RARE CASE OF DESMOID TUMOR OF URINARY BLADDER
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ABSTRACT: Desmoid tumor is a benign soft tissue tumor which belongs to a family of myofibroblastic fibromatoses. Occasionally, desmoid tumors have an unusual site of occurrence. We describe a case of incisional hernia in postmenopausal women with an intra operative incidental finding of a desmoid tumor from anterior wall of urinary bladder for which a wide excision was performed.

KEYWORDS: Fibromatosis, Urinary bladder, Wide excision.

INTRODUCTION: Desmoid tumor is also known as aggressive fibromatosis arising from fibroaponeurotic tissue and typically presents as slowly growing mass.¹ According to the World Health Organization, Desmoid tumor is defined as “clonal fibroblastic proliferations that arise in the deep soft tissues and are characterized by infiltrative growth and a tendency toward local recurrence but an inability to metastasize.”² Incidence of desmoid tumor in general is 2 to 4 individuals per million persons, however in literature only a few case reports are published on desmoid tumor of urinary bladder. Usually desmoid tumor is seen in pregnant patients who underwent cesarean section. Desmoid tumor usually presents with mass near cesarean section scar. Desmoid tumors usually occur in fertile females and are uncommon after the menopause. During pregnancy an increase in volume occasionally occurs in already existing tumors. This corroborates the estrogen-stimulated tumor growth hypothesis.³ Histologically these tumors consists of spindle-shaped cells in a collagenous matrix without the pleomorphic, atypical or hyperchromatic nuclei of malignancy.⁴ Desmoid tumors have been subdivided according to their location into extra-abdominal, abdominal and intra-abdominal, and the latter have been sub classified further into mesenteric fibromatosis and pelvic fibromatosis⁵. The connective tissue hyperplasia infiltrates locally, recurs frequently after resection but does not metastasize⁶. The correct pre and intra operative diagnosis of this rare disease is the most important factor for the respective treatment and prognosis of the destructive desmoid fibromatosis.

CASE: A 54 year old post-menopausal lady presented with history of midline swelling below the umbilicus of 4 months duration, which increases in size on coughing, straining and reduces on lying down. She had no history of pain abdomen, mass per abdomen or urinary complaints. She is a known case of Carcinoma of Cervix and underwent abdominal hysterectomy with pelvic lymph node dissection 6 months back. She had a history of pulmonary tuberculosis and received a full course of antitubercular treatment 6 years back.

Abdominal examination revealed a healed pfannelstein incision scar over lower abdomen with features of uncomplicated Incisional hernia through a defect of 4x5cms, without any lower
abdominal mass. All routine investigations were within normal limits. An ultrasound study of abdomen revealed Incisional hernia with a defect in anterior abdominal wall of 7cm with no features suggestive of lower abdominal mass or metastatic deposits.

Patient was planned for elective laparotomy and mesh repair. On exploration we found a solitary firm to hard mass of 6x6cms from anterior wall of urinary bladder abutting the lower anterior abdominal wall. [Fig. 1] Patient underwent wide excision of mass with primary repair of bladder wall in two layers along with anatomical repair of Incisional hernia. [Fig. 2] Excised specimen showed firm to hard solid mass, with homogeneous grey white to brown cut surface. [Fig. 3] Microscopic examination showed bland spindle shaped fibroblastic cells arranged in ill-defined fascicles and confirmed the diagnosis of desmoid tumor. [Fig. 4] Post-operative period was uneventful. Urinary catheter was removed after 2weeks. Patient is on regular follow up.

**DISCUSSION:** Desmoid tumors are monoclonal fibroblastic proliferations arising in musculoaponeurotic structures. They are benign but aggressive tumors of mesenchymal origin, forming a heterogenous group of pathologic entities resulting from the proliferation of well-differentiated fibroblasts. On microscopy desmoid tumors are poorly circumscribed, infiltrate the surrounding tissue, lack a true capsule and are composed of abundant collagen surrounding poorly circumscribed bundles of elongated, slender, spindle-shaped cells of uniform appearance.
Desmoid tumor is an uncommon neoplasm that can occur sporadically or with Familial adenomatosis polyposis (FAP) and Gardner’s syndrome. The risk increases 1000 folds in FAP. Inheritance (Or new mutation) of one copy of APC tumor suppressor gene is the cause of FAP and the two commonest causes of deaths in these patients are duodenal cancer and desmoid tumors.\(^\text{10}\) In FAP associated cases, desmoid tumors represent an extra-colonic manifestation of polyposis syndrome.\(^\text{11}\) Every patient with desmoid tumor should therefore be evaluated for the presence of associated polyposis syndrome by taking a detailed family history, performing colonoscopy and possibly upper gastrointestinal endoscopy.\(^\text{12}\) The vast majority of desmoid tumors are sporadic, typically in young woman during pregnancy or within a year of child birth.\(^\text{2}\) 25% of cases occur in abdominal wall.\(^\text{1}\)

These very rare tumors can develop in any musculoaponeurotic structure, they can be found in all regions of human body. The clinical behaviour and prognosis of desmoid is very diverse and depends on the anatomic location and proximity to vitally important organs. A corelation with the familial intestinal polyposis could be shown.\(^\text{13}\) Approximately 10-25% of patients with polyposis presents with intra or extra abdominal desmoid tumors.\(^\text{14}\) Supposed risk factors of desmoid are previous surgical interventions, pregnancy and hormonal treatment with estrogen. In the presented case previous surgical intervention is one possible risk factor for the disease.

The differential diagnoses for rectus abdominis lesions include hematoma, fibrosarcoma, lymphoma, rhabdomyosarcoma, liposarcoma, leiomyosarcoma, neurofibroma, benign fibrous tumor and primitive neuroectodermal tumor.\(^\text{15}\)

On ultrasonography, desmoid tumors appear as well-defined lesions with variable echogenicity. The lateral borders may appear ill defined or irregular.\(^\text{15,16}\) The CT appearance of desmoid tumors depends on their composition. They may appear homogeneous or heterogeneous and hypo, iso, or hyper intense compared with the attenuation of muscles. The degree of enhancement after the intravenous administration of contrast medium is variable.\(^\text{15,16,17,18}\) Magnetic resonance imaging (MRI) features of desmoid tumors also show wide variability depending on the stage they are imaged. Characteristic MRI findings include poor margination, low signal intensity on T1-weighted images and heterogeneity on T2-weighted images, and variable contrast enhancement. Low T2 signal intensity bands are characteristic and represent foci of high concentrations of collagen deposition.\(^\text{15,16,17}\)

Surgery remains the treatment of choice for desmoid tumor.\(^\text{1}\) Resection of the tumor with a wide margin of normal tissue is currently considered the optimal treatment. Abdominal wall desmoids are responsive to radiation therapy. Radiation alone is an acceptable treatment for unresectable Desmoid tumor\(^\text{2}\). Completeness of resection is an important prognostic factor

**CONCLUSION:** Desmoid tumors are the rare benign tumors with high local recurrence potential, arising most commonly from cesarean scar. The intra perotonial desmoid are very rare with very few case reports of urinary bladder desmoids. In our case, patient is a post-menopausal lady, without symptoms and signs of mass abdomen and urinary complaints. The desmoid tumor was from the urinary bladder which is very rare site for desmoid tumor occurrence. This makes our case as a rare presentation of desmoid tumor. As the patients with desmoid tumor can be
asymptomatic, desmoid tumor should be kept in mind even though rare, in cases with previous lower abdominal surgeries and treated with wide local excision.

REFERENCES:

1. Raphael E Pollock, atlas of clinical oncology, soft tissue sarcomas, 114-120
2. Biermann JS. Desmoid tumors. Curr Treat Options Oncol. 2000; 1: 262–266.
3. Enzinger FM, Weiss SW. Soft tissue tumors. 4th ed. St. Louis: CV Mosby; 2001. pp. 1472–1475.
4. Devita, hellman and rosenberg’s, Principles and practice of oncology, soft tissue sarcomas, 9th, 1539-1540.
5. Weiss S, Goldblum JR, editors. Enzinger and Weiss's Soft Tissue tumors. 4th ed. St Louis: Mobis; 2001. pp. 641–693.
6. Lewis JJ, Boland PJ, Leung DH, Woodruff JM, Brennan MF. The enigma of desmoid tumors. Ann Surg. 1999; 229: 866–872; discussion 872-873.
7. Moslein G, Dozois RR. Desmoid tumors associated with familial adenomatous polyposis. Perspectives in Colon and Rectal Surgery. 1998; 10: 109–126.
8. Sagar PM, Moslein G, Dozois RR. Management of desmoid tumors in patients after ileal-pouch-anal anastomosis for familial adenomatous polyposis. Diseases of the colon and rectum. 1998; 41: 1350–1355. doi: 10.1007/BF02237046.
9. Hartley JE, Church JM, Gupta S, McGannon E, Fazio VW. Significance of incidental desmoids identified during surgery for familial adenomatous polyposis. Diseases of the Colon and Rectum. 2004; 47: 334–340. doi: 10.1007/s10350-003-0063-0.
10. Latchford AR, Sturt NJH, Neale K, Rogers PA, Phillips RKS. A 10-year review of surgery for desmoid disease associated with familial adenomatous polyposis. British Journal of Surgery. 2006; 93: 1258–1264. doi: 10.1002/bjs.5425.
11. Julian N, Sturt H, Clark SK. Current ideas in desmoid tumours. Familial Cancer. 2006; 5: 275–285. doi: 10.1007/s10689-005-5675-1.
12. Rohrich RJ, Lowe JB, Hackney FL, Bowman JL, Hobar PC. An algorithm for abdominal wall reconstruction. Plastic and Reconstructive Surgery. 2000; 105: 202–216. doi: 10.1097/00006534-200001000-00036.
13. Mulik V Griffith AN, Beattie RB. Desmoid tumors with familial adenomatous polyposis in pregnancy. Journal of obstetrics and gynaecology. 2003; 23(3)307-308.
14. Sinha A,Gibbons DC.surgical prophylaxis in familial adenomatous polyposis:do pre-existing desmoids outside the abdominal cavity matter? Familial cancer.2010; 9(3): 407-411.
15. Teo HEL, Peh WCG, Shek TWH. Case 84: desmoid tumor of the abdominal wall. Radiology. 2005; 236: 81–84. doi: 10.1148/radol.2361031038.
16. Casillas J, Sais GJ, Greve JL, Iparraguirre MC, Morillo G. Imaging of intra- and extra abdominal desmoid tumors. Radiographics. 1991; 11: 959–968.
17. Overhaus M, Decker P, Fischer HP, Textor HJ, Hirner A. Desmoid tumors of the abdominal wall: a case report. World J Surg Oncol. 2003; 1: 11. doi: 10.1186/1477-7819-1-11.
18. Einstein DM, Tagliabue JR, Desai RK. Abdominal desmoids: CT findings in 25 patients. AJR Am J Roentgenol. 1991; 157: 275–279.
CASE REPORT

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