

## Case Report

# Pelvi-Ureteric Junction Obstruction In A Malrotated Ectopic Kidney With Concomitant Lithiasis – A Rare Clinical Scenario

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## Abstract

Kidney lying outside renal fossa is referred as ectopic kidney. Hereby we are presenting a rare case of Pelviureteric junction obstruction in a malrotated ectopic kidney with concomitant lithiasis in a 16 year male. He presented with pain in lower abdomen intermittently. On CT scan, a left malrotated ectopic kidney was located in the pelvis near the midline with a 10 mm stone in the lower pole, and having obstructed pattern drainage with moderate hydronephrosis. Open Anderson-Hynes dismembered pyeloplasty was done.

Thorough clinical approach with delineating aberrant anatomy of ectopic kidneys and their vascular supply with consideration of unusual functional data on conventional functional renogram is necessary for intervening in malrotated ectopic kidney with concomitant lithiasis.

**Keywords:** Ectopic; lithiasis; Malrotated; Pelviureteric junction obstruction.

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## How to cite this article:

Siddiqui MA, Sharma S, Shaw V. Pelvi-ureteric junction obstruction in a malrotated ectopic kidney with concomitant lithiasis- a rare clinical scenario. *J Soc Surg Nep.* 2023; 26(1):49-51.

## DOI:

<https://doi.org/10.3126/jssn.v26i1.57394>

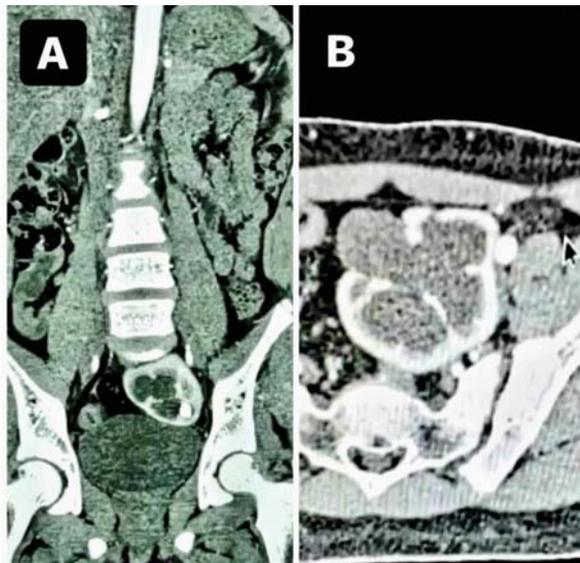
## Introduction

Congenital malformations of the genito-urinary system account for approximately 10% of all births. Upper tract anomalies contribute to 50% of these. Any kidney that lies outside the renal fossa since birth is referred to as ectopic kidney.<sup>1</sup> Pelvi-ureteic junction obstruction is seen in 22%–37% of ectopic kidneys. Abnormal renal anatomy in ectopic kidneys makes the management of symptomatic patients challenging.<sup>2</sup>

## Case Report

Hereby we are presenting a rare case of Pelvi-ureteric junction obstruction in a malrotated ectopic kidney with concomitant lithiasis in a 16 year old male. The patient

presented to us in OPD with complaints of pain in the lower abdomen intermittently. During evaluation, his blood parameters were found to be normal. On Computed Tomography, he had a left ectopic kidney which was malrotated with hilum facing anteriorly and located in the pelvic region just above the bladder near the midline having a stone in the lower pole of size 10 mm, and having obstructed pattern drainage with moderate hydronephrosis. The left renal artery was seen as a direct branch from left common iliac artery and renal vein was draining into left common iliac vein. However, the right kidney was normal. A functional renogram was done with F-15 protocol, which showed Glomerular filtration rate of 70 ml/minute of right kidney while 13 ml/minute of left kidney. However, a remark was made by the consultant that the left kidney which is located ectopically and is more anterior, the

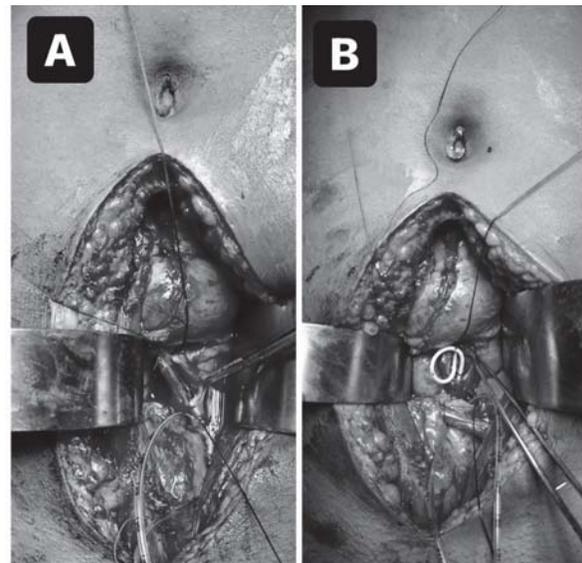


**Figure 1.** Radiological Image showing malrotated ectopic kidney in the pelvis region with a lower pole calculus with hydronephrosis- A. Coronal section. B. Axial section

quantitative information drawn using posterior gamma detector will not be accurate. Hence, repeat functional renogram was done using same F-15 protocol where the gamma detector was located on the anterior side and it reported a function of 25ml/minute on the left side and 29 ml/minute of the right side. Considering the different data and different positions of gamma detector, baseline GFR of left kidney was 25 ml/minute and of right kidney was 70 ml/minute. In view of patient being symptomatic with concomitant lithiasis, an open Anderson-Hynes dismembered pyeloplasty was done. Intraoperatively, an infraumbilical midline incision was taken and we tried to remain extraperitoneal. The left kidney was found near the



**Figure 3.** Post op radiograph showing position of DJ stent



**Figure 2.** Intra-op image showing -A. ectopic kidney with Anderson Hynes dismembered Pyeloplasty being done B. Pyeloplasty with DJ stent.

midline above the bladder, dissection was done to trace the pelvis and the ureter. Pelvis and ureter were dismembered, and stone was removed intact, the lateral end of ureter was spatulated and anastomosed with the pelvis in a tension free water tight manner over a DJ stent. Post operative period was uneventful and patient was discharged and is currently in follow up.

## Discussion

During the course of embryogenesis, when the kidney fails to ascend upwards from the pelvis in its metanephros phase, a number of anatomical abnormalities can be seen. Most commonly an ectopic kidney is seen inside the pelvis, opposite the sacrum and below the aortic bifurcation. However, most cases of pelvic kidney are asymptomatic, despite the risk of traumatic injury, urinary tract infections, renal calculi, and other urological problems; only 1 in 10,000 patients were clinically recognized.<sup>1,3</sup> Our case had an ectopic kidney which was malrotated and located in pelvis and patient was symptomatic.

PUJO is seen in 22%–37% of ectopic kidneys. The ectopia is typically related to malrotated unit with possible vascular aberrations.<sup>2</sup> Ectopic kidney may receive vascular access from a range of vessels like common iliac vessels, from the aorta, mid sacral vessels. Understanding this anatomy is vital for any urologist operating on patient with an ectopic kidney. An unusual anatomy with aberrant vascular supply poses difficulty in management, and it is, therefore, important to understand the anatomy and vasculature of pelvic kidneys.<sup>5</sup> In our case the left renal artery supplying the kidney was arising from the left common iliac artery.

Concomitant lithiasis of the urinary tract is not always

unusual and whether it co-exists as a separate entity or is the end result of a narrow renal outflow tract continues to be debated. The occurrence of lithiasis in renal malformations is believed to be higher than the general population. In a retrospective analysis of 1639 patients for the duration of forty-five years by Mayo Clinic, there was 70-fold increased prevalence in malformed kidneys. Overall, various factors seem to be responsible for lithiasis in PUJO other than urinary stasis. Metabolic abnormalities and genetic predisposition with associated urinary tract infection plays a role.<sup>6</sup> Our case also had an associated calculus in the lower pole of left ectopic kidney.

On evaluation of PUJO with nuclear functional imaging studies, further intervention should be done as per the degree of obstruction, renal function, patient symptoms and stone burden. Simultaneous treatment of PUJO and stone should be considered.<sup>6</sup> Our case underwent treatment of PUJO with removal of the stone in the same sitting.

In 1949, Anderson and Hynes defined open dismembered pyeloplasty method to treat PUJO. This was one of the proven versatile, secure and effective technique having 95% success rate. Till date, it is still regarded as gold standard for pyeloplasty.<sup>4</sup> Our case also underwent Anderson and Hynes open dismembered pyeloplasty.

## Conclusion

A thorough clinical approach with delineating aberrant anatomy of ectopic kidneys and their vascular supply with consideration of unusual functional data on conventional functional renogram is necessary for intervening in malrotated ectopic kidney with concomitant lithiasis so as to prevent long term complications like chronic kidney disease and non-functioning kidney.

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