

CASE REPORT

URETERIC ANGIOMYOLIPOMA CAUSING UNILATERAL PELVI-URETERIC JUNCTION OBSTRUCTION

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A 63 year old lady, presented to us with nonspecific abdominal pain. Ultrasonography (USG) and CT scan abdomen and pelvis, showed right moderate hydronephrosis, with no evidence of mass at pelvi-ureteric junction (PUJ) obstruction. Per-operatively mass upper ureter was found obstructing PUJ. Mass was excised and pyeloplasty done, with Double J (DJ) Stenting. Stent was removed after a week. Histopathology of specimen showed upper ureteric Angiomyolipoma.

Keywords: Pelvi-ureteric junction obstruction, Angiomyolipoma, Unilateral Hydronephrosis

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INTRODUCTION

Angiomyolipomas (AML) are unusual kidney tumours made up of blood vessels, muscle tissue and fat cells.¹ Extra-renal AML involving the genitourinary tract is rare with only few case reports.² Unilateral hydronephrosis is rarely caused by AML. To our knowledge there was no previously reported case of Angiomyolipoma of ureter that also caused unilateral hydronephrosis.

CASE REPORT

A 63 year old female presented to us with feeling of fullness after meal and vague pain in abdomen for one week. She had history of renal cell carcinoma in the family. On examination, the right kidney was palpable. Patient underwent USG abdomen that revealed right gross hydronephrosis. We did the CT scan abdomen/pelvis with and without IV contrast which showed gross hydronephrosis of right kidney with proximal slightly tortuous hydroureter with abrupt tapering. No definite was calculus noted in ureter. No mass could be identified near PUJ or upper ureter. We decided to do a cystoscopy, retrograde ureteropyelogram (RGUPG) and ureterorenoscopy

followed by pyeloplasty. Cystoscopy was normal. Retrograde ureteropyelogram was normal showing smooth outline of middle and distal third of ureter. Proximal ureter was very narrow with irregular lining and very narrow PUJ. There was a free floating tissue in proximal ureter, of which biopsy taken and sent for frozen section that came out to be benign inflammation. Right flank incision was given for open pyeloplasty. Per-operative findings were tough adhesions between kidney and surrounding tissue, and very tough tissue at PUJ causing complete blockage of right PUJ. Tissue was excised completely and also sent for frozen section that came out to be non-malignant. Right pyeloplasty was done, with DJ stenting.

Histopathology report showed there was a firm nodule measuring 1.6×1.0 cm, and microscopically it was well circumscribed nodule in muscularis of ureter. It was composed of proliferation of smooth muscle fibres, thick walled vessels and adipose tissue. Mild cytologic atypia of smooth muscle fibres was seen, it stained positive for HMB-45 stain (Figure-1), Diagnosis was Ureteric Angiomyolipoma.

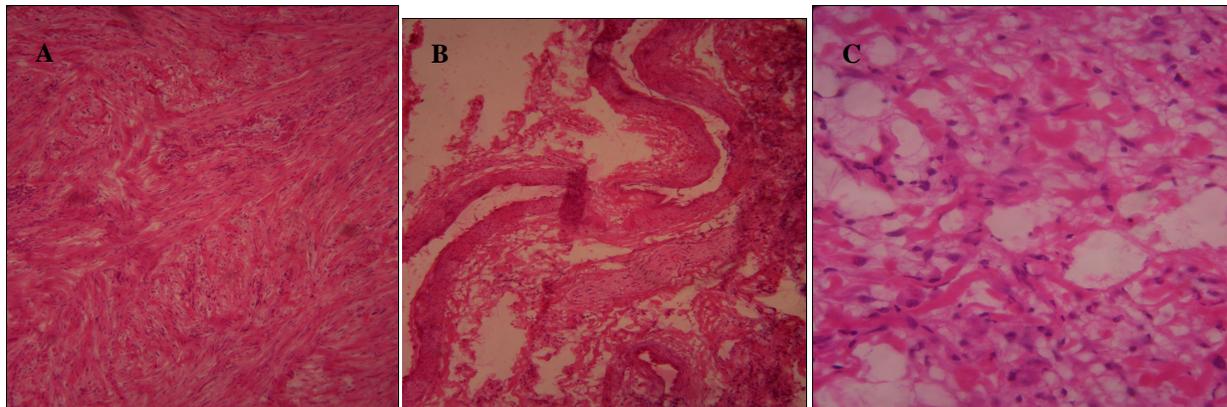


Figure-1: Histopathology slides of excised specimen showing (A) Muscle tissue (B) Thickened blood vessels (C) Fat cells

DISCUSSION

Extra renal AMLs are uncommon tumours but have been reported, most often in the liver and the uterus.³ Extra-renal AML involving the genitourinary tract is even rarer, though there are scattered case reports of AML involving left vesico-ureteric junction (VUJ), bladder wall and presenting as a bladder polyp.^{2,4,5} Diagnosis of AML is usually done on CT scan and ultrasound, although it becomes difficult in cases of fat poor AMLs. In fat poor AMLs, pathological confirmation is recommended. HMB 45 stain is employed to differentiate AML from sarcoma if significant cellular atypia is present. Our case was unique as it was AML of ureter causing ipsilateral hydronephrosis. Mass couldn't be identified radiographically and patient was operated for pain and hydronephrosis. Histopathology confirmed the diagnosis of AML and stained positive with HMB-45.

CONCLUSION

AML involving ureter is a rare case to date in literature, however keeping in view our case report; it should be included in differential diagnosis of PUJ obstruction leading to hydronephrosis.

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