Patología *1996; 29: 269-271*

Thoracic renal ectopia with diaphragmatic eventration.

A.W. BATES.

Department of Morbid Anatomy. The London Hospital Medical College and St Andrew's Hospital.London

SUMMARY

An intrathoracic ectopic kidney was an incidental finding at autopsy in an 85 year old woman with bilateral diaphragmatic eventration. The anatomical findings, including a normally positioned adrenal gland and abnormally long ureter, were indicative of a congenital lesion. The patient's normal renal function and normal post-mortem renal histology show that this rare form of ectopia had no functional consequences. The hypothesis that the normal diaphragm limits the craniad movement of the developing kidney was based on the association between thoracic ectopic kidney and diaphragmatic defects, however this case, in which the contralateral kidney was normally sited despite a diaphragmatic defect, suggests that the establishment of normal renal position does not depend on diaphragmatic position alone.

Key words: Diaphragmatic eventration. Urogenital abnormalities.

INTRODUCTION

Intrathoracic kidney is the rarest form of renal ectopia; a series of 15.919 paediatric autopsies included 22 cases of renal ectopia of which only one was intrathoracic (1). Thoracic kidney is usually asymptomatic and most of the approximately 150 cases in the literature were diagnosed by intravenous pyelography or ultrasound following the incidental finding of a mass on chest X-ray (2). A small proportion of cases which proceeded to thoracotomy have provided more detailed information on the anatomical relations of the kidney. Thoracic renal ectopia has been classified into four types: 1. True thoracic ectopia with a normally developed diaphragm. 2. Eventration of the diaphragm. 3. Diaphragmatic (Bochdalek's) hernia and 4. Traumatic rupture of the diaphragm (3). Here we report a case of previously undiagnosed thoracic kidney with bilateral diaphragmatic eventrations discovered at autopsy.

Correspondence: A.W. Bates. Department of Morbid Anatomy. The London Hospital Medical College. Whitechapel, London E1 1BB

CASE REPORT

An 85 rear old woman was admitted to hospital with an exacerbation of chronic airways disease. There was no history of urological symptoms. Laboratory investigations were within normal limits and a chest X-ray showed no abnormality of the lung fields, though bilateral diaphragmatic eventrations were noted. She responded well to treatment but after three days suffered a cardiac arrest and at autopsy death was found to be due to occlusive coronary arterial atheroma.

The right-sided diaphragmatic eventration contained the right kidney and peri-nephric fat; the kidney was related to the ninth to eleventh ribs posteriorly, the hilus facing infero-medially. The left sided diaphragmatic eventration contained fat only (fig. 1). Both renal arteries arose normally from the aorta at the level of the upper border of the twelfth thoracic vertebra. The renal artery, vein and ureter on the right side passed through a 3 cm diaphragmatic opening in their normal antero-posterior sequence; the kidney in situ within the eventration was not mobile. Both adrenal glands were morphologically normal and in their normal anatomical positions. The kidneys showed no macroscopic or histological abnormality, and no evidence of ischaemia.

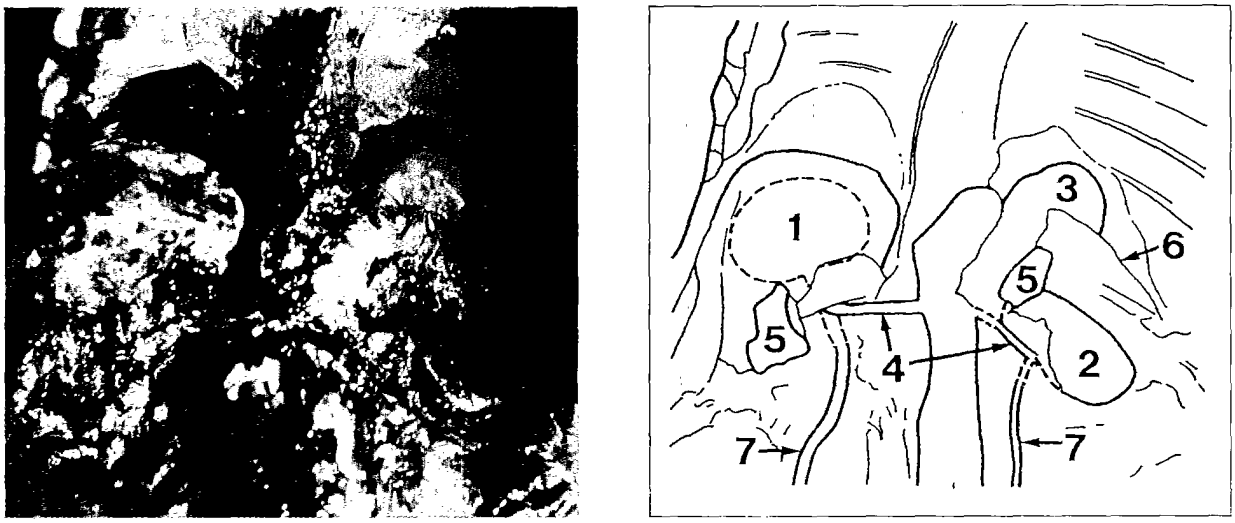

Figure 1.- Urinary tract and diaphragmatic eventrations in situ. 1) Intrathoracic kidney within diaphragmatic eventration. 2) Left kidney. 3) Diaphragmatic eventration. 4) Renal arteries. 5) Adrenal glands. 6) Cut edge of diaphragm. 7) Ureters.

The ureters and other abdominal organs were normal.

DISCUSSION

This case is, to our knowledge, the first report of thoracic kidney with diaphragmatic eventration at autopsy in an adult; three previous paediatric autopsy reports described thoracic kidney with diaphragmatic hernia (3,4) and eventration (5). Thoracic renal ectopia was initially assumed always to be congenital, but descriptions of two cases of apparently acquired thoracic kidney following diaphragmatic hernia due to trauma (6,7) raised the possibility of secondary renal displacement in adults, though in neither of these cases was it demonstrated that the thoracic kidney had once been normally sited. Diaphragmatic eventration may be congenital or acquired (8) and a Japanese radiographic study showed an incidence which increased with age to almost 1% in women over 60 (9), though as the authors suggest this unusually high incidence may be related to dietary or dressing habits in the population studied. In the present case therefore, owing to the age of the patient and the absence of previous radiographs, an acquired lesion cannot be entirely excluded, though the immobility of the kidney, the smallness of the diaphragmatic opening which admits the renal vessels, the presence of a normally formed liver and the abnormally long ureter all suggest a congenital abnormality. It is considered unlikely that the short renal artery would permit migration of the kidney to a thoracic position while preserving normal function (10), and in the present case no histological evidence of ischaemia was found.

The case does not satisfy all of the diagnostic creteria laid down for congenital thoracic kidney by Hawass et al (11), which are: 1. A rotation anormaly. 2. A long ureter. 3. Abnormally high renal vascular origins and 4. Medial deviation of the lower renal pole, however other authors accept the presence of a renal artery which arises normally from the aorta in congenital renal ectopia. One might expect an anomalous arterial supply if thoracic ectopia were due to abnormally cephalad development of the definitive kidney from the nephrogenic mesoderm, though not if the formed kidney became displaced during intra-uterine life, and cases of thoracic kidney with normal vascular origins have been described in very young children (5) and as part of a complex set of malformations (12), both of which suggest a congenital origin. Furthermore, there seems no reason to exclude cases such as the present one, in which the lower renal pole deviates laterally, from the congenital category.

It has been proposed that in thoracic renal ectopia with diaphragmatic eventration the diaphragmatic defect is attributable to the effect of the abnormally placed kidney on diaphragmatic development (13), though clearly most cases of eventration do not arise in this manner. An alternative theory is that a diaphragmatic defect fails to restrain the cephalad migration of the kidney (14). The low incidence of thoracic kidney with Bochdalek's hernia has been used as an argument against this hypothesis (2), and if diaphragmatic defects do predispose to thoracic kidney other factors must also be involved; the greater prevalence of thoracic kidney on the left side, and the association between right sided thoracic kidney and agenesis of the right lobe of the liver (15) seem to indicate a role for the liver in restricting abnormal renal movement. The present case indicates that longstanding thoracic kidney in diaphragmatic eventration may be both functionally and histologically normal.

ACKNOWLEDGEMENT

The author thanks Dr. S. I.Baithun for helpful discussions.

RESUMEN

Se comunica un caso de una paciente de 85 años con eventración diafragmática bilateral, en cuya autopsia se encontró incidentalmente un riñón ectópico intratorácico. Los hallazgos autópsicos, incluyendo una glándula adrenal de localización habitual y un uréter anormalmente largo, fueron indicativos de lesión congénita. La función e histología renal normales indicaron que esta rara forma de ectopia no tuvo consecuencias funcionales. La hipótesis que mantiene que los límites normales del diafragma limitan el movimiento craneal del riñón en desarrollo, estaba basada en la asociación entre riñones ectópicos torácicos y defectos diafragmáticos; sin embargo, este caso, en el que el riñón contralateral estaba en su posición habitual a pesar de la existencia de un defecto diafragmático, sugiere que el establecimiento de la localización normal del riñòn no depende únicamente de la posición diafragmática.

Palabras clave: Eventración diafragmática. Anomalías urogenitales.

BIBLIOGRAFIA

1. Campbell MF. Renal ectopy. J Urol 1930; 24: 187-198.
2. Donat SM. Donat PE. Intrathoracic kidney: a case report with a review of the world literature. J Urol 1988; 140: 131-133.
3. Pfister-Goedeke L. Brunier E. Die intrathorakale niere im kindesalter. Helv Paediat Acta 1979; 34: 345-357.
4. Campbell M. Clinical Pediatric Urology. Philadelphia, WB Saunders 1951; p 195.
5. N'Guessen G. Stephens FD, Pick J. Congenital superior ectopic (thoracic) kidney. Urol 1984; 24: 219-228.
6. Barrett NR. Right retroperitoneal diaphragmatic hernia. Br J Surg 1945; 32: 421-425.
7. Williams RG. Tillinghurst AJ. Diaphragmatic herniation of the kidney. Radiol 1949; 53: 566-568.
8. Dohler von R. Heinemann G. Angeborene zwerchfell-eventration. Fortschritte der Medizin 1979; 97: 1767-1830.
9. Okuda K. Nomura F, Kawai M, Arimizu N, Okuda H. Age related gross changes of the liver and right diaphragm, with special reference to partial eventration. Br J Radiol 1979; 52: 870-875.
10. Hill JE. Bunts RC. Thoracic kidney: case reports. J Urol 1960; 84: 460-462.
11. Hawass ND. Kolawole TM. El Badawi MG, Patel PJ, Malabarey T. Intrathoracic kidneys: report of 6 cases and a review of the literature. Eur Urol 1988; 14: 83-87.
12. Fusonie D. Molnar W. Anomalous pulmonary venous return, pulmonary sequestration. bronchial atresia, aplastic right upper lobe, pericardial defect and intrathoracic kidney. Am J Roentgen 1966; 97: 350-354.
13. Fleischner FG. Robins SA, Abrams M. High renal ectopia and congenital diaphragmatic hernia. Radiol 1950: 55: 24-26.
14. Spillane RJ. Prather GC. Right diaphragmatic eventration with renal displacement: case report. J Urol 1952: 68: 804-806.
15. Moschopoulos C. Bailly JM. Bruninx G, Delcour C. Agenesie du lobe droit du foie. A propos d'un cas. Annales de Radiologie 1993; 36: 323-327.