Case Report

Urology, Nephrology and Andrology International
Volume 5 Number 2, July–December 2020
DOI: http://dx.doi.org/10.21088/unai.2456-5016.5220.9

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A Rare Case of Desmoid Tumour Infiltrating into Bladder: Case Report

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How to cite this article:
Ravi Kumar BR, Prakash Prabhu, Abhilash Gautham Ramesh / A Rare Case of Desmoid Tumour Infiltrating into Bladder: Case Report. Urology, Nephrology and Andrology International. 2020;5(2):57–59.

Abstract

Introduction: Desmoid tumor also known as aggressive fibromatosis, is a rare, locally invasive, non-metastasizing soft tissue tumor. They arise from any part of the body in different types of connective tissues. Infiltration of the bladder is usually rare and very few cases have been reported till date.

Case report : 35/F presented with mass in the suprapubic region and lower abdominal pain since 1 month. Total abdominal hysterectomy + 8 years with pfananstiel incision.

Per abdomen- soft, mass about 5 × 6 cm was palpable over the lower abdomen in the suprapubic region with tenderness. Fnac showed features suggestive endometriosis of bladder wall. Cect revealed? Ca bladder/ endometriosis of bladder. In view of high clinical suspicion (no h/o cyclical bleeding and cyclical pain). Cystoscopy was done which showed mucosal edema in right lateral wall with no mass in bladder with normal cystoscopy findings. Diagnostic laparoscopy and biopsy from the mass and right lateral bladder wall was performed. Histopathological diagnosis was features are those of collagen forming fibrous lesion suggestive of fibromatosis. Hence surgical excision was planned. Complete excision of the mass was done. Final histopathology report was features suggestive of fibromatosis with no evidence of endometriosis/epithelial malignancy. Following surgery patient improved and was asymptomatic.

Discussion: Desmoid is a rare tumor, with reported incidence of 2–4 per million population and accounts for 0.03% Of all neoplasms. Desmoid tumor arises from myofibroblast, lacks a true capsule, usually infiltrates into adjacent muscle bundles. But the infiltration of the bladder is rare.

The etiopathogenesis is multifocal. Mutation in apc or beta catenin genes is seen. Complete excision of the mass with sufficient margins of anterior rectus sheath with deroofing of the tumor with bladder wall was done. Final histopathology report was features suggestive of fibromatosis with no evidence of endometriosis/epithelial malignancy.

Conclusion: The course of desmoid tumor is unpredictable, there can be spontaneous regression, long-lasting mass and/or progression of disease can occur.[5] Currently preferred management for asymptomatic cases may be "wait and see" approach. Tumors showing increase in size and/ or causing symptoms require active management.

Keywords: Desmoid; Bladder; Excision.

Introduction

Desmoid tumor also known as aggressive fibromatosis, is a rare, locally invasive, non-metastasizing soft tissue tumor.

They arise from any part of the body in different types of connective tissues including muscle, fascia and aponeurosis.[1]

The most common primary sites are the abdominal wall, limbs, girdles, and mesenteric area.

Infiltration of the bladder is usually rare and very few cases have been reported till date.

Case Report

A 35-year-old female presented with a mass in the suprapubic region and lower abdominal pain since 1 month. She underwent abdominal Hysterectomy 8 years ago with pfananstiel incision.

Her Obstetric history is P2L2, both FTND and last child birth 20 years back.

On examination, Per abdomen was soft,mass about 5×6 cm was palpable over the lower abdomen in the suprapubic region with tenderness. (Fig. 1)

USG abdomen showed soft tissue mass of 5×3 cm involving the right lateral and dome of bladder, FNAC from the mass showed scattered small cells with compact nuclei resembling stromal cells and hemosiderin pigment laden macrophages with features suggestive endometriosis of bladder wall

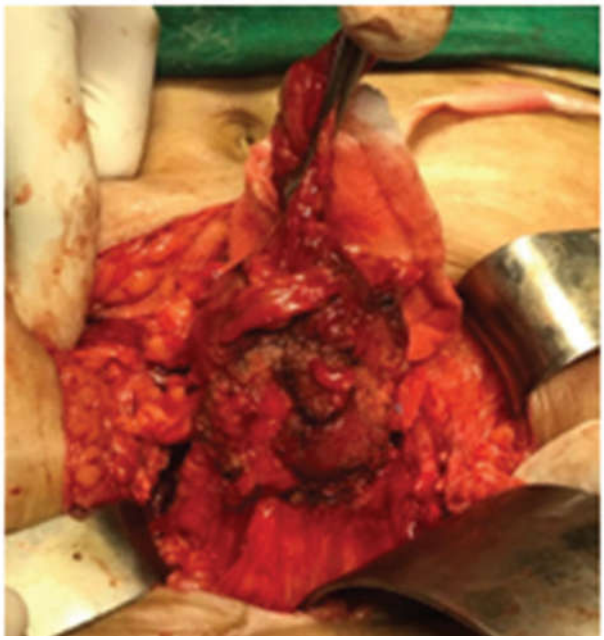

Fig. 1: Intra operative picture indicating tumour with bladder invasion.

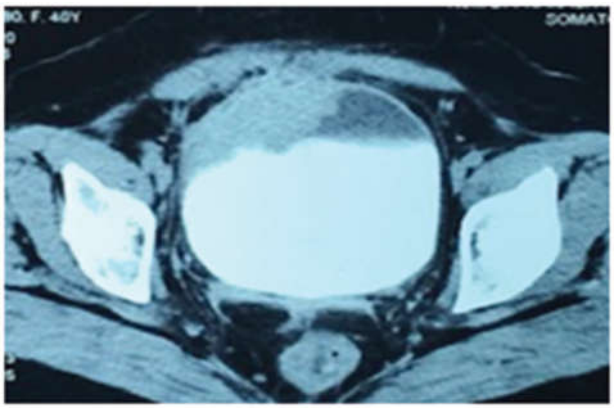

Fig. 2: CT - Scan inage with bladder wall infiltration of tumour.

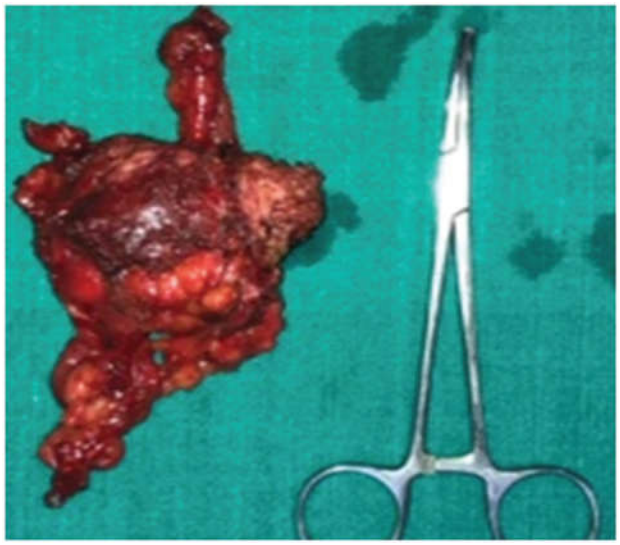

Fig. 3: Post operative image with resected specimen.

Cect Abdomen and Pelvis revealed heterogeneously enhancing soft tissue density mass lesion arising from the bladder with extravesical extension - ca bladder/endometriosis of bladder (Fig. 2)

Patient did not respond to LHRH agonist which was given for 6 weeks and continued to have pain.

In view of high clinical suspicion (no h/o cyclical bleeding and cyclical pain)

Cystoscopy was done which showed mucosal edema in right lateral wall with no mass in bladder with normal cystoscopy findings.

Diagnostic laparoscopy and biopsy from the mass and right lateral bladder wall was performed. Histopathological diagnosis was features are those of collagen forming fibrous lesion suggestive of fibromatosis. (Fig. 3)

Hence surgical excision was planned. Mid line laparotomy inscion was placed. Mass was involving the anterior rectus with extension upto right lateral wall of bladder without mucosal involvement. Complete excision of the mass with sufficient margins of anterior rectus sheath with deroofing of the tumor with bladder wall was done. Final Histopathology report was features suggestive of fibromatosis with no evidence of endometriosis/ epithelial malignancy.

Following surgery patient improved and was asymptomatic.

Discussion

Desmoid is a rare tumor, with reported incidence

of 2–4 per million population and accounts for 0.03% of all neoplasms.[2] Desmoid tumor arises from myofibroblast, lacks a true capsule, usually infiltrates into adjacent muscle bundles. But the infiltration of the bladder is rare.

The common tumors of bladder are transitional cell carcinoma, epithelial metaplasia, inverted papilloma, leukoplakia.

Desmoid tumors are typically diagnosed in young adults (peak incidence 35–40 years) mainly in women at reproductive age. The etiopathogenesis is multifocal.[3] It may be sporadic or familial in nature. Mutation in APC or beta catenin genes is seen.[4] Surgical trauma accounts for 68-86% of all desmoids. Pregnancy and use of oral contraceptives have also implicated in the etiopathogenesis. Multimodality imaging including USG, CT and magnetic resonance play a key role in the diagnosis, staging & follow up.

Conclusion

The course of desmoid tumor is unpredictable, there can be spontaneous regression, long-lasting mass and/or progression of disease can occur.[5] Currently preferred management for asymptomatic cases may be "wait and see" approach. Tumors showing increase in size and/ or causing symptoms require active management.

In this case patient was symptomatic and hence surgical excision was performed.

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