UNILATERAL POLYCYSTIC KIDNEY WITH RENAL HYPOPLASIA*

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Unilateral polycystic disease of the kidney is rare as is indicated by the paucity of reports in the literature. It is unilateral in only 3% of cases.[1] In a large series reported by Bell[2] only 2 cases were diagnosed between the ages of 1 month and 25 years. The youngest patient in a report of 85 cases of polycystic kidney by Walters and Braasch was 21 months old.[3]

CASE REPORT

An African mother of 3 children was admitted to the labour ward as an unbooked case on 3 January 1966. General examination was normal. On abdominal examination the uterine fundus was consistent with a pregnancy at term. There was no evidence of hydramnios. She was in advanced labour. A normal vaginal delivery of a healthy male infant weighing 7 lb. 7 oz. ensued. Routine examination of the infant after birth showed abdominal distension due to a partly solid mass measuring about 12 cm. across. The mass was limited in mobility and was bulging into the left flank. The surface was not clearly defined, but appeared lobulated and was resonant. No other abnormality was detected. The diagnosis of polycystic kidney or Wilms's tumour was considered.

Investigations, including full haematological examination, serum chemistry, blood urea, urinalysis and radiological chest examination, were within normal limits. An intravenous pyelogram showed complete absence of excretion of dye on the left side; the right kidney excreted normally. No abnormality was noted in the calyceal system of the right kidney. The pelvis, right ureter and bladder shadows were normal.

On 15 January 1966 laparotomy was performed through a left paramedian incision. A lobulated, tensely cystic mass was present in the position of the left kidney. The left ureter appeared as a thin, solid cord. The right renal system, liver, spleen and the pancreas appeared normal. Transperitoneal nephrectomy of the abnormal organ was performed.

Recovery was uneventful and mother and baby were discharged on the 9th postoperative day. When last seen at follow-up examination 21 months after operation, the infant was progressing very satisfactorily.

Pathologist's Report

A reniform mass measuring 10 × 8 × 7 cm. showed multiple cysts containing clear fluid. There was a complete absence of normal architecture. Sections showed a few infantile glomeruli surrounded by normal and cystic tubules of varying sizes and shapes. No communication between the cysts and renal pelvis was detected in any of the sections examined. The blood-vessels were normal. The features were consistent with the diagnosis of congenital polycystic kidney.

DISCUSSION

The pathogenesis of polycystic disease of the kidney remains obscure. Hildebrandt (quoted by Aird)[1] postulated a failure of the collecting tubules, which develop from the Wolffian duct, to join the secreting tubules which originate in the metanephros. Although this is the most acceptable theory today, it does not explain the association with similar cysts in other organs.[1]

It is now generally believed that the disease, as it occurs in adults, is different from that encountered in infants. The adult form is transmitted as a Mendelian dominant, while the infantile form is transmitted as a recessive trait. Tubular cysts in adults communicate with the renal pelvis; those in infants rarely do so.[4,5] The connective tissue in infants may appear embryonic.[4] The cysts are lined by flattened to cuboidal epithelium and contain clear or brownish fluid with urate, oxalate, cholesterol, sodium, potassium, chloride, urea, albumin and a few red blood cells.[3] The infantile type may be associated with other congenital abnormalities, such as hydrocephalus, spina bifida, cardiac malformations and, rarely, cysts in the lungs, spleen and pancreas.

Clinically, the infantile type of disease is more common in females. Depending on the size of the kidneys, obstructed labour may result, or the mass may be discovered after birth. According to Cooke,[6] 25% of children are stillborn or die soon after birth, 4% die in the first year and another 1½% succumb in the first 20 years. Renal rickets may develop in the last group.

Intravenous pyelography may demonstrate the typical 'spider pelvis', or show a non-functioning kidney as in our case.

In the differential diagnosis Wilms's tumour, hydronephrosis and a solitary renal cyst must be considered.

When the condition is bilateral it is not amenable to surgery. Walters and Braasch[3] believe that 'adequate visualization of the pelvis of but one kidney can be regarded as sufficient evidence of normal renal function to warrant removal of the other kidney'.

SUMMARY

A case of unilateral polycystic disease of the kidney, diagnosed at birth, is reported.

The clinical and pathological features are reviewed. Nephrectomy is considered the treatment of choice in the case presented.

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REFERENCES

1. Aird, I. (1958): *A Companion in Surgical Studies*, 2nd ed., p. 1124. Edinburgh: E. & S. Livingstone.
2. Bell, E. T. (1946): *Renal Disease*, p. 85. Philadelphia: Lea & Febiger.
3. Walters, W. and Braasch, W. F. (1933): Surg. Gynec. Obstet., **57**, 647.
4. Ackerman, L. V. (1964): *Surgical Pathology*, 3rd ed., p. 588. St Louis: C. V. Mosby.
5. Nelson, W. E. (1964): *Textbook of Pediatrics*, 8th ed., p. 1100. Philadelphia: W. B. Saunders.
6. Cooke, L. (1936): Lancet, **1**, 312.

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