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A MULTIDISCIPLINARY APPROACH TO THE CORRECTION OF UREMIC SYNDROME IN A PATIENT WITH KIDNEY ANGIOMYOLIPOMA

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Abstract

Patients with uremic syndrome are at high risk of developing various complications that can lead to fatal consequences. Uremic syndrome usually develops on the background of chronic kidney disease, especially its later stages. However, uremia may also occur as a result of tumor, surgical treatment of which can lead the patient to renoprival state. According to various authors, the development of uremic syndrome occurs in 7-10% of patients suffering from kidney tumors.

Renal angiomyolipoma is a benign mesenchymal tumor, which accounts for 3% of all tumors of renal parenchyma and 90% of all benign tumors of the kidney. It is known that angiomyolipoma is characterized by slow growth and the associated asymptomatic course.

Present study describes a case of uremic syndrome developed as a result of kidney angiomyolipoma in a 32-year-old female patient, who earlier underwent nephrectomy in connection with angiomyolipoma of the contralateral kidney. The computer tomography allowed to reveal a large retroperitoneal space-occupying lesion and to detect a spontaneous rupture of this formation, in connection with which the patient underwent a revision of the retroperitoneal space. While making the revision of the tumor no kidney authentic tissues were clearly detected. That was the reason for performing the nephrectomy. The patient has begun renal replacement therapy using haemodialysis. The mother of the patient expressed a desire to become a kidney donor for the daughter. As a result of examination in the potential donor a renal artery aneurism was revealed.

After examination of the donor and recipient, the patient has successfully undergone living-related kidney transplantation, and the kidney donor – renal artery plasty.

Thus, the specialized urological management, haemodialysis as a method of renal replacement therapy and the living-related kidney transplantation, concentrated in one center, have given the chance to provide maximal rehabilitation of the patient with the uremic syndrome as well as to prevent fatal complications in the kidney donor.

Keywords: *uremic syndrome, kidney angiomyolipoma, renal replacement therapy, living-related kidney transplantation.*

Introduction

Uremia is a clinical syndrome associated with fluid and electrolyte, metabolic and hormonal disorders developing on the background of decreased kidney function. Kidney failure leads to the development of hypertension, anemia, acidemia, hyperkalemia, hyperparathyroidism, protein-energy malnutrition [*Meyer T, Hostetter T, 2016*]. Uremic syndrome usually develops with the decrease in glomerular filtration rate of less than 10 *ml/min*. However, in some patients the above-mentioned symptom complex occurs in earlier stages of the kidney damage.

Currently a number of uremic toxins, resulted from metabolism (especially proteins) and accumulated in patients with impaired renal function, have been identified. In addition to the urea, ammonia,

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cyanate, creatinine, guanidine, uric acid, β2-microglobulin, β2-glycoprotein, peptides of average molecular mass, amino acid, pyridine derivatives, aliphatic and aromatic amines, polyamines, indole, phenols, myoinositol, mannitol, acetone, lipochromes, cyclic adenosine monophosphate, glucuronic and oxalic acids, some hormones, including parathyroid hormone, some enzymes and other substances are also accumulated [*Yavuz A et al., 2005*].

Uremic syndrome usually develops on the background of chronic kidney disease, especially its later stages. However, uremia may also occur as a result of tumor, surgical treatment of which can lead the patient to renoprival state.

Renal angiomyolipoma is a benign mesenchymal tumor, which accounts for 3% of all tumors of renal parenchyma and 90% of all benign tumors of the kidney [*Eble J, 1998; Matveev V et al., 2002*]. It is known that angiomyolipoma is characterized by slow growth and the associated asymptomatic course. Clinical management of patients with angiomyolipoma includes observation, embolization of the renal vessels, partial or complete removal of the kidney. Recommendations for treatment are usually based on the patient's symptoms or the injury size [*Blute M et al., 1988; Dickinson M et al., 1998*]. Although from a modern point of view while choosing a surgical treatment method for patients with angiomyolipoma the most protective method is organ-sparing approach, but in the presence of clinical manifestations of angiomyolipoma – large sizes or tumor growth, complications of the tumor, retroperitoneal or intrarenal hemorrhage, compression of the cup-pelvis-plating system, pain syndrome, renal hypertension, the nephrectomy is indicated [*Matveev V et al., 2002*]. Another group of indications for nephrectomy is associated with diagnosis uncertainty and difficulties of preoperative diagnosis [*Ellingson J et al., 2008*].

The prognosis for patients with uremia is unfavorable without using renal replacement therapy – dialysis or transplantation. Timeliness and completeness of treatment that requires a multidisciplinary approach are essential for the life safety of patients with uremic syndrome [*Santos A et al., 2014*].

We present our own observation of the uremic syndrome development in a patient with kidney angiomyolipoma.

Clinical observation

A 32-year-old female patient was admitted to Kharkiv Regional Clinical Center of Urology and Nephrology named after V. I. Shapoval with complaints of nausea, dry mouth, general weakness, abdominal pain (mainly on the right side) and decreased urine output. The complaints appeared 3 days ago.

It is known from the anamnesis that left nephrectomy, ovariectomy and splenectomy were performed in this patient with angiomyolipoma in 2012.

On admission, the general state of the patient was assessed as severe. The patient was exhausted, with a pale skin and puffy face. Body temperature was 37°C, tachycardia – up to 100 beats per minute. Blood pressure – 90/60 *mm Hg*. The abdomen is increased in volume, tense, sharp pain, positive Pasternatsky symptom. On palpation of the abdominal cavity a dense formation is determined. Daily diuresis is reduced to 100 *ml*.

The examination revealed anemia (red blood cells $2.8*10^{12°}/l$, hemoglobin 87 *g/l*), increased levels of urea up to 20.3 *mmol/l*, creatinine – up to 626,3 *mmol/l*, potassium – 5.6 *mmol/l*. In the clinical analysis of urine – red blood cells in the entire visual field, 15 to 25 cells in total amount.

During a CT scan in the right retroperitoneal space there was a multinodular formation of irregular shape having dimensions 187×141×261 *mm* with uneven outlines and heterogeneous structure due to dense fatty inclusions, septa, solid and liquid component, on the background of which the parenchyma and renal cavity system are not clearly differentiated. This formation was determined as angiomyoliposarcoma of the retroperitoneal space with invasion into a single right kidney.

Diagnosis: Retroperitoneal space-occupying lesion. Spontaneous rupture of the tumor. Hematoma of the retroperitoneal space.

The patient was offered a surgical intervention within the retroperitoneal space margins on the right with possible nephrectomy. Cystoscopically preoperative stenting was performed in connection with the predictable difficulty of visualization of ureter in the tumor tissues.

Multiple adhesions are suboperationally observed in the abdominal cavity. The retroperitoneal space-occupying lesion on the right occupies

2/3 of the abdominal cavity and pushes the abdominal organs to the left. Bowel loops are distended and soldered together. In the abdominal cavity a hemorrhagically colored ascitic liquid is detected; the total volume is up to 2.5 *l*. Gradually using blunt and sharp dissection, the isolation of the space-occupying lesion spread out from the subhepatic space pushing the liver upwards and to the pelvis is made.

The ureter containing the catheter is dislocated to the pelvis. The kidney tissue is not detected and differentiated from the tumor tissues. The renal artery and vein were also isolated. While isolating the renal vessels the artery was injured. Vascular clamps were applied, and the vascular defects were sutured with prolene 6/0 thread.

The ischemia time was 4 minutes. In order to visualize the renal tissue and determine the condition of the kidney it was decided to remove the tumor by fragments. The space-occupying lesion was isolated from surrounding tissues on the renal vessels. During fragmentary removal of the tumor, the renal cavity system was detected in several areas. While conducting thorough revision of the tumor tissues no authentic kidney tissues were clearly detected. In this connection it was unadvisable to preserve the kidney and it was decided to perform nephrectomy and removal of the tumor tissues. Fyodorov's forceps was applied on the vascular pedicle, the vessels were crossed. The removal of the tumor and kidney was performed.

The vessel pedicle was tied with a nylon ligature. In the tumor bed and retroperitoneal space a careful hemostasis was performed. During the removal of the tumor the adrenal gland was visualized, the latter was partially resected. The sanitation and drainage of the retroperitoneal space and abdominal cavity through counterpuncture on the anterior abdominal wall from two points was produced.

Immunohistochemical study diagnosed angiomyolipoma.

In postoperative period a replacement therapy was started using hemodialysis.

Later on the patient's mother expressed a desire to donate a kidney to her daughter. Standard examination of the potential donor was conducted. During computer tomography the aneurysm of the right renal artery up to 5 *mm* in diameter was revealed. Due to the risk of saccular aneurysm development, which could provoke the bleeding, the donor was indicated urgent surgical intervention – right nephrectomy with subsequent preservation of the kidney and renal artery plasty. As the recipient had no other potential donors, a decision was made about the right-side nephrectomy and plasty of the renal artery aneurysm with subsequent kidney transplantation of the daughter.

Under endotracheal anesthesia a lumbotomy was performed to the donor on the right side. The access to the right kidney was performed gradually layer-by-layer. After that the kidney with a vascular pedicle was isolated. During the revision a saccular aneurysm of the renal artery is defined. The kidney was deemed suitable for transplantation. The right nephrectomy in the donor was performed. The kidney was washed with a solution of 1000 *ml* Custodiol. While examining the renal artery after the aneurysm excision it was decided to carry out its plasty by a great saphenous vein fragment. The additional access in the top right third of the right thigh toward the great saphenous vein was produced; the latter was allocated over 4 *cm*, ligated, the fragment of the vein with a diameter of about 8 *mm* was resected. The plasty of renal artery with a fragment of saphenous vein was carried out. The kidney was washed well, with the vessels of the renal pedicle were tight.

The kidney transplantation was performed in the left iliac region of the patient. The end-to-end anastomosis of the renal artery with the internal iliac artery and end-to-side anastomosis vein with external iliac vein were performed. Urine was obtained immediately after incorporation of the transplant into the bloodstream.

Postoperative period was uneventful. The level of creatinine and urea was normalized on the third day after surgery. The donor and recipient were discharged on the 7th postoperative day in a satisfactory condition.

Conclusion

Among the reasons of development of chronic renal insufficiency urologic diseases, including kidney tumors occupy a significant place. In such patients renal failure develops as a renoprival condition due to removal of kidneys.

The above-described observation demonstrates

that in young patients the removal of single or both kidneys followed by the patient's renoprival condition and the medical management using renal replacement therapy (long-term hemodialysis) and subsequent kidney transplantation, currently recognized as the most effective treatment, is justified in the final stage of the renal failure.

Implementation of a multidisciplinary approach in a specialized centre made it possible to provide medical rehabilitation of the patient with severe kidney damage, as well as prevent life-threatening complications of a kidney donor.

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