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Case report

# Polycystic Horseshoe Kidney

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## Abstract

Horseshoe kidney is a renal fusion anomaly during embryogenesis with incidence ranges of 1 in 400 to 1 in 1800 live births. Adult polycystic kidney disease is a hereditary disorder and transmitted in autosomal dominant pattern with a reported incidence of 1 in 1000 to 1 in 5000 cases. Polycystic horseshoe kidney is a very rare occurrence with incidence ranges of 1 in 134 000 to 1 in 8 000 000 live births. We here present a patient who was admitted with headache, hypertension and polycystic horseshoe kidney.

**Keywords:** Horseshoe Kidney; Polycystic Kidney Disease

## Introduction

Horseshoe kidney is a fusion anomaly occurring during embryogenesis in which the kidneys are conjuncted in the lower poles of the medium line with and incidence between 1/400 and 1800 [1]. Adult type autosomal dominant polycystic kidney disease (ADPKD) is a hereditary disorder developing due to polycystin 1 (ADPKD1) ve polycystin 2 (ADPKD2) with a reported incidence of 1/ 1000-5000 [2]. Polycystic horseshoe kidney anomaly is a very rare disorder with an incidence between 1/134 000 and 1/ 8 000 000 [3]. Polycystic horseshoe kidney is considered as two seperate diseases.

## Case

A 54 years old female patient presented to our hospital with complaint of headache. From her history, we learnt that she had been treated for hypertension for 2 years. We found that our patient had gone to the controls irregularly and used angiotensin receptor blocker + hydrochlorothiazide group antihypertensive medicine. On her familial history, there was history of hypertension in her mother and there was not a history of kidney disease in her family. Her blood pressure was 180/100 mm Hg without any additional pathology. On the laboratory examinations, creatinine value was found as 0,8 mg/dL. Full blood count and the other biochemical serum values were in the normal ranges. Microscopic hematuria was found in her urine examination. On the abdominal ultrasonography, it was found that the size of both kidneys increased, there were numerous number of cysts in the liver and kidneys and lower poles of the kidneys were conjuncted. Furthermore, the patient underwent computed abdominal tomography. On her computed tomography; enlarged kidneys, numerous kidney cysts with various sizes and horseshoe kidney anomalies (Figure 1) and multiple liver cysts (Figure 2) were reported. Her mother and siblings were screened with kidney ultrasonography for ADPKD, but no polycystic kidney disease was found.

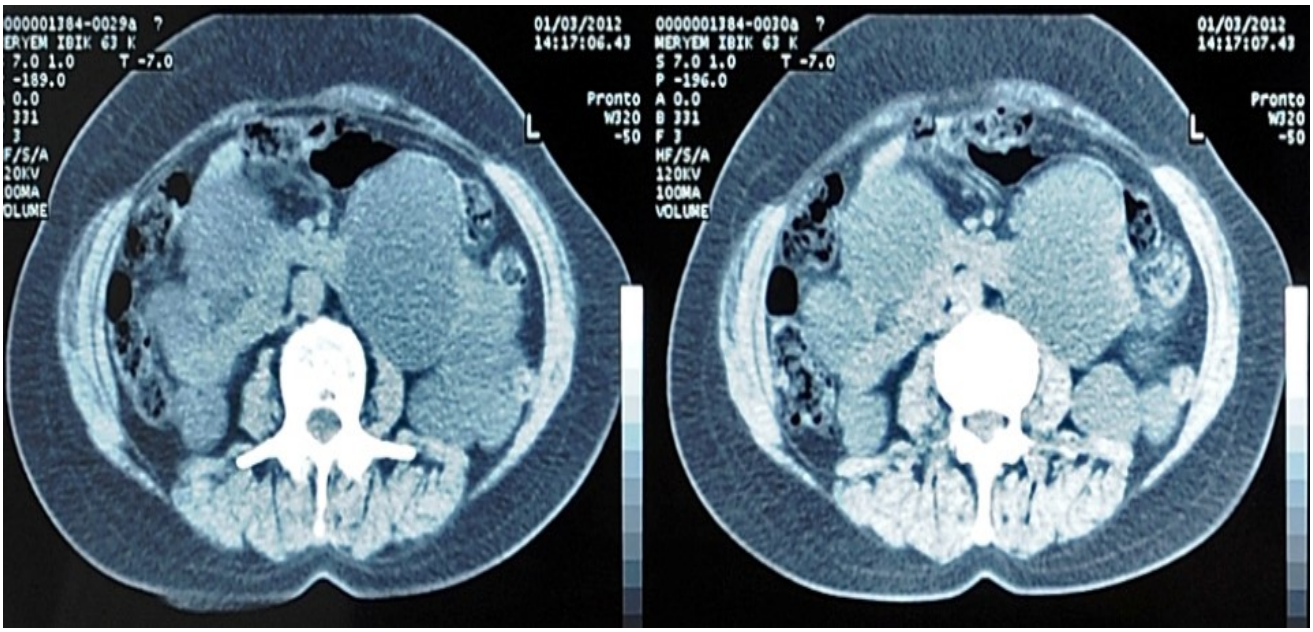


**Figure 1.** Multiple cysts in various sizes and horseshoe anomaly in both the kidneys are seen on computed abdominal toımography.

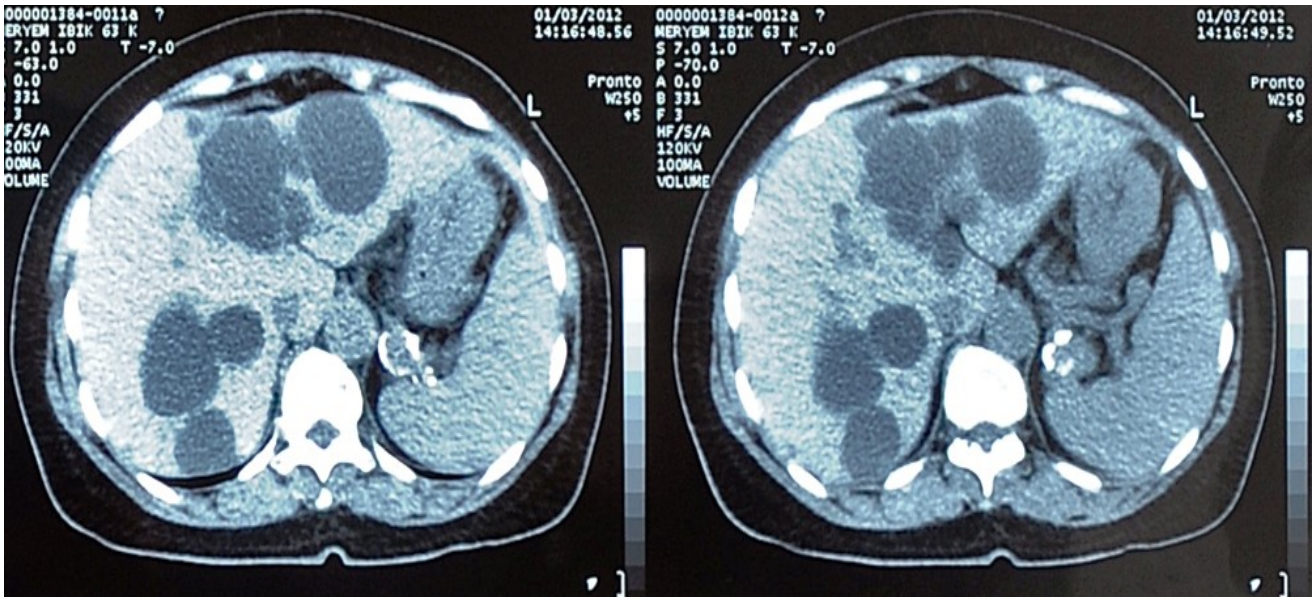


**Figure 2.** Multiple cysts in various sizes in the liver are seen on computed abdominal toımography (polycystic liver).

## Discussion

Horseshoe kidney which occurs in the lower poles of the kidneys during embryogenesis is probably the most common form of the renal fusion anomalies. Its incidence has increased in the cases with Turner syndrome and in men [4]. Although, horseshoe kidney cases are asymptomatic, yet they can be seen with stone disease, ureteropelvic junction syndrome and renovascular hypertension [5-8].

Adult polycystic kidney disease is characterized with diffuse cystic lesions in the kidneys and other viscera. Mutation in the PKD1 gene which is located in the 16th chromosome and PKD2 gene which is located in the 4th chromosome, coding polycystin-1 and polycystin-2 in the kidneys and other tissues play a role in the pathogenesis [9-11]. Since PKD1 and PKD2 genes may be found also in the non-renal tissues, polycystic kidney disease must be considered as a systemic disorder. In adult polycystic disease, systemic involvement may be seen such as diffuse cyctic lesions in the liver, colonic diverticula, valvular heart disorders and cerebral artery aneurysmas [12-15]. There were diffuse hepatic cysts in our patient.Adult polycystic kidney disease may affect all the ethnic groups. Of ADPKD

cases, 5% are seen without a familial history [3]. This probably reflects the new mutation causing to a new cohort likewise our case.So far no any genetic association was described between PKD gene loci and horseshoe kidney anomaly.

We could not find data in the literature about clinical progression of polycystic horseshoe kidney. Diagnosis for polycystic horeshoe kidney is often established incidentally with ultrasonography during the routine screening or screening of the persons having familial ADPKD history.

Polycystic horseshoe kidney is, extremely rare with about 20 cases reported in the literature. Polycystic horseshoe kidney presented by findings of the combination of two distinct renal disorders, loss of kidney function and end stage renal failure [16]. Occasionally polycystic horseshoe kidney presented by such as subjuctive findings increased belly volume, abdominal pain, headache and hematuria. In the patients with polycystic horseshoe kidney disease, treatment is in the form of individual therapy especially for hypertension and serious complications including pain, hematuria, infection, nephrolithiasis, diverticulosis, and arterial aneurysms. Isthmus undivided bilateral nephrectomy has been reported in polycystic horseshoe kidney patients [16].

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