Sigmoid Kidney Associated with Double Urethra

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Abstract An unusual case of crossed renal ectopia (sigmoid kidney) in an adult, compounded with double urethra, is being presented for its rarity and mode of presentation.

Keywords Sigmoid kidney · Duplication of urethra · Ureteric calculus · Intravenous urography

Introduction

Unilateral fused kidney with inferior ectopia is a rare congenital anomaly, where the long axes of both the kidneys are in continuous line with the pelvis facing in opposite direction assuming a 'S' or sigmoid appearance to the entire inner outline (see inset-Fig. 1). Such kidneys are source of trouble and urogram appearance is striking.

The estimated occurrence of sigmoid kidney is 1:1000 and a left to right crossed over occurs somewhat more frequently than shift in opposite direction [1].

This communication reports an adult patient with ureteric colic, which was subsequently diagnosed through USG and urogram as a case of sigmoid kidney associated with double urethra.

Case Report

A 31 year old male, was admitted in the hospital in April 2004 with the history of severe episodic pain in the left iliac fossa, usually characterised by left ureteric colic radiating to left scrotum and tip of the penis. He had been diagnosed and admitted for acute appendicitis with lump in recent past, and had been treated conservatively in some other hospital. The diagnosis of left ureteric colic was made in our hospital and was placed on expectant treatment.

Urinalysis showed numerous RBCs, and haemogram showed normal values. Straight x-ray abdomen discerned a radio opaque calculus at the lower end of left ureter, just above the urinary bladder.

Sonogram demonstrated a definable right renal parenchyma with a complex appearance of renal parenchyma in the lumbar region with absence of kidney elements on left side.

IVU revealed presence of both kidneys on right loin, a rare congenital anomaly with fusion of lower pole of right kidney with upper pole of left kidney, termed as "sigmoid kidney" (unilateral 'S' fusion). Left hydroureter and early hydronephrosis of pelvicalceal system was observed due to presence of a stone, 5 mm×5 mm size, in the lower third of the normally inserted left ureter, just above the U.V. junction (Fig. 1). Patient was placed on antispasmodics and forced diuresis. Fortunately on 3rd day he had retention of urine with strangury, due to impaction of urethral calculus. On inspection, he had two external urethral openings lying dorsiventrally. Palpation of ventral urethral meatus revealed an impacted stone, which could be retrieved. Dorsally placed urethra was found to be blind at the proximal end (Fig. 2).

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Fig. 1 IVU showing unilateral fused right kidney mass, lower moiety showing hydroureter and hydronephrotic change. Straight skiagram discloses a R.O. calculus at the lower end of ureter on left side. The in-set shows the anomaly in a line diagram

Discussion

Congenital diseases of the kidney, though present since birth, may manifest during childhood or in adult life with symptoms of urinary infection or upper urinary tract obstruction.

Crossed renal ectopia, with or without fusion, is rarely encountered among congenital anomalies, and may per chance be detected during investigation, as in the present case of left ureteric colic. Patient with cross ectopia have the tendency for infection, UPJ obstruction, calculus formation, apparently linked with the anatomical defects of the fused organs, which are conducive to the stasis of urine in the upper urinary tract and to disturbances of the circulation of lymph and blood.

In general, the occurrence of an associated anomaly in patients with crossed renal ectopia, excluding solitary crossed ectopia is low; the most frequent entities are imperforate anus (4%), orthopaedic anomalies (4%), skeletal abnormalities, and septal cardiovascular defects [1].

Crossed renal ectopia is a situation in which the fused mass of total kidney substance is located on one side of the midline. One moiety may be the seat of hydronephrosis, infection or calculus formation. The adrenal glands are normally situated [1]. Most individuals with crossed renal ectopia have normal longevity and prognosis. However, some patients, with a potentially obstructive collecting system, are at risk of developing urinary tract infection, or renal calculus formation or both [1].

Fig. 2 Photograph showing double urethra: The bougies show dorsiventral placement of urethral openings

Symptoms of crossed ectopia are rare. Patients generally present in adulthood with urinary tract infection symptoms or calculi as in the present case of ureteric colic, are thought to be secondary to stasis. The blood supply to the crossed kidney is usually anomalous. The ureters usually open normally in the bladder and are not ectopic; but at times the ectopic ureter, as it crosses the pelvis, may pass behind the rectum.

The clinical importance of this anomaly is linked with the difficulties in its diagnosis. Dystopic kidneys often receive unnecessary treatment for erroneously diagnosed diseases and may undergo unwarranted operation (appendectomy & laparotomy, the latter when a tumour is found in the abdomen close to the vertebrae) [1]. The abnormality is readily detectable on USG, IVU, CT or MRI.

In duplication of urethra, the two urethras commonly lie dorsiventrally in relation to each other, or may be side by side. Bladder, ureteric and urethral anomalies are usually associated with solitary crossed ectopic anomaly [2]. The Sigmoid kidney associated with double urethra is known in children [3], but the authors are unaware of any record of this anomaly in an adult.

Double urethra is usually associated with bladder and ureteric anomalies, but its occurrence in an adult, coupled with a sigmoid kidney is a very rare situation.

In brief this case report of sigmoid kidney, associated with double urethra in an adult, has been described with sequence of events, which led to detection of these anomalies.

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