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Case report

Ask-Upmark kidney with bilateral pelvi-ureteric junction obstruction – A rare entity

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Developmental arrest

Abstract
Renal segmental hypoplasia (Ask-Upmark kidney) is a congenital disorder, first described by Eric Ask-Upmark in 1929. Habib et al. called it "segmental hypoplasia of the kidney in 1965. Ask Upmark kidney is more in females and present with hypertension or sometimes as recurrent urinary tract infections. Usually unilateral, bilaterally asymmetrical segmental hypoplasia has also been reported. The pathogenesis of Ask-Upmark kidney is controversial, attributing to vesicoureteral reflux (VUR) with intrarenal reflux or possibility of localized developmental arrest. We report a case of two years' male child presenting as abdominal swelling with respiratory distress. On evaluation he was found to have bilateral pelvi-ureteric junction obstruction and left non-functioning kidney for which he underwent right pyeloplasty and left nephrectomy later on. Histopathology report of left kidney suggestive of segmental renal hypoplasia (Ask-Upmark kidney).

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Introduction

Renal segmental hypoplasia (Ask-Upmark kidney) is a rare renal developmental anomaly which usually presents in young female as hypertension and/or its complications. Pathogenesis of renal segmental hypoplasia remains controversial. We report an unusual association of renal segmental hypoplasia with bilateral pelvi-ureteric obstruction in a male child. The case is unique in presentation and this association may reveal new aspects of this rare entity.

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Case report

A two years' male child presented with left flank fullness since 3 months. Lump was painless, gradually increasing in size and associated with difficulty in breast feeding and respiration. On general physical examination, temperature – 98.6 F, pulse rate – 122/min at radial artery in supine, respiratory rate – 36/min and blood pressure – 108/56 mmHg, were recorded. On local examination, abdomen was distended with soft cystic lump palpable in bilateral flank region. Left flank lump was approximately 10 cm × 8 cm and right lump was approximately 8 cm × 6 cm in vertical and transverse dimensions respectively. Hernial sites and external genitalia were normal without any surgical scar. Laboratory investigations displayed hemoglobin 9.6 g/dl, total leukocyte count 12,000 mm^{-3}, blood urea 26 mg/dl, serum creatinine 0.71 mg/dl, serum sodium and potassium 130 and 4.5 mmol/L respectively. Urine analysis was having 1–2 pus cells/high power field and urine culture was sterile. Ultrasonography abdomen revealed left kidney grossly hydronephrotic with thinned out parenchyma (Fig. 1) and right kidney with moderate hydronephrosis. On further study, 99m technitium-ethylene cysteine renal scan revealed a non-visualized nonfunctioning left kidney

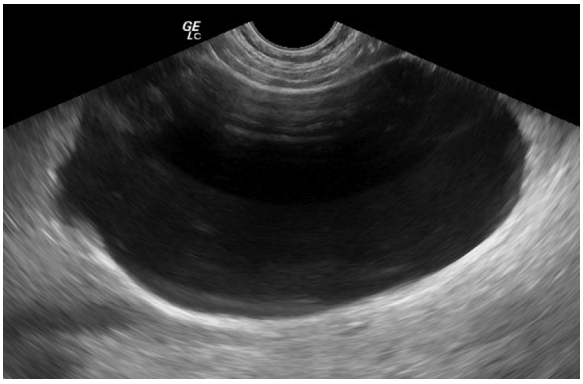

Fig. 1 Ultrasonography abdomen showing left kidney grossly hydronephrotic with thinned out parenchyma.

with grossly hydronephrotic obstructed right kidney with preserved function (Glomerular filtration rate – 50.1 ml/min) (Fig. 2). Patient presented with bilateral obstructive uropathy with respiratory distress and difficulty in breast feeding so patient was planned for

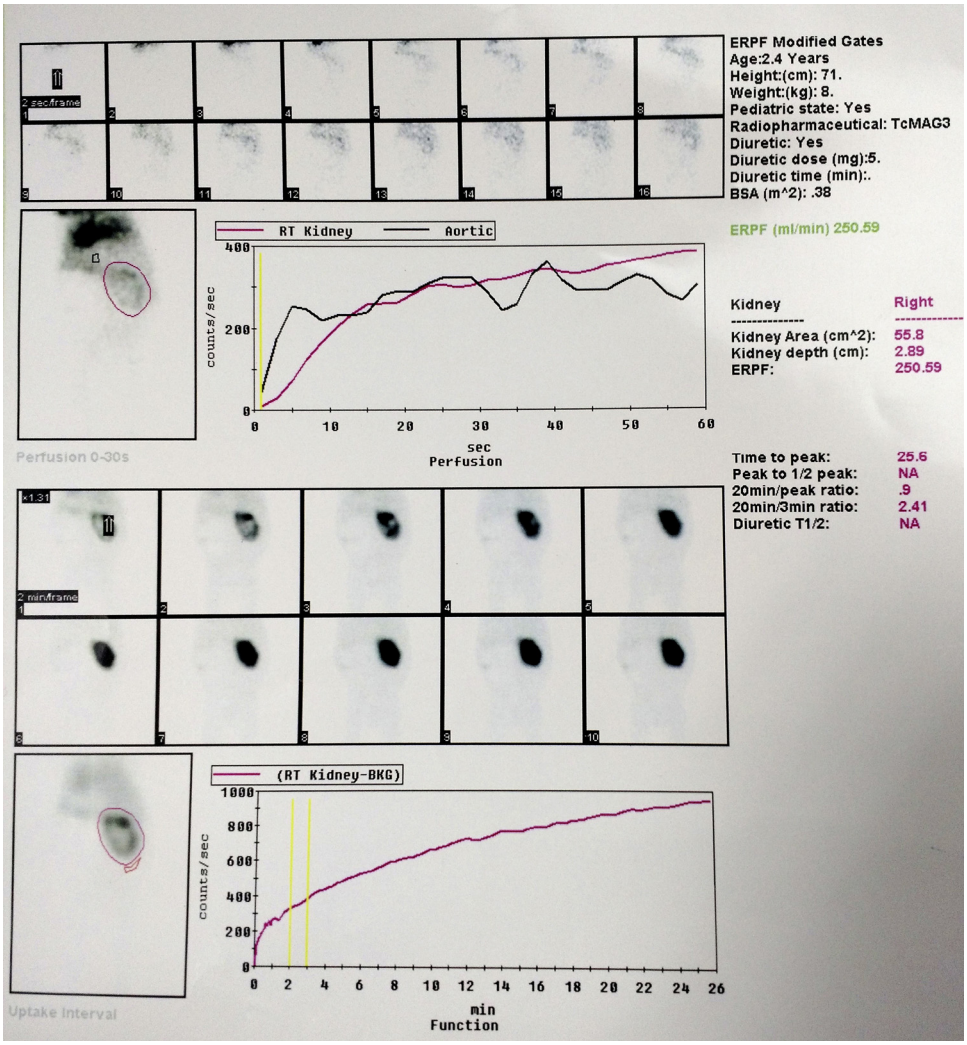

Fig. 2 99m technitium-ethylene cysteine renal scan shows a non-visualized left kidney with grossly hydronephrotic obstructed right kidney with preserved function.

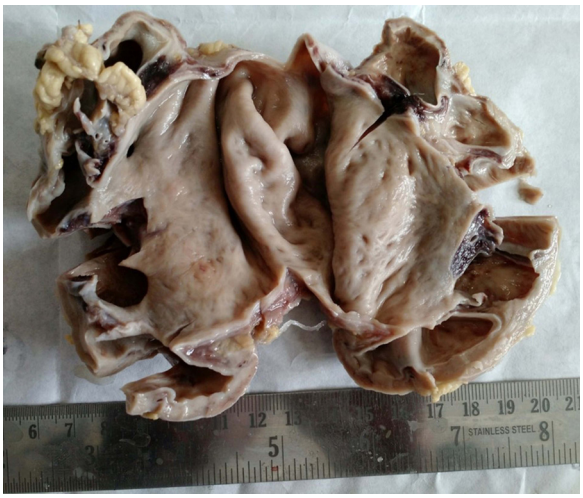

Fig. 3 Specimen left kidney: gross examination shows left kidney smooth, grossly dilated with thinned out parenchyma.

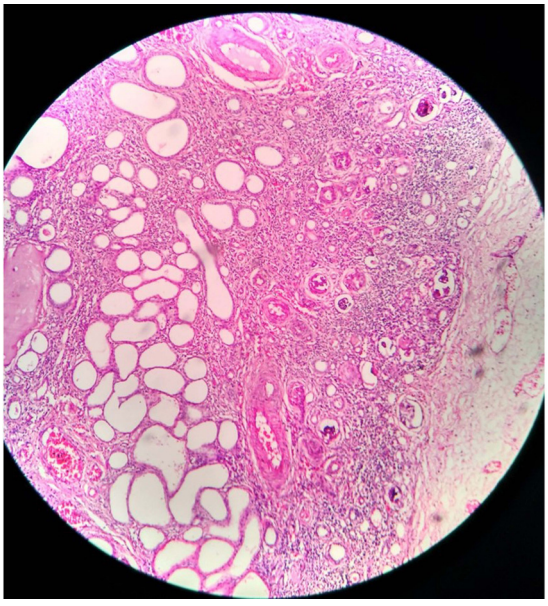

Fig. 4 (Hematoxylin and Eosin, 40×): histopathological examination of resected left kidney shows thyroidisation of tubules, thick walled arteries and absence of glomerulus with sharp delineation from surrounding normal renal parenchyma.

right side pyeloplasty and left side percutaneous nephrostomy tube placement under general anesthesia. Postoperative period was uneventful with marked improvement in respiratory pattern. Patient underwent left side open nephrectomy after 3 weeks. On gross examination, left kidney was smooth, grossly dilated with thinned out parenchyma. Ureteric stump of approximately 1–2 cm was removed. Specimen was sent for histopathological examination (Fig. 3). Histopathological examination of resected left kidney revealed thin cortex with thyroidisation of tubules, thick walled arteries and absence of glomerulus with sharp delineation from surrounding normal renal parenchyma. Sections from ureter showed thickened epithelium with underlying stroma showing dense mixed inflammatory infiltrate. Histopathological findings were consistent with segmental renal hypoplasia (Ask-Upmark kidney) (Fig. 4). Patient was discharged in satisfactory general condition with normal feeding and respiratory pattern and normal blood pressure.

Discussion

Majority of the patients of Ask-Upmark kidney present with hypertension or sometimes with history of recurrent urinary tract infections. More frequent in females [1,2]. Usually unilateral, but bilaterally asymmetrical segmental hypoplasia had also been reported [3,4]. Grossly, the hypoplasia is characterized by capsular grooves and decrease in number of pyramids [5]. We reported a case presenting as bilateral pelvi-ureteric junction obstruction with segmental hypoplasia in left non-functioning kidney. As described in the literature, hypoplastic areas contain no glomeruli but have thyroid like tubules and thick walled arteries [4,5]. This case exhibited similar findings with thickened vessels and thyroidisation of the tubules and absence of glomeruli with sharp delineation from normal renal parenchyma. The pathogenesis of the Ask-Upmark kidney is still controversial. Vesicoureteral reflux with intrarenal reflux proposed as a possible mechanism, however not all patients demonstrated a vesicoureteral reflux at the time of diagnosis. Shindo et al. conducted a radiographic and morphologic study of nine patients with renal segmental "hypoplasia at mean age of 2.9 years (range 0.1–10 years) and recognized a strong relationship between vesicoureteric reflux and renal segmental hypoplasia" [6]. Localized developmental arrest remained another explanation; however this alteration in metanephric development could probably be a consequence of an intrauterine reflux [3–5]. It had been argued that the absence of reflux at the time of diagnosis did not rule out as the initial cause of the hypoplastic kidney [7]. In our case, renal segmental hypoplasia was found in association with pelvi-ureteric junction obstruction, which indicated possibility of intrauterine/early neonatal intrarenal urine stasis and reflux as pathogenesis of Ask-Upmark kidney, as described in literature. Association of segmental renal hypoplasia with bilateral pelvi-ureteric junction obstruction is a rare association, not reported in literature till date as per author's best knowledge.

Conclusion

Segmental renal hypoplasia in grossly hydronephrotic non-functioning kidney supports intrarenal urine stasis and reflux as a possible pathogenesis of Ask-Upmark kidney. Ask-Upmark kidney may present in association with pelvi-ureteric obstruction with normal blood pressure.

Author's contributions

Dr Bhupendra pal Singh and Dr S.N. Sankhwar managed the case including surgical procedure. Dr Ashok Kumar Sokhal contributed in conception and design of the study and he worked on drafting the article or revising it critically for important intellectual content. Dr Durgesh Kumar Saini helped in acquisition of data.

Conflicts of interest

No authors have any conflicts of interest or financial ties to disclose.

Source of funding

None.

Consent

Oral consent was given by the patient for this case report.

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