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Case Report

The Novel Occurrence of Non-Syndromic Unilateral Renal Hypoplasia and Unilateral Aplasia in two Brothers

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Abstract:

Background: Non-syndromic unilateral renal hypoplasia is a congenital condition associated with one small kidney (hypoplastic) that generally has normal residual parenchyma but smaller calyces. We have previously reported the first case of non-syndromic unilateral renal hypoplasia transmitted in an autosomal dominant fashion from the mother.

Patients and methods: The brother of the boy who was the patient in the medical literature reported to have unilateral renal hypoplasia transmitted from the mother in an autosomal dominant was seen and renal ultrasound performed.

Results: The present patient, the brother was seen late during June, 2022 at about the age of eighteen years, for a routine check and was asymptomatic and otherwise healthy. However, abdominal ultrasound showed unilateral right renal aplasia. The right kidney was absent, while left kidney showed compensatory hypertrophy.

Conclusion: This paper reports the novel occurrence of non-syndromic unilateral renal hypoplasia and unilateral aplasia in two brothers.

Key words: autosomal dominant; non-syndromic; unilateral renal hypoplasia; unilateral renal aplasia iraq

Introduction

Non-syndromic unilateral renal hypoplasia is a rare congenital condition associated with one small (hypoplastic) kidney that generally has normal residual parenchyma but smaller calyces. The size of the hypoplastic kidney is less than two standard deviations below the expected mean. The condition can be asymptomatic or presents with urinary tract infections and/or renal urolithiasis. Unilateral renal hypoplasia is typically diagnosed with renal ultrasound.

We have previously reported the first case of non-syndromic unilateral renal hypoplasia transmitted in an autosomal dominant fashion from the mother [1].

Patients and methods

The brother of the boy who was the patient in the medical literature reported to have unilateral renal hypoplasia transmitted from the mother in an autosomal dominant was seen and renal ultrasound performed.

Results

The present patient, the brother was seen late during June, 2022 at about the age of eighteen years, for a routine check and was asymptomatic and otherwise healthy. However, abdominal ultrasound showed unilateral right renal aplasia. The right kidney was absent. The left kidney showed compensatory hypertrophy with dimensions of 10 x 5 cm, cortex 14 mm with homogeneous texture. The cortico-medullary junction was intact with no mass or stone. There was no dilatation of the pelvi-calyceal system or the left ureter. There was no other abnormality on abdominal ultrasound.

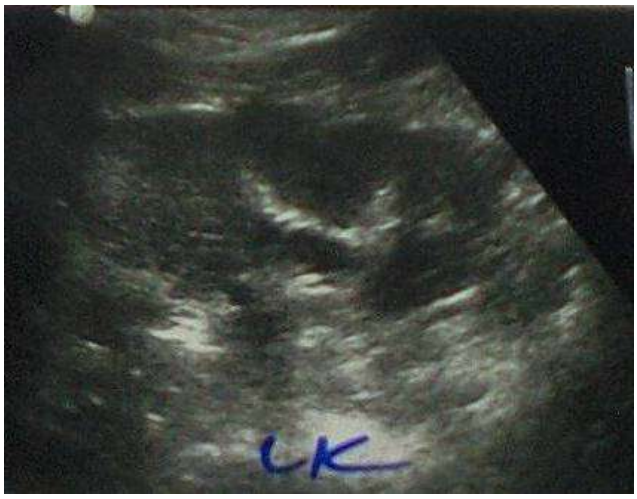

Figure-1: The left kidney

Discussion

The occurrence of non-syndromic unilateral renal hypoplasia has been reported early during the 1900s [1]. Taño Pino et al (1996) described two brothers, not twins who had unilateral congenital renal hypoplasia with vesico-urethral reflux. The diagnosis in the two brothers was made at about the age of 24 years and without any urological history, when they presented with a dysuria-pollakiuria syndrome [3].

Cain et al (2010) emphasized a genetic basis for unilateral renal hypoplasia [4], but the transmission of the condition from a parent to offspring has never been reported. Autosomal dominant non-syndromic unilateral renal hypoplasia has not been reported in Iraq or in any other country [3-9].

The occurrence of non-syndromic isolated unilateral renal aplasia in two siblings has been reported once during the early 1960s.

Conclusion

This paper reports the novel occurrence of non-syndromic unilateral renal hypoplasia and unilateral aplasia in two brothers.

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