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Crossed fused renal ectopia with vaginal agenesis – a case report

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Abstract

Ectopic kidney occurs as a result of a halt in migration of kidneys to their normal location during embryonic period. Due to their ectopic position and aberrant vascularity, they are more prone to many diseases such as urinary tract infection, renal stones, hypertension etc. Crossed fused renal ectopia is frequently associated with other congenital anomalies. Here we presented a case report of crossed fused renal ectopia with vaginal agenesis for its rarity and clinical significance.

Key Words: Crossed fused renal ectopia, vaginal agenesis

INTRODUCTION

Crossed fused renal ectopia refers to a rare congenital anomaly where the kidneys are fused and located on the opposite side of the midline in relation to its ureteric orifice in the urinary bladder.[1] It is remarkable for its associated anomaly in the urogenital tract and other systems.[2] Usually it is asymptomatic, but may cause diagnostic problems when acute disease develops in the kidney[3]. Here our patient had two anomalies-crossed fused renal ectopia and vaginal agenesis. This case is presented here for its rarity and clinical significance.

CASE REPORT

A 17 year old female patient presented with cyclical lower abdominal pain for last one year. Clinical examination revealed absence of vagina with normally developed secondary sexual characteristics. Routine laboratory investigations were normal. On ultrasonography, uterus with endometrial collection, absence of kidney on right side and a large left kidney with possibility of unilateral fused kidney was identified. Computer tomography intravenous pyelography(CTIVP) (FIGURE I) revealed no kidney on right side and unilateral(large) fused kidney with anterolaterally facing hilum of both kidneys on left side with normal left ureter and ureter of crossed fused right kidney crossed the midline and had normal course in its lower 1/3rd to open into the normal right vesicourteric junction. Left renal vessel was normal and that of other kidney was narrow and measured 3 mm at origin. Uterus was anteverted with collection inside. Diagnosis of crossed fused renal ectopia with vaginal agenesis was thus established.

DISCUSSION

Crossed fused renal ectopia is a very rare congenital abnormality with incidence of 1 in 1000 live births[4]. It is two times more common in males than females,[4] which further added

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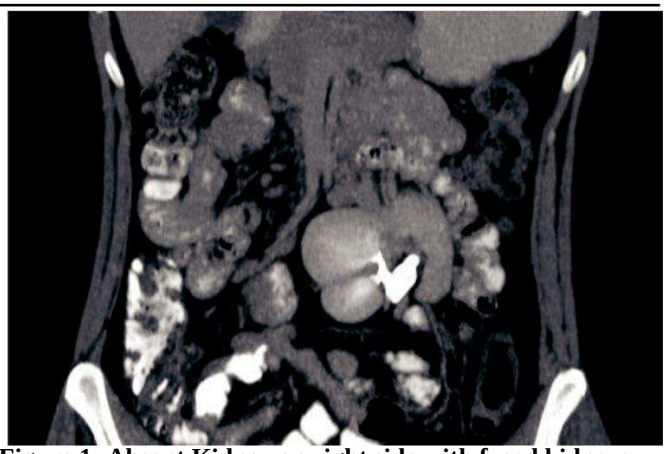

Figure 1: Absent Kidney on right side with fused kidneys on left side with anterolatterally facing pelvises

the rarity in our case as in our case, a female is affected. However, it is common on left side as in our case.[4]

Renal fusion and anomalies were first studied and classified by wilmer, (1938) later it was revised by McDonald and McClellan, (1957). They classified the ectopic sequel into four major types as:[4]

(1) Crossed ectopia with fusion which is further subclassified depending on extent of fusion and rotation into

(a) Unilateral fused kidney with inferior ectopia

(b) Sigmoid or S shaped kidney

(c) Unilateral loop kidney

(d) L shaped kidney

(e) Unilateral fused kidney with superior ectopia

(2) Crossed ectopia withour fusion

(3) Solitary crossed ectopia

(4) Bilaterally crossed ectopia

In our case, it was crossed renal ectopia with fusion of loop

type with anterolatterally facing pelvises

The factors responsible for this type of ectopia and fusion were still undetermined. However, Wilmer suggested that crossover occur as a result of pressure from abnormally positioned umbilical arteries that prevent the normal ascent of the kidney which then follows a path of least resistance to the opposite side. Other suggested factors are faulty ureteric bud development, teratogenic factors, abnormal variation in the growth of the hind gut, abnormal position of caecum.[4]

Crossed fused ectopia of kidney is frequently associated with other anomalies of cardiovascular system[2] (dextrocardia, aortic and multiple iliac aneurysms, Takayasu's arteritis), imperforate anus, skeletal abnormalities,[5] multicystic dysplasia in a fused or unfused cross kidney,[6] ureterocele, patent urachus[2], hydronephrosis, ectopic ureteral orifice, vaginal agenesis, hypospadias.[7,8] In our case crossed fused ectopia of kidney is associated with vaginal agenesis.

Most of the presenting symptoms of crossed renal ectopia are nonspecific and most cases remain asymptomatic through their life and are diagnosed incidentally,[2,4,9] as in our case. However, it frequently predisposes to certain complications like urolithiasis, urinary tract infections and hypertention[4]. Therefore its early detection not only helps in prevention of these diseases but awareness of presence and association of crossed renal ectopia also helps to avoid iatrogenic injuries to the kidneys and /or the ureters during abdominal and pelvic surgeries[9]. Ultrasonography, CTIVP, Renal Scintigraphy have presently become very important for the evaluation of such cases.[9] In our case, CTIVP revealed narrow vessel of crossed fused right kidney which may responsible for hypertention in future.

CONCLUSION

Most patients with crossed fused renal ectopia are asymptomatic and diagnosed incidentally. Other congential anomalies always must be identified in its presence. As it predisposes to certain complications, sophisticated investigations are needed for its early diagnosis.

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