Horseshoe Kidney Complicated by Xanthogranulomatous Pyelonephritis in a Young Girl: A Case Report and Review of the Literature

Jamal Musayev[1,*], Rashad Sholan[2], Adalat Hasanov[1], Rizvan Rustamov[3]

ABSTRACT

The cases of horseshoe kidney presented by xanthogranulomatous pyelonephritis are very rare. In this study, the case of XGP developing in HSK in a young female patient was presented due to its rare incidence and the previously reported cases were reviewed, as well. The patient, who has end-stage renal disease and was under treatment, admitted to the clinic for preemptive kidney transplantation. Bilateral open en bloc nephrectomy was performed before the kidney transplantation. The histopathological examination of the specimen was reported as XGP. Eight months later, living-donor organ transplantation was performed to the patient with the kidney obtained from her father. XGP can present as a complication of HSK. Moreover, HSK may rarely be manifested by end-stage renal disease in young patients. In such cases, who would undergo kidney transplantation, it is important to examine the HSK in detail and perform bilateral nephrectomy to prevent complications after transplantation.

KEYWORDS

horseshoe kidney; xanthogranulomatous pyelonephritis; bilateral nephrectomy; end-stage renal disease

AUTHOR AFFILIATIONS

[1] Department of Pathology, Azerbaijan Medical University, Baku, Azerbaijan
[2] Department of Kidney Diseases and Transplantology, Republican Treatment and Diagnostic Center, Baku, Azerbaijan
[3] Institut of Pathology, SRH Wald Klinikum, Gera, Germany
* Corresponding author: Sherifzade 212, Patoloji Anatomiya Burosu, AZ1012, Baku, Azerbaijan; e-mail: patolog.jamalmusaev@gmail.com

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INTRODUCTION

Horseshoe kidney (HSK) is the most common renal fusion anomaly with a prevalence of 0.25%; and is twice more common in the male compared to the female (1, 2). Approximately 1/3 of the cases with such anomalies are asymptomatic and they are detected randomly during autopsies or through the imaging methods (1, 2). The remaining cases occur as complications such as obstruction, infection, urolithiasis in the ureteropelvic junction, and rarely through secondary findings due to tumor and trauma (2).

Xanthogranulomatous pyelonephritis (XGP) is a rare disease causing renal parenchymal destruction; and it constitutes 0.6 to 1.0% of the cases of chronic pyelonephritis. It can be seen at all ages; however, it is more frequent in middle-aged women (3, 4).

The cases of HSK presented by XGP are very rare. In this study, the case of XGP developing in HSK in a young female patient was presented due to its rare incidence and the previously reported cases were reviewed, as well.

CASE REPORT

A 17-year old female patient, who has end-stage renal disease and was under treatment, admitted to the clinic for preemptive kidney transplantation with complaint of chronic fatigue. The partially palpable abdominal mass was determined on physical examination. The daily amount of urine was around 1 liter; and the protein amount in the 24-hour urine was 4000 mg. According to the laboratory tests, WBC was 10.3, creatinine was 4.5 mg/dL, GFR was 18 ml/min/1.73 m^2, hemoglobin was 8.5 gr/dL, and hematocrit was 27%. There were signs of pyuria in the urine and *E. Coli* reproduction in the urine culture. Native computed tomography (CT) revealed enlargement in both renal pelvis and calyces, numerous formations compatible with stone in their cavity, signs of atrophy in the renal parenchyma, and isthmus combining both kidneys in their lower poles (Figure 1). Considering the findings mentioned above, the patient was recommended bilateral open en bloc nephrectomy to prevent complications after the kidney transplantation.

For the nephrectomy, the abdominal cavity was opened with a section starting with supraumbilical incision and extending laterally with a Mercedes-type incision. The colon was medialized by incising the parietal peritoneum from the white line of Toldt, and the retroperitoneum was reached. Surrounding tissues were removed from both kidneys, and the arteries and veins were ligated. Both kidneys were removed as en bloc, together with the isthmus connecting the two kidneys. The amount of blood loss during surgery was 200 mg. The creatinine level was 7–9 mg/dL in the postoperative period. The patient received hemodialysis 3 times a week following the bilateral nephrectomy. Eight months later, living-donor organ transplantation was performed to the patient with the kidney obtained from her father. No complications and rejection have been observed in the patient within the seven years of follow-up.

In the macroscopic examination of the specimen, fusion was detected in the lower pole of the horseshoe kidney (Figure 2). There was dilatation in the renal pelvis and calyces. Signs of atrophy were observed in the renal parenchyma. Abundant purulent exudate and stones were detected in the renal pelvis. In the microscopic examination, renal tissue was atrophic. There were neutrophil infiltration areas and microabscess in the background of diffuse lymphocyte and plasma cell infiltration. Lipid-laden macrophage accumulations were observed in certain areas (Figure 3). The histopathological examination of the specimen was reported as XGP.

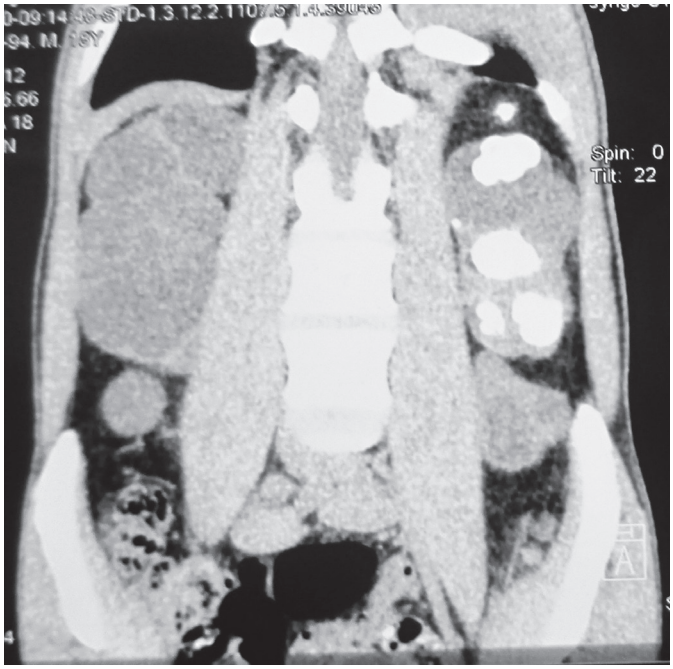

Fig. 1 CT scan of the abdomen revealed features of the bilateral hydronephrosis and pelvic urolithiasis.

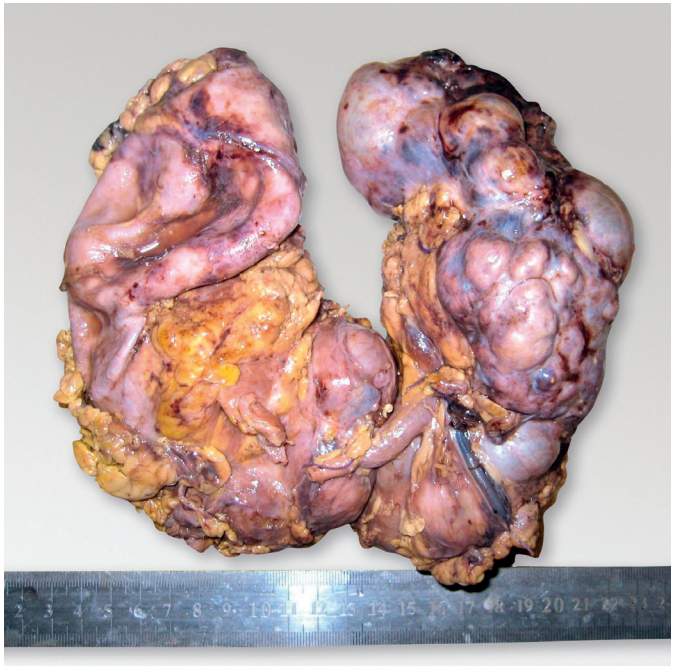

Fig. 2 Macroscopic view of the nephrectomy specimen.

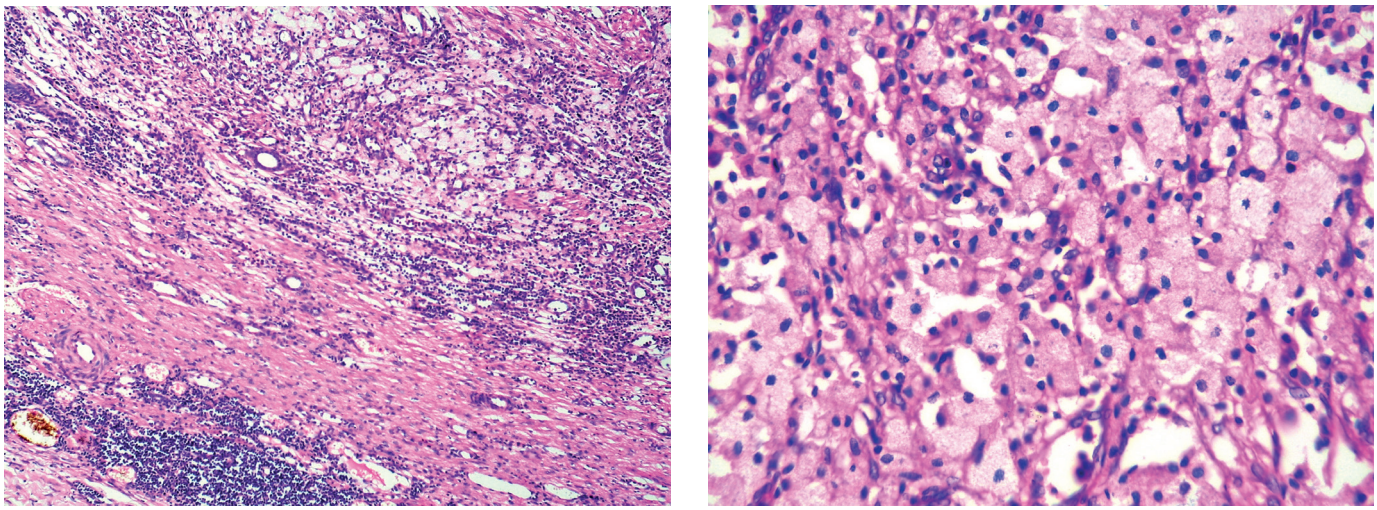

Fig. 3 Diffuse infiltration of atrophic renal parenchyma by lymphocytes, plasma cells and neutrophils (a), and aggregates of foamy macrophages (b) (×100, ×200; HE).

DISCUSSION

The combination of HSK and XGP is extremely rare and the first case was reported in 1994 by Hammadeh et al. (5). There have been only 7 cases reported in the English literature so far (5–11). The detailed characterization of all cases is presented in Table 1. What makes our case different from the previously reported cases is the concomitant end-stage renal disease, implementation of bilateral nephrectomy and subsequent kidney transplantation.

Congenital anomalies of the kidney and the urinary tract have been detected more frequently and earlier, particularly with the widespread of imaging methods within the last 30 years. Some of these anomalies, which form a wide spectrum, can be identified within the first years of life and in the antenatal period; however, asymptomatic anomalies such as unilateral renal agenesis, ectopia, malrotation, and fusion anomalies manifest themselves in older ages and adults (1, 12).

HSK is the most common fusion anomaly. It has been reported in the literature that the incidence rate in newborns is 1/400–1600 (1, 2). In the vast majority of the cases, fusion is present in the lower pole of the kidneys; therefore, U-shaped HSK is more common. Reverse U-shaped HSK, which results from the fusion of the upper pole, and L-shaped HSK, which is shaped due to lateral fusion, are rarely observed (2). In our case, fusion was in the lower pole and U-shaped HSC was present.

Approximately half of the complications related to HSK consists of obstructions and kidney stones (1, 2, 13).

Tab. 1 The general characteristics of horseshoe kidney cases complicated by xanthogranulomatous pyelonephritis.

Authors	Age (years)	Gender	Symptoms	Imaging method	Presence of stones	Involvement	Treatment
Hammadeh et al. (5), 1994	7	Male	Right flank mass	X-ray	Yes	Right kidney	Right heminephrectomy
Samuel et al. (6), 2001	6≥	NK	Pain, fever, hematuria, renal mass	X-ray, US, CT	Yes	Left kidney	Left heminephrectomy
Sausville et al. (7), 2009	48	Female	Fever	CT	Yes	Left kidney	Laparoscopic left hemine-phrectomy
Mongha et al. (8), 2010	62	Male	Pain, fever	US, CT	Yes	Left kidney	Left heminephrectomy
Basson and Witt (9), 2013	18≤	NK	NK	X-ray, CT	Yes	Right kidney	NK
Sawazaki et al. (10), 2017	75	Male	General fatigue, breathlessness	CT	No	Right kidney	Right heminephrectomy
Fernandez et al. (11), 2018	64	Female	Pain, decreased appetite, abdominal discomfort, chronic fatigue	CT	Yes	Bilateral	Laparoscopic left hemine-phrectomy
Musayev et al. Present case	17	Female	Chronic fatigue	CT	Yes	Bilateral	Bilateral nephrectomy

NK: Not known, US: Ultrasonography, CT: Computed tomography

Among the cases, 4% can manifest with malignancies and the most common malignancy accompanying HSK is the renal cell carcinoma (2, 13). Urinary tract infections are present in 19% of the cases; however, the cases of HSK complicated with XGP are very rare (13).

XGP was defined for the first time by Schlagenhaufer in 1916 (3, 4). The reasons for its occurrence are not definite; however, it is common in cases with long-term obstruction and infection. The presence of stones in the pelvis was reported in 47–100% of the cases (4). Obstruction findings and stones were present in both moiety of the kidney in our case.

The preoperative diagnosis process of XGP is quite complicated. In some of the cases, specific diagnosis can be determined by CT. However, the fact that there are not stable clinical and radiological findings of XGP as well as it can resemble tuberculosis, pyelonephritis, perinephric abscesses, and malignant tumors in the imaging methods, are important factors making the specific diagnosis difficult (3, 4). In such cases, determining the specific diagnosis and ruling out the above mentioned entities requiring different management is possible only with histopathological examination.

As can be seen from the table 1, the majority of cases of XGP developing in HSK are adults; only 2 pediatric cases have been reported (5, 6). XGP is more common in the female, while HSK is more common in the male. Our case was a 17-year-old female.

The most frequent imaging method used in the diagnostic process of the cases was CT, which was also used in our case. CT is valuable in these cases in terms of displaying the kidney stones, the involvement of perinephric adipose tissue characterizing XGP, and the anatomic findings specific to HSK (4, 9).

The incidence of urinary stones is high in the cases of both XGP and HSK. Urinary stone has been reported in the vast majority of cases with HSK manifesting with XGP, including our case. Only one case, who did not have a urinary stone, had a history of nephrolithotomy that was performed 34 years ago (10).

The risk of end-stage renal disease in HSK is higher than that of the general population. However, it has not been reported in the cases with the combination of HSK and XGP before our case (13). Six of the cases had unilateral and one case had bilateral involvement. Unilateral heminephrectomy was performed in the 6 cases where the treatment method was specified (5–8, 10, 11). Mortality was reported in only one case (10). Our case is the first case of HSK bilaterally complicated with XGP in the literature that underwent bilateral nephrectomy and was performed kidney transplantation subsequently.

CONCLUSION

Although rare, XGP can present as a complication of HSK. Moreover, HSK may rarely be manifested by end-stage renal disease in young patients. In such cases, who would undergo kidney transplantation, it is important to examine the HSK in detail and perform bilateral nephrectomy to prevent complications after transplantation.

ABBREVIATIONS

HSK	Horseshoe kidney
XGP	Xanthogranulomatous pyelonephritis
US	Ultrasonography
CT	Computed tomography
WBC	White blood cells

PRIOR PUBLICATION

This study was presented as an poster presentation at the 1st International Uroanatomy Congress, Izmir-Turkey, on June 14–16, 2013.

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