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Submucosal cystic neurofibroma of the bladder- A case report

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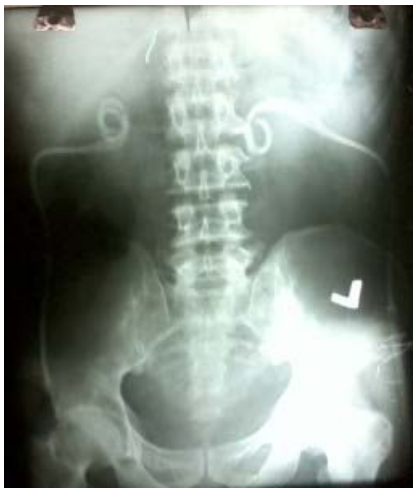
Abstract :
50 year old male, farmer by occupation presented with dysuria and urinary incontinence 3months duration, hematuria 2 episodes, only in the initial stream, painless, no passage of clots or tissue bits. Abdominal examination was normal with no organs or mass palpable. Digital rectal examination revealed grade 1 prostatic enlargement and above prostate,there was large boggy mass , superior extent could not be felt . Routine biochemical and hematological investigation revealed raised renal parameters. Urine routine examination was normal. Urine culture revealed - no growth. Urine cytology - negative for malignancy. Chest x ray is normal. USG study, showed multiloculated mixed echogenic mass appearing within bladder. Both kidneys showed hydroureteronephrosis. Suprapubically usg guided trucut biopsies from the mass done twice- were negative for malignancy. Plain CT abdomen and pelvis done previously showed heterodense lesions inside the bladder both anterior and posteriorly, with peri vesical extension posteriorly. Antegrade study showed multiple filling defects inside the bladder. MRI pelvis showed hetero intense mass lesion, soft tissue component predominantly in posterior aspect, and multiple cystic components filling rest of the bladder, both ureteric orifices encroached by the mass. Initially right percutaneous nephrostomy (pcn) was done, left pcn done subsequently because of persistently elevated renal parameters. Cystoscopy revealed normal bladder mucosa , with indendations seen - (right) lateral wall, (right) anterior wall, and in the posterior wall midline above trigone, (right) ureteric orifice could not be visualised. Trans urethral resection biopsy - possibility of benign leiomyoma. Surgery through lower abdominal, extraperitoneal approach. Bladder was opened. Tumour found to be intramucosal, pedunculated arising from the base of the bladder, mostly from the right side, involving right ureteric orifice also. Entire tumour was resected and right ureteric reimplantation done . Post operative histopathology - sub mucosal cystic neurofibroma. The case is presented for its rarity.

Keyword :neurofibroma bladder, ureteric reimplantation, benign bladder tumor

INTRODUCTION -

Neurofibroma is a rare, benign, neoplastic tumor of the nerve sheath, composed of Schwann cells, perineurium-like cells, fibroblasts, and intermediate-type cells.[1] Most occur in the setting of neurofibromatosis type 1, rare cases of isolated sporadic bladder neurofibroma have also been reported . Von Recklinghausen's neurofibromatosis - an autosomal dominant disorder characterized by multiple subcutaneous neurofibromas and cutaneous pigmentation. >50 cases of genitourinary neurofibroma have been reported and the bladder is the most common genitourinary organ to be involved.[2] Histologic findings are the same regardless of the site of occurrence. Differential diagnosis: other spindle cell tumors such as Leiomyoma , Inflammatory myofibroblastic tumor; Postoperative spindle cell nodule, low-grade leiomyosarcoma, malignant peripheral nerve sheath tumor (MPNST), and Rhabdomyosarcoma. Continued surveillance for malignant transformation is required.



CASE HISTORY -

50 years old male presented with dysuria – 3months ,urinary incontinence – 3months, hematuria – 2 episodes, only in the initial stream, painless, no passage of clots or tissue bits. No comorbid illness.No previous surgical illness.Treated elsewhere, as large multi cystic mass in the bladder with bilateral hydroureteronephrosis , increased renal parameters (urea-120 mgs ,cr-4.8mgs). patient underwent bilateral percutaneous nephrostomy (PCN). USG

Bilateral PCN:

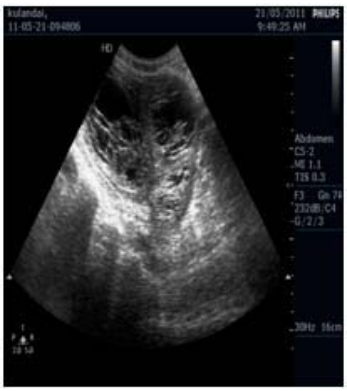
guided biopsies from the mass done twice – negative for malignancy ***.****initially treated elsewhere and referred to our institute for further management later. Abdominal examination was normal with no mass or organs palpable. Digital rectal examination revealed Grade I prostate, and a large boggy mass above the prostate, superior extent could not be felt.*

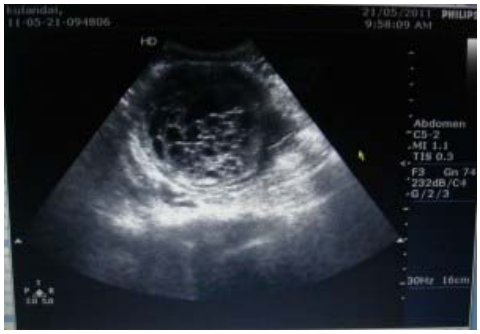
Evaluation

-*Hemogram – normal , Urine culture and sensitivity -no growth, Urine cytology -negative for malignant cells , Urea - 58 mg%, Creatinine - 1.6 mg% , (PCN INSITU), Sugar - 82 mg%.*

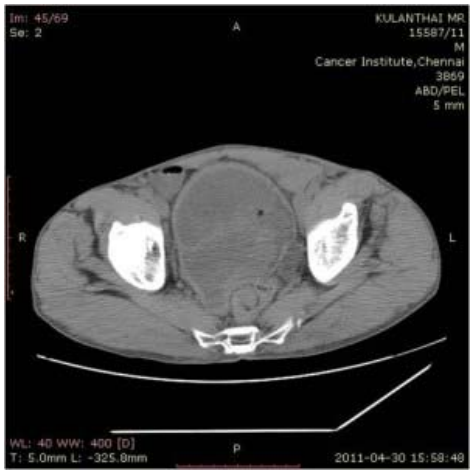
USG study

-*multiloculated mixed echogenic mass appearing within bladder, Both kidneys were normal, bilateral pcn insitu.*



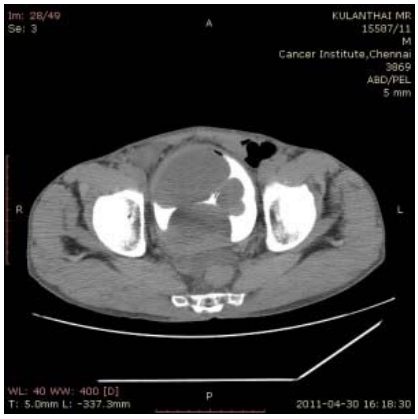


USG

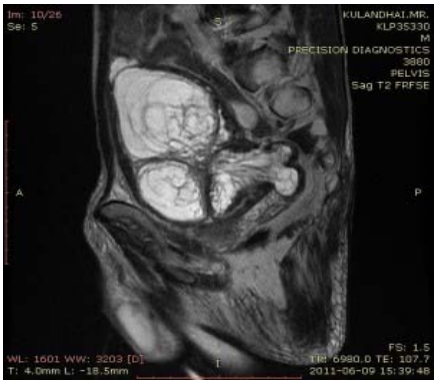


Plain CT abdomen & pelvis –
showed heterodense lesions inside the bladder both anterior andposteriorly, with peri vesical extension posteriorly.
Antegrade study -showed multiple filling defects inside the bladder.

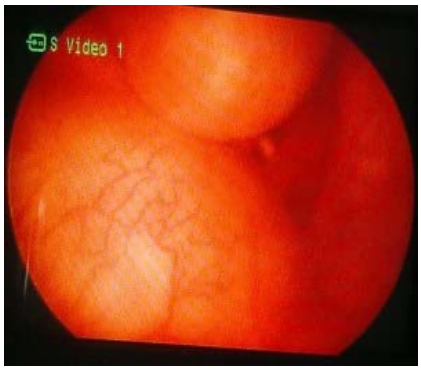
CT picture Antegrade study:



MRI pelvis – showed hetero intense mass lesion, soft tissue component predominantly in posterior aspect, and multiple cystic components filling rest of the bladder, both ureteric orifices encroached by the mass.



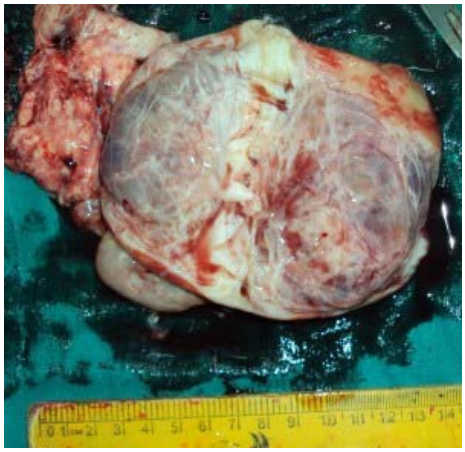
MRI



Cystoscopy -urethra - normal , veru - normal , prostate - grade 1 lateral lobes, bladder mucosa normal , indendations seen -right lateral wall, right anterior wall , posterior wall midline above trigone, right ureteric orifice could not visualized and left ureteric orifice was normal.
Cystoscopy picture
Transurethral resection biopsy - Extensive proliferation of interlacing bundles of smooth muscle spindle cells in the subepithelial tissue, with

patchy areas of lymphocytic infiltrate in the sub mucosa, sub mucosal smooth muscle hyperplasia with chronic cystitis - Possibility of Benign Leiomyoma

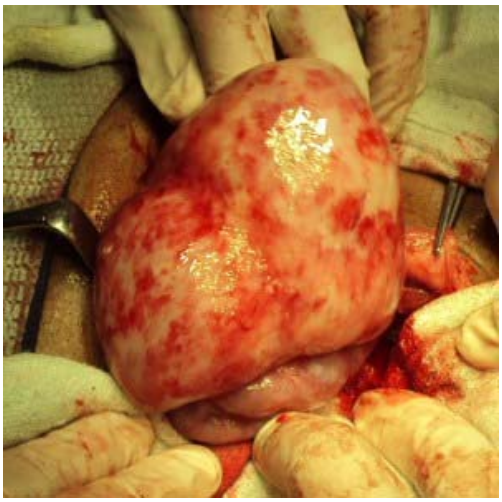
On exploration -Surgery through lower abdominal, extraperitoneal approach. Bladder opened , multilobulated submucosal mass of size 11 X 9cm, arising from base of the bladder mostly from the right side involving right ureteric orifice, intravesical excision of mass with right ureteric reimplantation done. Cut section showed edematous cystic degeneration



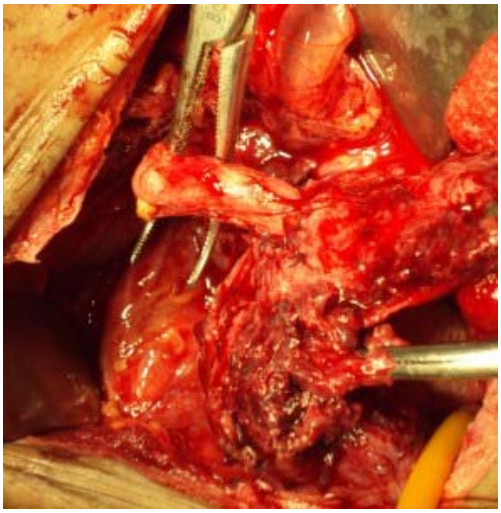
Ureteric reimplantation picture
Resected specimen

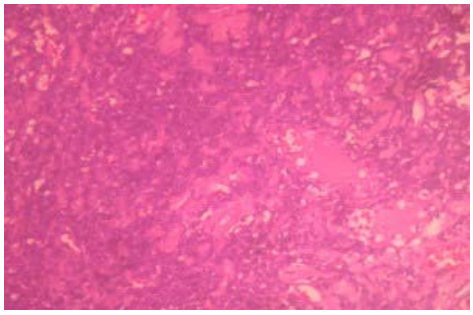
Postoperative Histopathology: Submucosa showing multiloculated cystic tumour mass composed of interlacing bundles of short spindle cells with wavy small nuclei, Stroma shows hyaline/ collagen deposits. Final HPE - Submucosal cystic benign neurofibroma.

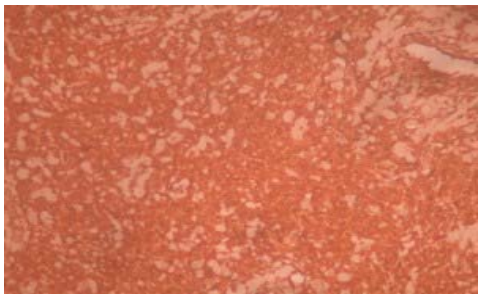
Immunohistochemistry : Neurofibromin ++ / DESMIN – HPE slide



intraoperative picture







Immunohistochemistry
Case is presented for its rarity -isolated visceral bladder neurofibroma.

DISCUSSION -
Neurofibroma is a benign, neoplastic tumor of the nerve sheath, composed of Schwann cells, perineurium-like cells, fibroblasts and intermediate-type cells. Neurofibroma of the bladder usually occurs in the setting of generalized neurofibromatosis type 1 rather than as isolated visceral neurofibromatosis. >50 cases of neurofibroma of the urinary bladder have been reported in the literature. Histologic findings are the same regardless of the site of occurrence.Immunohistochemical staining positive for S-100 protein & type IV collagen. Differential diagnosis-other spindle cell tumors such as Leiomyoma , Inflammatory myofibroblastic tumor, Postoperative spindle cell nodule, low-grade leiomyosarcoma, malignant peripheral nerve sheath tumor (MPNST), and Rhabdomyosarcoma.[3,4] Immunohistochemistry is useful in differentiating these tumors.Conservative treatment - transurethral resection or partial cystectomy, appropriate for patients without upper tract obstruction. When the lesion is extensive and symptomatic, radical cystectomy or pelvic exenteration often becomes necessary. Continued surveillance for malignant transformation is required.

REFERENCES:

1. Cheng L, Scheithauer BW, Leibovich BC, Ramani DM, Cheville JC, Bostwick DG.Neurofibroma of the urinary bladder. Cancer 1999;86:505-513.

2 Cabrera castillo PM, Alonso V Gregorio S, Cansino Alcaide JR, Aquilera Basan A, De la Pena Barthel JJ. Bladder neurofibroma: case report and bibliographic review. Arch Esp Urol. 2006;59:899-901.

3 Wang W, Montgomery E, Epstein JI. Benign nerve sheath tumors on urinary bladder biopsy. Am J Surg Pathol. 2008;32:907-12.

4 Lott S, Beltran AL, MacLennan GT, Montironi R, Cheng L. Soft tissue tumors of the urinary bladder, part I: myofibroblastic proliferations, benign neoplasms, and tumors of uncertain malignant potential. Human Pathology 2007;38: 807– 823.