with interferon alfa treatment: a case report. Cytokines Cell Mol Ther. 2000;6(3):155-156.
[16]. Hansmann A, Adolph C, Vogel T, Unger A, Moeslein G. High-dose tamoxifen and sulindac as first-line treatment for desmoid tumors. Cancer. 2004;100(3):612-620
[17]. Joseph K Wanjeri et al. A massive abdominal wall desmoids tumor occurring in laparotomy scar: A case report. World J Surg Oncol.2011; 9 :35.
[18]. Shields CJ et al. Abdominal desmoid tumors Eur J Surg Oncol. 2001 Dec;27(8):701-6.
[19]. Harvey **E. L. Et al.** Case 84: Desmoid Tumor of the Abdominal Wall 10.1148/ Radiology 2005**;** 236:81–84
[20]. Sutton RJ, Thomas JM. Desmoid tumours of the anterior abdominal wall. Eur J Surg Oncol. 1999;25(4):398-400.