

Keratinizing squamous metaplasia of the upper urinary tract in a child with a solitary kidney

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ABSTRACT

We report a rare case of keratinizing squamous metaplasia of the upper urinary tract in a child with a single kidney. Squamous metaplasia has rarely been reported in the upper urinary tract, and is even rarer in children.

Key words: Cholesteatoma, child, keratinizing, metaplasia, squamous

INTRODUCTION

Metaplastic changes in the urothelium of the upper urinary tract are relatively infrequent. No definite terminology has been used for this entity. The process may be secondary to chronic obstruction, infection or congenital.

We report a very rare case of case of keratinising squamous metaplasia (KDSM) of upper urinary tract in a child with single kidney and reviewed the etiopathogenesis of the disease.

CASE REPORT

A 3-year and 6-month-old male child presented to us with complaints of intermittent fever for 1 month. The parents complained of refusal for feeds and lethargy. He had a history of polydipsia and polyuria. Antenatal ultrasound scans had identified an absent right kidney with hydronephrosis of the left kidney. His parents

had been advised surgery for left pelviureteric junction obstruction at the age of 2 months, but they had refused.

At presentation to us, the child had chronic renal failure with severe metabolic acidosis. A large renal lump was palpable in the left flank. Blood investigations revealed Hb of 6 gm%, total leukocyte count of 15,900/cumm, serum sodium of 116 mmol/L, serum potassium of 2.1 mmol/L, serum creatinine of 2.8 mg/dL and blood urea nitrogen of 97 mg/dL. Urine examination revealed pyuria with significant growth of *Escherichia coli*. Ultrasound of the kidney revealed absent right kidney with gross hydronephrosis and parenchymal thinning on the left side. Debris was noted in the dilated pelvicalyceal system.

The child was stabilized in pediatric intensive care for fluid and electrolyte imbalance and was administered culture specific antibiotics.

The DTPA scan revealed non-visualization of the right kidney with severely diminished parenchymal uptake on the left side without any excretion over 24 hours.

After stabilization, he was taken for open left pyeloplasty. The kidney was grossly dilated with thinned-out parenchyma. The dilated pelvicalyceal system was full of an unusual, white creamy material with thin flakes, not resembling pus [Figure 1]. The system was also lined by the same material. The flakes were gently aspirated out and sent for culture and histopathology. Anderson-Hyne's dismembered pyeloplasty was performed with a nephrostomy tube. The post-operative course was uneventful. Histopathology revealed focal areas of mature squamous epithelium with keratinization. The flakes were suggestive of an acellular keratin-forming membranous sheath [Figure 2].

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Access this article online

Quick Response Code:	Website: www.indianjurol.com
	DOI: 10.4103/0970-1591.126916

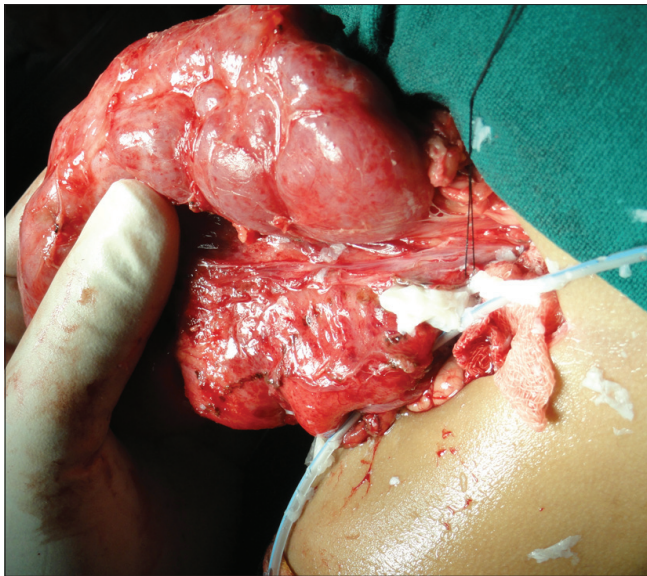


Figure 1: Intraoperative photograph showing dilated pelvis full of thin membrane-like flakes

At a follow-up of 1 year, the child is asymptomatic with normal renal function.

DISCUSSION

Keratinizing desquamous metaplasia (KDSM) is described as a condition in which the urothelium of the urinary tract is replaced with keratinized squamous epithelium.^[1] Although the condition commonly involves the lower urinary tract, KDSM of the renal pelvis and ureter is rare.^[2]

The condition seems to afflict older individuals in the third to sixth decades of life, with very few cases being reported in children.^[3] Most of the literature reported is in adults, and the terminology has been confusing.

Squamous metaplasia refers to replacement of the normal transitional epithelium by squamous epithelium and keratin production may or may not be present, atypia is not a feature and, when present, the condition is referred to as squamous metaplasia with dysplastic change, which is a more ominous diagnosis. Leukoplakia refers to the presence of a grossly discernible white patch on the mucosal surface and has no histological significance. Urinary cholesteatoma is nothing more than a mass of desquamated keratin lying free in the lumen of the renal pelvis or ureter, keratin ball. The latter is not without squamous metaplasia.

The term KDSM is descriptive of the histology involved. The cells are metaplastic, not dysplastic. Therefore, the basal cells are well-differentiated, completely normal squamous cells.^[3] For uniformization of the terminology, a histologic description was given in the form of KDSM by Hertle

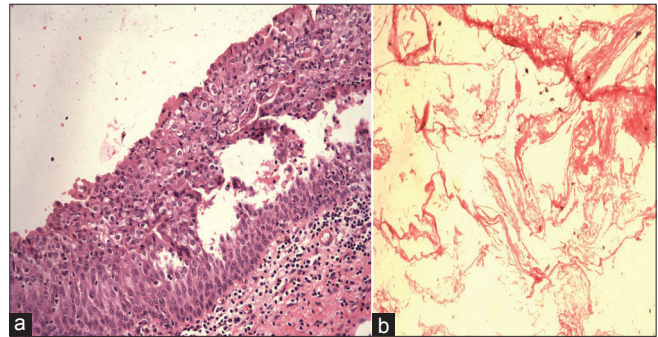


Figure 2: (a) Section of the renal pelvis with transitional epithelium along with squamous metaplasia. (b) Keratin flakes

and Andraulakakis.^[4] In their extensive review, squamous metaplasia of the transitional epithelium was noted with keratinization and subsequent desquamation. Majority of the lesions were noted in the renal pelvis, and were focal. Of the total 78 cases, only two cases of KDSM occurred in children. The exact etiology of this entity is unknown and various theories have been proposed. It may be a response of the urothelium to chronic inflammatory process like recurrent urinary tract infections, urolithiasis, hydronephrosis, etc,^[5] spontaneous epithelial migration^[6], or the condition has also been described as congenital in origin on the basis of the observations in the newborn and in children. Congenital presence of abnormal epithelial cells within the normal upper tract urothelium, originating from the primitive Wolffian duct may lead to this condition. In our patient, it is possible that the pathology was congenital rather than acquired. A congenital obstruction in a child, without recurrent episodes of infection, is less likely to cause metaplasia.

The condition is difficult to diagnose radiologically. Presence of radiolucent stringy defects in the renal pelvis with cornified epithelium in the urine should confirm the diagnosis.^[6] The condition has been inconsistently correlated with squamous cell carcinoma, as the progression from metaplasia to neoplasia has never been demonstrated. There is still debate as to whether KDSM is a pre-cancerous or more of a benign process. Some cases of KDSM have been seen in association with squamous cell carcinoma, but, in other cases, long-term observation has not revealed any cancerous transformation.^[7] Hence, close follow-up or conservative surgery has been recommended.^[4,8] KDSM has been described as being coincident with squamous cell carcinoma of the upper urinary tract in 8-12% of the cases; however, it has never been demonstrated that squamous cell carcinoma arises from KDSM.^[8]

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How to cite this article: Jain P, Mishra A, Misra DS. Keratinizing squamous metaplasia of the upper urinary tract in a child with a solitary kidney. Indian J Urol 2014;30:230-2.

Source of Support: Nil, **Conflict of Interest:** None declared.

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As a continuation of this series, three workshops are planned at SGPGI, Lucknow during 2014 as follows: (i) Workshop on 'Scientific Paper Writing' on April 18-20, 2014, (ii) Workshop on 'Basic Biostatistics' on July 18-20, 2014, and (iii) Workshop on 'Observational Studies' in September-October, 2014 (exact dates to be announced later).

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