

A Case of Acrorenal Syndrome

Mehdi Esmaeili Jadidi,¹ Farshid Oliaei^{2,3,4}

¹Student Research Committee,
Babol University of Medical
Science, Babol, Iran

²Cellular and Molecular Biology
Research Center; Cancer
Research Center; Health
Research Institute, Babol
University of Medical Sciences,
Babol, Iran

³Clinical Research
Development Center, Shahid
Beheshti Hospital, Babol
University of Medical Sciences,
Babol, Iran

⁴Department of Internal
Medicine, School of Medicine,
Babol University of Medical
Sciences, Babol, Iran

Urinary tract anomalies are common and comprise about 20% to 30% of total congenital anomalies. This spectrum consists of many different anomalies of the urinary tract that may be syndromic or nonsyndromic with different etiologies. In this case report, a patient with single kidney and urinary tract signs is introduced that was diagnosed accidentally. The finding of different anomalies in different organ systems should lead us to examination of the intactness of the urinary tract. In these disorders, if there is no need for immediate intervention, long-term follow-up can be helpful to postpone chronic kidney disease progression.

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INTRODUCTION

Urinary tract anomalies are common and comprise about 20% to 30% of total congenital anomalies.¹ This spectrum consists of unilateral or bilateral renal agenesis, hypoplasia, dysplasia, multicystic kidneys, horseshoe or duplex kidneys, vesicoureteral reflux, hydroureter, hydronephrosis, and ureteropelvic or ureterovesical junction obstruction.^{2,3}

Congenital renal anomalies can be sporadic, familial, syndromic (with disorders in other organs), or nonsyndromic. The etiology can be environmental (nutrition and drugs) or genetic.^{4,5} In this case report, a single-kidney patient with urinary tract signs is introduced that was diagnosed with a syndrome.

CASE REPORT

A 29-year-old man was referred to hospital with severe left flank pain and reduced urine output. On physical examination, in addition to left costovertebral angle tenderness, loss of right thumb was prominent as a congenital anomaly

(Figure). Abdominal ultrasonography represented a single left kidney with moderate hydronephrosis and a small lower calyceal calculus.

The patient was hydrated and then he passed a calculus that was lost. The history of urinary calculus was positive in his father. He was azotemic that recovered completely after calculus passage. The patient was unaware about his single kidney and there was no similar history in his family. Repeated history taking revealed right undescended testis that had been surgically corrected in childhood. In order to examine other anomalies associated with acrorenal syndrome, vesicocystoureterography was done that was negative. He was discharged with a recommendation for follow up of the left kidney function.

DISCUSSION

Acrorenal syndrome is part of many different syndromes occurring with congenital anomalies of the kidney and urinary tract, and it can be due to chromosomal, teratogenic, or dysplastic disorders.⁶ In this syndrome, limb disorders can be unilateral or



Radiographic image of the right hand shows oligodactyly (no thumb).

bilateral; upper or lower; polydactyly, oligodactyly, syndactyly, brachydactyly, or ectrodactyly (loss of a part or whole of a finger like this case); and shortness of radial bone or forearm. This problem can be seen in a wide range of disorders of oromandibular area, trachea, lungs, skin and appendages, uterus, testes and vas deferens, heart, eyes, and spine. Simultaneous formation of the urinary tract and other systems in 4 to 12 weeks of embryonic period is the main reason of these syndromes.⁷

It seems that many acrorenal syndromes are transmitted as autosomal recessive inheritance, but the reported case did not match with syndromes like Dieker,^{8,9} Johnson-Munson,¹⁰ or Siegler.¹¹ In these syndromes, the probable gene (Formin gene) is located at 15q13-14.¹¹ In reported case, due to no similar patient in his family, it must be

a sporadic mutation.

Unilateral renal agenesis occurs in 1 in 1500 to 3200 live births. This is more common in men and more prevalent in the left side.¹² Regardless of morbidities from other nonurinary system involvement and their impact on the quality of life, unilateral renal agenesis alone with making hyperfiltration, glomerulosclerosis, and chronic kidney disease can be impressive on life.

The finding of different anomalies in different organ systems should lead us to examination of the intactness of the urinary tract. In these disorders, if there is no need for immediate intervention, long-term follow-up can be helpful to postpone chronic kidney disease progression.

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CONFLICT OF INTEREST

None declared.

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Correspondence to:
Farshid Oliaei, MD
Department of Internal Medicine, School of Medicine, Babol
University of Medical Sciences, Babol, Iran
Tel: 011-32254899
Fax: 011- 32251664
E-mail: ol_1964@yahoo.com

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