Nephron sparing surgical intervention as an effective method of treatment of bilateral xanthogranulomatous pyelonephritis in a 6-year-old boy

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KEY WORDS

xanthogranulomatous pyelonephritis ◗ decapsulation ◗ heminephrectomy ◗ xanthoma cells

ABSTRACT

Xanthogranulomatous pyelonephritis is an extremely rare form of tubulo-interstitial nephropathy, characterized by the progressive destruction of the renal parenchyma, which is then replaced by the xanthogranulomatous tissue. This process is usually present in one kidney and in the perirenal fat tissue and its clinical course imitates a carcinoma. Nephrectomy of an affected organ is often the only treatment.

We would like to present a case of a 6-year-old boy with bilateral xanthogranulomatous pyelonephritis, in whom a sparing surgery, namely decapsulation of a swollen, infiltrated kidney with heminephrectomy of its inferior pole, combined with broad-spectrum antibiotic therapy, allowed saving the child's life and retaining active renal tissue. The diagnosis was confirmed by numerous xanthoma cells found in a microscope examination of resected tissues.

CASE REPORT

Chronic xanthogranulomatous pyelonephritis (XGP) is a clinical and morphological form of chronic tubulo-interstitial nephropathy, which is extremely rare in children. XGP is characterized by progressive destruction of the renal parenchyma, which is then replaced by the xanthogranulomatous tissue. Its etiopathogenesis is unknown, its clinical image is non-characteristic and the pathomorphological image of inflammatory infiltration is a decisive factor for its diagnosis. The inflammatory infiltration is present in the region of the renal pelvis and interstitial tissue; frequently it passes the renal capsule and expands to the perirenal fat tissue and other structures of the retroperitoneal space. The presence of xanthoma (foam) cells, which are lipid-laden macrophages, as well as of blood-originated cells, lymphocytes and neutrophils is a pathognomic feature of the inflammatory infiltration. The foci of hyalinization, calcification and microthrombi are formed inside the infiltrate [1, 2, 3].

We are presenting a case of a 6-year-old boy with bilateral xanthogranulomatous pyelonephritis, in whom combined surgical and pharmacological treatment was successful.

A 6-year-old boy (W.S.) was transferred to the Ward of Nephrology from a regional hospital with suspected urosepsis. A few days earlier the child had a high fever and experienced vomiting and strong abdominal pain. History: since infancy the child had experienced recurring urinary tract infections and therefore imaging diagnostics of the urinary tract were performed. A bilateral double calyceal-pelvic system was diagnosed based on ultrasound examination and urography. Cystography excluded the presence of vesicoureteral outflow. A routine ultrasound examination in March 2007 revealed normal renal structure: the left kidney of 7.3 cm and the right kidney of 8.4 cm. Within 3 months preceding the hospitalization the following facts were observed: recurrent infections of the upper respiratory tract, episodes of high fever of unknown etiology, general weakness, and body weight loss.

At admission to the Department of Nephrology the patient's condition was moderately severe, he was apathetic and suffering. The physical examination revealed dehydration, severe abdominal bloating, and general abdominal tenderness, lack of peristalsis and positive peritoneal symptoms. In the palpation examination a suspicious mass in the left middle abdomen was recognized, which suggested a tumor inside the abdominal cavity. Laboratory tests revealed very high levels of inflammatory markers (CRP – 372 mg/l), increased leukocytosis with predominant granulocytes (L – 18.53 G/L), moderate urine leukocyturia and albuminuria, hemostasis disturbances (APTT – 46.2 s; prothrombin index – 55.6%; D – dimers 7.5 mg/l, ATIII 40%; Fibrinogen – 4.75 g/l) and electrolyte disturbances (Na – 126 mmol/l; K – 3.5 mmol/l). A urine culture revealed the growth of *Pseudomonas aeruginosa* at the concentration of 10^3, while blood cultures were negative. An ultrasound examination revealed enlarged kidney dimensions, especially in the case of the left kidney, and its perfusion was disturbed (RK – 10.7 cm; LK – 12.2 cm), and features of intraparenchymal edema in both kidneys. Due to increasing peritoneal symptoms and worsening of the patient's clinical condition, the decision was made to perform an exploratory laparotomy. However, during the surgical procedure no defined pathological condition was observed in the abdominal cavity that could explain the existing clinical picture. For that reason, intensive symptomatic treatment was continued. Treatment included broad-spectrum antibiotic therapy (Tazocin, Ciprofloxacin), immunoglobulin infusions, compensation of water-electrolyte disturbances and red blood cell concentrate infusions. During treatment the boy's clinical condition improved gradually; however, the symptoms of a "tumor" inside the abdominal cavity persisted, as well as a subfever and high values of inflammatory markers. An abdominal CT examination revealed massive infiltrating-inflammatory lesions in both kidneys, predominant in the left kidney and a hypodense region in the parenchyma suggesting the formation of multiple abscesses (Fig. 1, 2).

The lesions described above were accompanied by infiltrative-inflammatory lesions in the fascia and perirenal fat tissue.

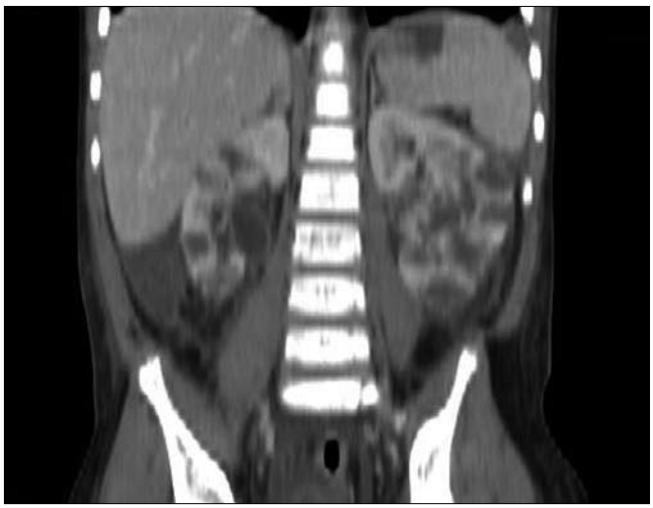

Fig. 1. CT scan reveals significant enlargement of both kidneys, especially of the left kidney and multiple hypodense areas in the median part of the left kidney and its bottom pole.

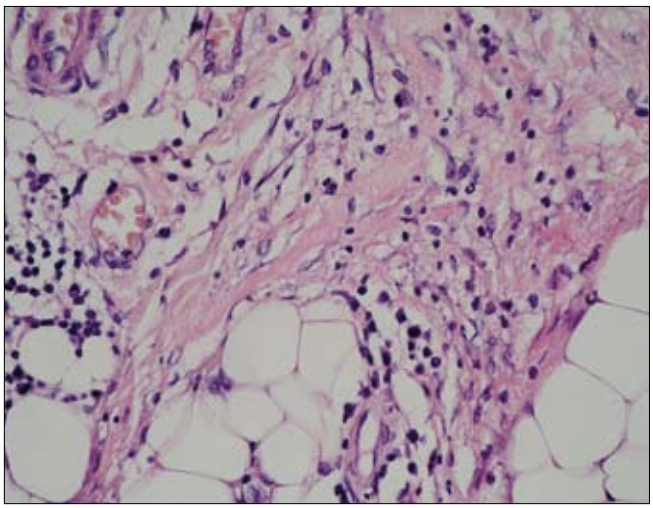

Fig. 3. Part of the fibros – membranous capsule with inflammatory lesions that spread into the perirenal fat tissue. Many xanthomatous cells are visible.

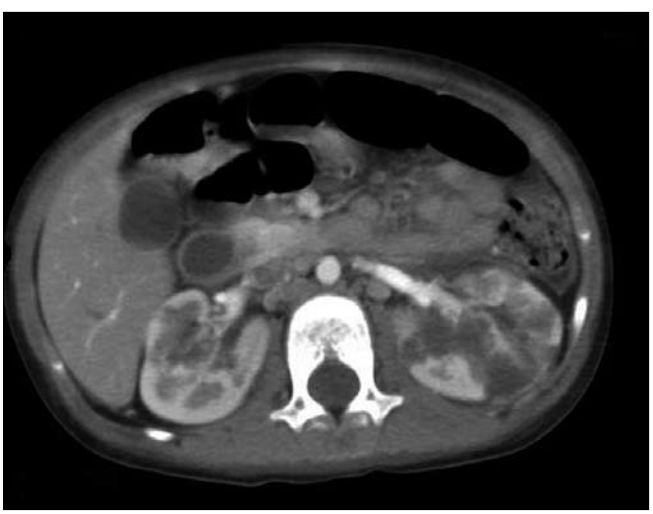

Fig. 2. CT scan showing bilateral solid hypodense renal masses, which in the left kidney form a "bear paw" sign. Thickening of the Gerota fascia and perirenal inflammatory changes are visible.

Fig. 4. A scintigraphic image – areas of impaired radioisotope uptake especially in the bottom poles of both kidneys.

Moreover, an exudate was observed in the pleural and peritoneal cavities. Based on a CT scan the suspicion of granulomatous nephritis was made. Despite intensive pharmacotherapy the boy's clinical condition did not improve as expected. Abdominal pain and fever persisted, body weight loss increased and weakness also persisted. Increasing sterile pyuria and anemisation were observed in the laboratory tests. Daily diuresis and the values of the arterial pressure were normal. A follow-up CT scan revealed lack of progression of inflammatory-destructive lesions in the kidneys and perirenal tissues, and the density of inflammatory lesions, especially in the bottom pole of the left kidney, increased. In addition to that, thickening of the renal pelvic wall and bottom ureter in the right kidney were observed. Exudates in the body cavities regressed. Based on the findings described above the decision was made to perform a surgical procedure. Decapsulation of the left kidney and resection of its bottom pole with an abscess were performed. In the postoperative course we observed quick improvement of the clinical condition and the values of inflammatory markers returned to normal (CRP – 4.5 mg/l; L – 7 G/L). The histopathological assessment of the resected kidney revealed numerous foam cells and therefore it was possible to diagnose xanthogranulomatous pyelonephritis (Fig. 3.) Follow-up imaging tests (abdominal CT and ultrasound) revealed gradual evolution of lesions described above, namely kidney dimensions decreased, the pelvic-calyceal systems in both kidneys widened discreatly and inflammatory-post-inflammatory lesions were formed in the renal cortex. Static scintigraphy performed after 3 months confirmed that excretory functions of both kidneys were maintained and small post-inflammatory foci were present in the parenchyma (Fig. 4). The laboratory markers of renal functions were within the norm (creatinine – 0.7 mg/dl).

DISCUSSION

The issues related to xanthogranulomatosis as a rare form of tubulo-interstitial nephritis were discussed in detail in a recently published review [1]. The evidence for a rare incidence of XPG is a low number (slightly above 200) of cases of this form of nephropathy, which have been published in medical literature [2, 4]. According to our knowledge, there is no case of bilateral XPG in a child in Polish literature, therefore we have decided to present this patient in detail. The factors predisposing for XPG include urinary tract infections, which are chronic and recurrent, hindered urinary outflow especially due to nephrolithiasis, congenital anomalies of the urinary system (subpelvic stricture, vesicoureteral outflow and especially double kidneys and ureters), ineffective treatment for urosepsis, chronic hypoxia of the renal parenchyma with subsequent focal impairment of tissue metabolism, lymph circulation disturbances, and congenital or acquired immune defects favoring the development of infections, especially with gram-negative bacteria [4, 5]. Double kidneys and ureters as well as recurrent urinary tract infections are of notice in this case. Although the clinical picture depends on the extensiveness of inflammatory lesions in the kidneys and surrounding tissues, the most frequent symptoms in-

clude fever, weakness, body weight loss, anemisation, symptoms of a "tumor" inside the abdominal cavity, and pain in the lumbar area [1, 2]. The differential diagnosis should include disease conditions associated with enlarged kidneys, especially neoplasms (Wilms' tumor, clear cell carcinoma, non-Hodgkin lymphoma, leukemia), hydronephrosis, pyonephrosis, fungal infections and other forms of chronic, granulomatous nephritis (megalocytic interstitial nephritis (MIN), malacoplakia) [1].

CONCLUSIONS

The result of a renal CT scan (a "bear paw" sign) (Fig. 2) and the detection of characteristic xanthoma cells in the inflammatory infiltrate in the kidneys and perirenal tissues or urine [1, 6] are extremely important to the diagnosis.

The recommended form of treatment for this nephropathy is nephrectomy, which is often life-saving. When the pathological process has affected both kidneys, it is necessary to choose organ-sparing techniques (decapsulation, abscess drainage, heminephrectomy) [1].

Intensive antibacterial and resuscitation treatment (mainly fluid therapy) combined with surgical treatment saved the patient's life and restored almost normal excretory functions of the kidneys (despite the loss of half of a kidney and a history of inflammatory infiltration in both kidneys).

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