Afr. J. Med. med. Sci. (1987) 16, 187–191.

Renal ectopia with and without fusion in Ibadan, Nigeria

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Summary

Thirty-five patients with renal ectopia have been received. Forty-three per cent of the patients presented with a palpable lower abdominal mass, abdominal pain and gastrointestinal symptoms. In 37% of the cases it was an incidental finding at urography done for other conditions. The vagueness of symptomatology and findings in renal ectopia indicate that a high index of clinical suspicion is needed in its diagnosis with subsequent performance of excretory urography. Needless laparotomy, as undertaken in three children in the series, will, thus, have been avoided.

Résumé

Les cas de trente-cinq patients souffrant d'ectopie rénale sont passés en revue. 43% des cas examinés se présentent avec une masse abdominable inférieure palpable, accompagnée de douleur admoninale et des symptomes gastro-intestinaux. Dans 37% des cas, l'urographie pratiquée pour d'autres indications ont révélé l'incidence de la maladie en question. Le caractère vague de la symptomalogie et des constats dans l'ectopie rénale indique la nécessité d'un index élevé de soupçon clinique pour le diagnostic, suivi de l'urographie excrétoire. La laparotomie, pratiquée inutilement dans le cas de 3 enfants de la série, serait alors évitée.

Introduction

Among the body organs, the kidneys are unusual in that they ultimately lie at a level, which is higher than that of their site of embryologic origin. Having arisen deep in the pelvis in the first few weeks of foetal life, they gradually ascend to their final anatomic position. Such a complex embryologic development can produce many renal and ureteric anomalies, among which renal ectopia and fusion are frequent (Farman, 1968). This paper documents experience with thirty-five patients with renal ectopia with and without fusion.

Renal ectopia is defined clinically as a kidney that is located in an abnormal anatomical position. It may be iliac in type when it is situated in the iliac fossa. That below the brim of the bony pelvic inlet is considered a pelvic kidney while a kidney located above the diaphragm is intrathoracic.

Patients and methods

Thirty-five cases of renal ectopia with and without fusion were diagnosed in the 4-year period 1981–1984 at the University College Hospital, Ibadan (Table 1). The clinical features leading to the diagnosis are as summarized in Table 2. The diagnosis was confirmed in a few cases by ultrasonography and in all cases by excretory urography. Three patients who had obstructive uropathy of their ectopic pelvic kidneys also had renal angiography and micturating cysto-urethrogram. The latter investigation was done to exclude vesico-ureteric reflux while angiography was done for contemplated surgical intervention since nephrectomy in such cases can be difficult and blood loss excessive owing to multiplicity and variability of their blood supply.

Results

The age at presentation ranged from 1 week of life to 70 years with the majority presenting between the ages of 25 years and 55 years.

Table 1. Classification of thirty-five cases of renal ectopia

Classification			Number of cases
Simple renal ectopia (without fusion)			
	Unilateral:	Thoracic	1
		Iliac	10
		Pelvic	14
	Bilateral:	Pelvic	1
Crossed renal ectopia with fusion (iliac)			1
Fused ectopic and horse shoe pelvic kidneys			8
Total	Thoracic	1	35
	Iliac	11	
	Pelvic	23	

(1), prune bell syndrome (1), testicular feminzation syndrome with urethro-vaginal fistula (1), and spina bifida occulta defect of L5 and S1 (2).

Management was conservative and symptomatic in all except in the three cases who had nephrectomy for gross hydro-nephrosis and ureterohydronephrosis. Figure 1 is an intravenous urogram on a child with a palpable left abdominal mass. The procedure revealed gross hydronephrosis of the left ectopic kidney. Of particular interest in the series are three children who underwent needless laparotomy, (two for suspected intussusception and one case for suspected appendix mass), only to discover that the palpable pelvic mass in each case was an ectopic pelvic kidney. All the fourteen adult female patients have had uncomplicated pregnancies and deliveries. In four of the pregnant women an associated 'pelvic tumour' was palpable with a suspected diagnosis of an overian tumour or uterine fibroid. Ultrasonography and elective post-partum urography revealed that the pelvic masses were indeed ectopic kidneys (Fig. 2).

Table 2. Presentation in thirty-five cases of renal ectopia

Presentation	Number	%
Palpable lower abdominal mass	15	43
Abdominal or flank pain	14	40
Gastro-intestinal symptoms: nausea, vomiting and/or diarrhoea	10	28.6
Gross and microscopic haematuria	6	17.1
Proven urinary tract infection	4	11.4
Enuresis	2	5.7
Incidental finding at urography for conditions such as benign prostatic hypertrophy, vesico-vaginal fistula, carcinoma of cervix, uterine fibroid, hypertension and urethral stricture	13	37

There were twenty-four adults with a male to female ratio of 10 : 14. Eleven cases were seen in the paediatric age group with a male to female ratio of 6 : 5. The overall male to female ratio was 16 : 19, which appears statistically insignificant.

In simple unilateral renal ectopia, fourteen cases occurred on the left and eleven on the right (Table 1). There was a case of an asymptomatic left ectopic intra-thoracic kidney in a 70-year-old man who was being managed for urethral stricture.

Co-existing congenital abnormalities were found in eleven cases: renal malrotation (4), imperforate anus (2), posterior urethral valves

Discussion

Renal ectopia with, and without, fusion are relatively uncommon (McDonald & McLellan, 1957), while ectopic intrathoracic kidney is an even rarer developmental anomaly with only about fifty cases reported in the world literature (Kirshenbaum, Puri & Race, 1981).

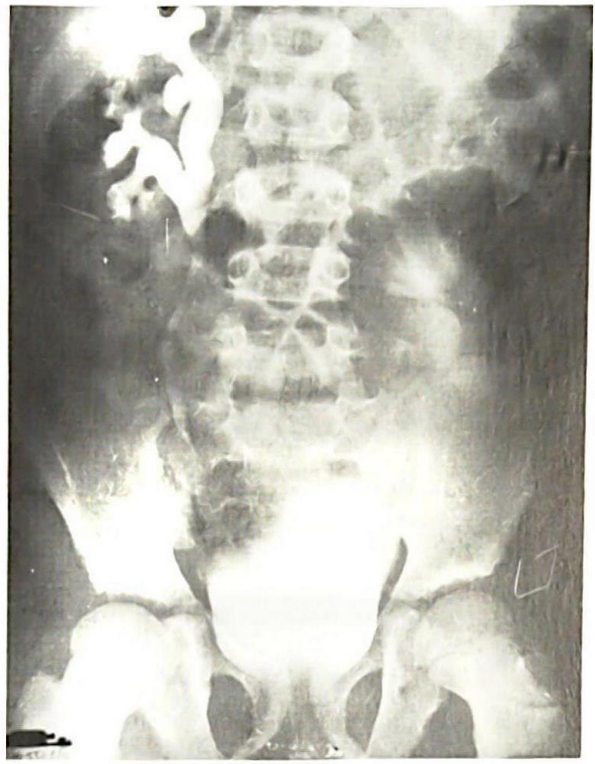

Fig. 1. Intravenous urogram showing gross hydronephrosis in a left ectopic iliac kidney. Clinically unsuspected.

In the human embryo at the 5-mm stage, i.e. about 5 weeks, the ureteric buds begin to appear from the Wolffian duct, and proceed dorso-cephalad into the metanephric masses. Apposition of the developing paired metanephric masses from whatever cause, within the foetal pelvis may produce fusion. The normal position of the kidney is reached at the 25–30-mm stage, i.e. about the end of the second month of development. Therefore, it may be assumed that any factors arresting the cephalad migration of the ureteric buds will result in an ectopic kidney. Clinically undefined and unspecified, these probable aetiological factors are vascular obstructions, faulty ureteric buds, pelvic anomalies developing at that stage, defective metanephros, genetic defects or maternal illness in early pregnancy.

Thoracic renal ectopia on the other hand has been attributed to delay in the transformation of the mesonephros (Malter & Stanley, 1972), which results in the continuation of the ascent beyond the eighth embryonic week.

It is noteworthy that when the case histories of all the patients were reviewed, there was nothing in the record to suggest to the attending doctor, either pre-operatively or before excretory urography, that renal ectopia was present. From this review three facts emerge about renal ectopia, which may be highlighted to the clinician.

The first is the vagueness of present symptoms. The clinical symptoms and findings related to renal ectopia do not have special characteristics. Therefore, a high index of suspicion is needed in its diagnosis.

In about 37% of the present series, the diagnosis was made incidentally at urography for other conditions such as carcinoma of cervix, hypertension or prostatic enlargement. Thirty-four per cent of the patients presented with symptoms pertaining to the urinary tract

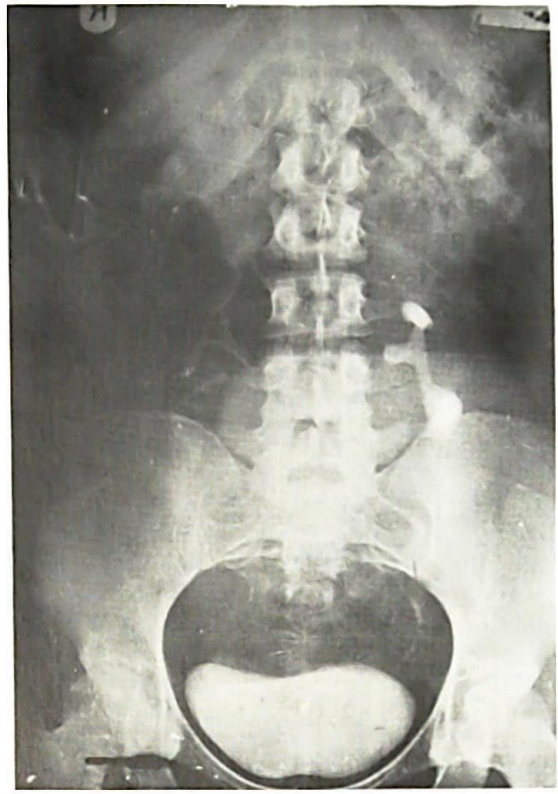

Fig. 2. Post-partum intravenous urogram revealed that the 'pelvic tumour' palpated during antenatal care is indeed an ectopic fused iliac kidney.

such as haematuria, enuresis, urgency and frequency. These were due to coincidental urinary tract disease, and cannot be assumed specific for renal ectopia.

In 43% of the series the presentation was with a palpable lower abdominal mass with vague abdominal or flank pain and/or gastrointestinal symptoms. The differential diagnoses in these patients included an appendix mass, overian tumour or cysts, omental cyst, large bowel tumour, intussusception or pedunculated fibroid.

Three children actually underwent needless laparotomy for either suspected intussusception or appendix mass that turned out to be a low ectopic kidney.

It may, therefore, be inferred that the finding of a lower abdominal mass of questionable aetiology with vague abdominal symptoms must raise the suspicion of an ectopic kidney with subsequent performance of an excretory urogram for confirmation. Kyrayianris, Stenos and Deliveoliotis (1973), after reviewing sixty-one patients with renal ectopia over a 15-year period, also reached a similar conclusion.

The second fact noted in this series is that the position of the ectopic kidney usually does not interfere with pregnancy and labour. Provided that there is no obstruction to the presenting part engaging in the pelvis, no difficulty should arise. Four patients were actually pregnant with an associated palpable 'pelvic tumour' shown subsequently at ultrasonography and post-partum urography to be ectopic kidney. They experienced no difficulty in pregnancy and labour. Another ten of the nineteen adult females in this study claimed to have experienced no difficulties in their many pregnancies and deliveries.

These observations are similar to that of

Thompson and Pace (1937) who reported a series of pregnancies in the presence of ectopic pelvic kidney. Out of fourteen women who had one or more children, none experienced any difficulties in labour, and they concluded that pregnancy will generally be uneventful and labour normal in spite of the presence of an ectopic pelvic kidney.

The third fact is the presence of co-existent congenital anomalies such as imperforate anus, renal malrotation, posterior urethral valves, prune belly syndrome etc. The incidence of associated malformations of the urinary tract and other organ systems is not as high as in other reported series of Malek, Kelalis and Burke (1971) and Kelalis, Malek and Segura (1973). Nevertheless, these findings are in general agreement with observations that renal ectopia, with or without fusion, appears to be only one facet of the malformative process involving many organ systems. Thus, the presence of any such congenital anomalies of the kidney is an indication for their thorough investigation and accurate definition. Conversely, when various anomalies are recognized, renal ectopia or fusion should be excluded by proper urological evaluation.

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(*Accepted* 16 *April* 1986)