Acute Calyceal Obstruction In an Adolescent with Infundibulopelvic Dysgenesis

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Introduction

Infundibulopelvic dysgenesis is an obstructive process of the renal collecting system that is associated with a variety of congenital malformations [1].

Patients with this condition often have associated abnormalities such as multicystic dysplastic kidney (MCDK), infundibulopelvic stenosis, and calyceal diverticula [2].

While some patients may remain asymptomatic, others may experience intermittent flank pain, depending on the degree of obstruction.

Rarely, patients will present with acute onset flank pain resembling that of acute ureteral obstruction.

Our case describes the unusual presentation of a 14-year-old adolescent, with no previous symptoms found to have significant hydrocalycosis and pain secondary to acute obstruction of a stenotic infundibulum.

Case

HPI: 14-year-old male with no past medical history presented to the emergency department with a chief complaint of acute onset nausea, vomiting, and flank pain.

Vital signs were unremarkable.

Relevant physical exam findings: Exquisite right flank tenderness to palpation.

Laboratory findings were unremarkable.

Imaging: A contrasted CT scan with delayed films was then performed and revealed a delayed right nephrogram as well as a massively dilated central calyx. A nephrostogram was performed with the catheter in dilated calyx. This study revealed no communication with the true collecting system.

A Nephrostomy tube was placed without complication. The patient was discharged home.

The patient presented as an outpatient for diagnostic uretero-renoscopy. Retrograde pyelogram revealed a dilated megaureter and a grossly abnormal collecting system. There was an abnormally shaped renal pelvis with multiple stenotic infundibular segments leading to malpositioned calyces. Methylene blue was injected through the nephrostomy tube for better identification of the stenotic infundibulum.

A punctate stone was encountered at the entrance of a stenotic calyx. Laser lithotripsy was used to fragment this stone. Methylene blue was immediately seen emanating from this stenotic calyx. A retrograde pyelogram was again performed and now suggested continuity between the two systems. A double J ureteral stent was placed to allow for healing and decompression of the collecting system.

Repeat antegrade nephrostogram was performed, which confirmed resolution of the obstruction. The patient recovered well and has remained without flank pain.

Initial Imaging

Figure 1: Axial view on CT abdomen pelvis

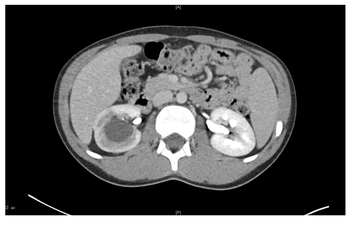
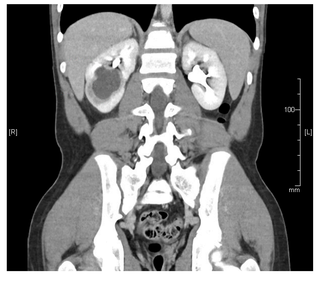


Figure 2: Coronal view on CT abdomen pelvis



Operative Imaging

Figure 4: Nephrostomy tube placement

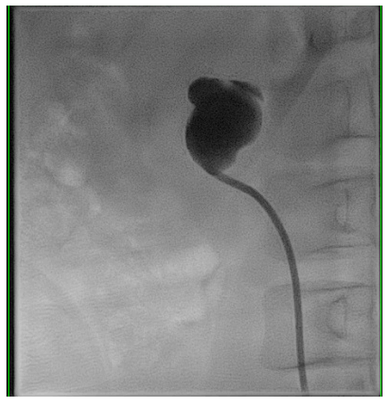


Figure 4: Placement of nephrostomy tube into a dilated central calx. Contrast has been injected through the nephrostomy tube. There is no communication between this calyx and the rest of the collecting system.

Figures 5 and 6: intra-operative fluoroscopy - Contrast has been injected through the nephrostomy tube in an antegrate fashion. Contrast has also been injected through the ureteroscope in a retrograde fashion. The stenotic entrance to the dilated calyx is triangulated using a combination of live fluoroscopy and direct ureteroscopic visualization.

Figure 5: Intra-operative fluoroscopy

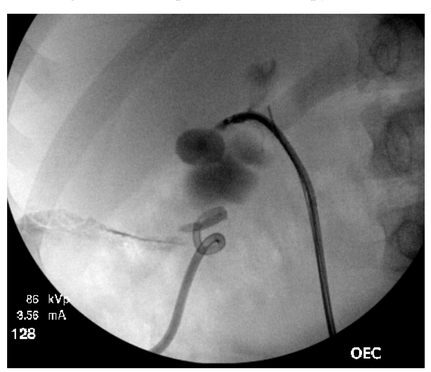
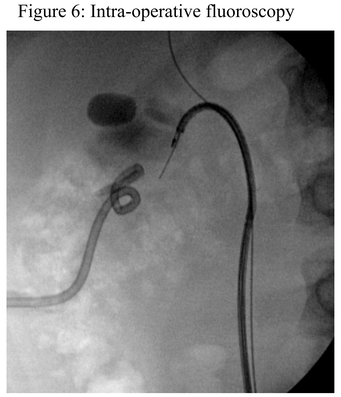


Figure 6: Intra-operative fluoroscopy



Post-operative imaging

Figure 7,8: Post-operative antegrade nephrostogram

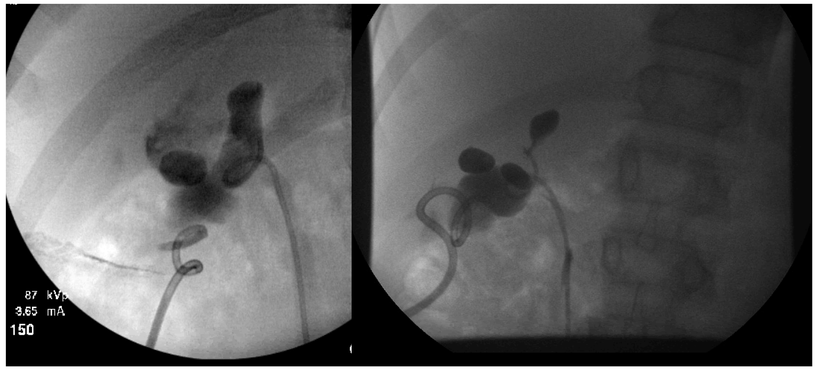


Figure 7,8: Antegrade nephrostogram was performed 1 week post operatively and revealed communication between the dilated calyx and the rest of the native collecting system.

Discussion and Conclusion

This case highlights the unusual presentation of acute stone obstruction of a stenotic infundibulum in a patient with infundibulopelvic dysplasia.

This is a rare presentation, as such cases of stenotic infundibulum are generally asymptomatic. Further, when symptomatic these cases are often associated with chronic vague symptoms and long term obstruction.

Often symptoms may go unnoticed for many years, resulting in renal scarring and diminished function.

In such cases, an endoscopic approach is not feasible. Partial nephrectomy, or radical nephrectomy may be needed.

This case serves to present management strategies for this atypical presentation.

References

Bayne CE, Peters CA. Congenital infundibulopelvic stenosis: Indications for intervention, surgical technique, and review of literature. J Pediatr Urol. 2016

Uhlenhuth E, Amin M, Harty JI, Howerton LW. Infundibulopelvic dysgenesis: a spectrum of obstructive renal disease. Urology. 1990