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Case Report

Polycystic horseshoe kidney

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ARTICLE INFO

Article history:

Received 29 July 2012

Accepted 11 December 2013

Available online 12 March 2014

Keywords:

Polycystic kidney

Horseshoe kidney

Fetal autopsy

Introduction

Polycystic kidney disease (PKD) is a hereditary disorder of autosomal dominant or recessive inheritance. Horseshoe kidney is a common renal fusion anomaly during embryogenesis. Combination of both PKD and horseshoe kidney is rare, with an incidence of one in 1,34,000–8,00,000 live births.¹ Less than twenty cases have been reported till date in adults and only one in the fetus.^{2,3} We report the autopsy findings of this rare anomaly in a fetus with brief review of literature.

Case report

A 22-year-old female patient G2, P2, L1 married to her maternal uncle, had a normal antenatal course until 27 weeks of gestation. She presented to obstetric department with history of

loss of fetal movements for two days. Her past medical history was unremarkable. Routine blood and biochemical investigations were within normal limits. Antenatal Ultra Sonographic Scan (USG) revealed a single fetus with severe oligohydramnios and bilateral polycystic kidneys. Other family members were found to be normotensive and they did not give consent for their abdominal sonographic screening. Medical termination of pregnancy was performed with informed consent, and the fetus was sent for pathological evaluation.

At autopsy examination, it was a male fetus weighing 1600 g. On external examination there was low set ears and depressed nasal bridge (Potter's facies). On internal examination all organs were in-situ. The kidney was enlarged and fused at the lower pole measuring $9 \times 8 \times 1.2$ cm and revealed multiple cysts of varying sizes, largest measured 1.5×1.5 cm (Fig. 1). Microscopic examination of kidney revealed cysts of varying sizes lined by flattened epithelium which were surrounded by thick peritubular mesenchyme. Absence of immature cartilage within it ruled out dysplasia (Fig. 2A and B). Ureter and bladder were unremarkable. Microscopic sections from the liver showed preserved architecture with multiple small cysts lined by flattened epithelium along with mild periportal fibrosis (Fig. 2C and D). All other visceral organs were unremarkable. Umbilical cord showed three vessels. With the above findings a pathological diagnosis of polycystic horseshoe kidney (Potter's type III) was offered.

Discussion

Horseshoe kidney is the commonest renal fusion anomaly with an incidence of 1 in 400 persons. The abnormality occurs

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<http://dx.doi.org/10.1016/j.mjafi.2013.12.002>

Fig. 1 – A: Gross photograph of fetus with opened abdomen showing polycystic horseshoe kidney. B: Gross photograph of cut section of polycystic horseshoe kidney showing cysts of different sizes.

between 4th and 6th weeks of gestation. It is less commonly associated with cystic diseases. The cystic changes commonly associated are polycystic kidney disease (PKD), multicystic renal dysplasia (MCRD) and glomerulocystic diseases (GCD).^{4–8} A combination of horseshoe kidney and PKD is extremely rare.² Campbell reported two cases of polycystic disease in an autopsy series of 122 cases of horseshoe kidney. Rathbun on reviewing the literature prior to 1924, found three cases of polycystic kidney amongst 108 cases of horseshoe kidney.⁴ Boatmen et al noted one polycystic kidney in a review of 96 horseshoe kidney patients.⁵ Dalgaard noted one case of horseshoe kidney in his study of 284 patients with polycystic kidneys.⁶

Coexistence of horseshoe kidney and MCRD is rare. Less than twenty cases have been reported till date.⁷ PKD is usually bilateral or involves both the components of horseshoe kidney but the lower urinary tract is unaffected; in contrast MCRD is unilateral or involves only one component of horseshoe kidney and is associated with some form of lower urinary tract obstruction.⁸ GCD is an uncommon type of cystic disease; the condition may be familial, non-familial or associated with other heritable form of cystic disease like ADPKD or ARPKD. The involved kidney grossly appears sponge like, hypoplastic or normal in size. Microscopically tissue shows dilatation of Bowman's space with absence of tubular dilatation.⁹ There are reports of isolated GCD involving one component of horseshoe kidney.¹⁰

Fig. 2 – A&B: Microphotograph of kidney showing loss of architecture and cysts of varying sizes lined by simple epithelium with thick peritubular mesenchyme (H&E, ×40). C: Microphotograph of liver showing cyst lined by simple epithelium (H&E, ×10). D: Microphotograph of liver showing mild periportal fibrosis and cyst lined by simple epithelium (MTS, ×10).

USG can diagnose cystic conditions involving kidney but sometimes underlying horseshoe kidney is missed as observed in our case. The classical USG findings in PKD are enlarged homogeneous hyper echogenic kidneys with absence of corticomedullary differentiation and difficulty in identifying the structure of urinary bladder.² The ultrasonographic criteria for the diagnosis of MCRD includes identification of enlarged kidney with grape like clusters of non-communicating cysts, absence of accumulation of urine in urinary bladder and severe oligohydramnios. There may be associated non renal anomalies like dilatation of bowel loops. A dimercaptosuccinic acid scan reveals absence of function in the affected segment. GCD also shows multiple cysts in kidney but with lower urinary tract obstruction.⁹

Horseshoe kidney is an incidental renal anomaly and mostly asymptomatic condition.³ Neonates with PKDs surviving beyond first month usually have hypertension or renal insufficiency.² The development of renal failure associated with PKD is not greater in the presence of a horseshoe kidney.¹ Indication for nephrectomy for polycystic horseshoe kidney include: persistent pyelonephritis, uncontrollable hypertension and in preparation for renal transplantation.¹ Cystic diseases of the kidney pose diagnostic challenges to the radiologist and sometimes necessitates autopsy for final confirmation.

Conflicts of interest

All authors have none to declare.

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