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Segmental cystic dilatation of the ureter in a Sudanese adult. A case report

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ABSTRACT

Objective: Segmental cystic dilatation of the ureter is extremely rare in adults. This article presents a case of segmental cystic dilatation of the left ureter with proximal dilated upper left ureter and ipsilateral atrophied kidney, while the distal ureter with normal calibration.

Case: Excision of the cyst with proximal ureter and kidney was done. Segmental cystic dilatation of the ureter should be considered in the differential diagnosis of intra-abdominal cystic mass.

Keywords: Megaureter, Segmental megaureter, Segmental cystic dilatation, Adult

INTRODUCTION

Congenital anomalies of the kidneys and urinary tract represents abroad range of disorders which result from abnormalities in the development of the urinary collecting system, abnormal embryonic migration of the kidney, and abnormal renal parenchymal development (1). Megaureter is defined as the presence of an enlarged ureter with or without concomitant dilatation of the upper collecting system. The normal diameter of the ureter is 3 mm and in practice ureter of 7 mm and more is considered a megaureter (2). Megaureters are categorized as primary and secondary megaureters. Primary megaureter is related to those with idiopathic congenital alteration of the vesicoureteral junction. The primary megaureter is categorised into obstructed, refluxing, and non-refluxing non-obstructing types. Secondary megaureter is due to distal obstruction in the urethra, bladder, or distal ureter (3). However, segmental megaureter or segmental cystic dilatation of the ureter is a very rare entity, and only a few cases were reported in the literature (4, 5, 6, 7, 8, 9, 10, 11). This paper will present a case with segmental cystic dilatation of the ureter with dilated proximal upper ureter and atrophied ipsilateral kidney.

CASE

Thirty-Seven years old male presented to us in Ribat university hospital, Sudan, in September 2017 with slowly progressive abdominal swelling over two years period until it reached its maximum size and occupied the entire abdomen. The swelling was associated with dyspeptic symptoms and burning micturition. No loss of weight or loss of appetite. No abdominal pain, vomiting, jaundice, or change in bowel habits. The patient is not known as diabetic, hypertensive, or had a chronic illness. His past medical history was unremarkable. No family history of similar condition or malignancy. The patient is not a smoker or alcohol consumer.

On examination, the patient was well, not pale or jaundiced. He was on good nutritional and performance status. Abdomen was distended all over with full flanks, and the umbilicus was flat. Scar, visible pulsation, peristalsis, or dilated veins have not been observed. Hernial orifices were intact. There were a pelvi-abdominal mass about 35*25 cm, which firm, smooth, immobile, dull to percussion with positive fluid thrill. Systemic examination was normal. CT abdomen with oral and I.V. contrast was showed, large pelvi-abdominal cystic mass containing fluid with thin and smooth regular wall. No calcification, septation, or intra-lesion complex mass. The cyst was displacing the bowel to both sides of the abdomen with no feature of obstruction.

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The right kidney was normal. The left kidney was atrophied and not taking the contrast. No other abnormality. Possible differential diagnoses were mesenteric cyst and pancreatic pseudo-cyst.

Pre-operative workup included blood investigation, urine analysis, and cardio-respiratory assessment and all were unremarkable.

Exploration revealed a large retroperitoneal cystic mass about 35*25 cm and had been found to continue distally with normal caliber left ureter and proximally with dilated left upper ureter ipsilateral atrophied left kidney. A urologist was consulted and involved in the surgery. The consent was taken for removal of the left kidney and ureter. The cyst was removed along with the dilated upper ureter and kidney after ligation and division of the left renal pedicle and left gonadal vessels. The normal distal ureter was ligated and left in situ. The cyst was occupying the left mid-ureter.

The kidney, ureter, and cyst weigh 7.7 kg. The cyst size was 34*23.5*7 cm with thin walls containing hemorrhagic fluid. The attached left kidney and cystically dilated upper ureter measure was 27*5*2 cm. the cut surface of the kidney showed a multi-locular cyst and no renal tissue was identified (figure 1).

Figure 1: Ureteric cyst with proximal dilated ureter and atrophied kidney (K).

Microscopically, the cyst showed a thin fibrous wall lined with partially attenuated epithelium. The attached kidney showed cystic spaces lined with urothelium replacing renal parenchyma. And the ureter was lined by hyperplastic urothelium. No evidence of malignancy in the specimen.

The postoperative course was uneventful. No postoperative complications from the surgery or anaesthesia. Follow up in outpatient clinic to assess the function of remaining kidney, no raise in renal functions or any abnormality seen. The patient is in good health and on follow-up every 6 months with renal function test and abdominal ultrasound.

DISCUSSION

Few cases of segmental megaureter and segmental cystic dilatation were reported in the literature and most of them were in children. In 1986, Mandell et al reported 4 cases of congenital megacalycosis associated with ipsilateral segmental megaureter in children. Of their cases, the megaureter is due to narrowed a peristaltic ureter (4). In 1995, Ramaswamy reported one pediatric patient with segmental megaureter with sparing of proximal and distal ureter (5). A similar case was reported in 2010 by Karman et al in two months old infant with segmental ureteral dilatation with sparing of proximal and distal ureter (6). In 1997, Pinter et al reported a case with bilateral segmental megaureter (7). Soler et al. reported a case of multi-cystic dysplastic kidney and contralateral megacalycosis associated with ipsilateral distal segmental megaureter, in 2004 from Spain (8). In 2007, Prieto et al reported a case of congenital severe mid-ureteral dilatation associated with mild proximal ureteral dilatation and normal calibre of distal ureter similar to our case (9). In 2015, Dutta and Harsh reported a 7 years old male child with segmental megaureter and ipsilateral megacalycosis and contralateral vesico-ureteric reflux. Cystoscopy of this child revealed an absence of ipsilateral ureteric meatus and golf hole opening of refluxing ureter. The ureter distal to dilated part is of normal calibre but opened in the bladder neck (10).

We found only 2 cases of congenital segmental megaureter in adults associated with urolithiasis in the literature, which Rosenblatt et al. reported in 2009. The first one (58 years old) had a 2.3 cm stone in a dilated segment of his left distal ureter with normal-sized proximal ureter and mild dilatation of ipsilateral collecting system. The other case is 48 years old with dilated left distal ureter with ipsilateral 9 mm renal stone (11).

Several theories have been speculated regarding the pathophysiology of congenital segmental megaureters. Ramaswamy suggests that the segmental megaureter was a variant of non-refluxing megaureter and attenuated nexuses and thin myofilaments might be responsible for this entity, as their case (5). In another study, it was suggested that aganglionosis of the distal segment of the ureter resulted in dilatation of the proximal segment as in cases of achalasia and Hirschsprung's disease (4). Pinter suggests that recanalization of solid ureteral duct, if abnormal, might produce segmental megaureter (7).

The current belief is that primary obstructive megaureter is present primarily in adults when the congenital abnormality does not cause symptoms or illness and is not seen via an imaging study performed during childhood.

Spontaneous regression fails to occur, yet patients remain asymptomatic during childhood. Eventual symptoms include urinary tract infection, renal parenchymal disease, and recurrent stone formation (10).

Treatment of segmental cystic dilatation of the ureter should be tailored according to ipsilateral kidney function and the length of the segment involved (6). If the residual length of the proximal and distal ureteral segment is enough with good ipsilateral kidney function, end to end uretero-ureterostomy after excision of the cyst can be performed. If excision is not possible, tailoring or trimming of the dilated segment should be considered. If the ipsilateral uretero-renal function is poor, hypoplastic, or dysplastic, a nephroureterectomy is advised, as in our case.

CONCLUSION

Segmental cystic dilatation of the ureter should be distinguished from ureterocele and other conditions such as ureteral diverticulum. At the same time, patient should be investigated for possible associated urinary tract system anomalies such as megacalycosis, duplication of the collecting system, and hypoplastic, dysplastic, or non-functioning kidney (6).

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