CASE REPORTS
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INTRAOPERATIVE CELL SALVAGE IN THE TREATMENT OF WUNDERLICH SYNDROME CAUSED BY RENAL ANGIOMYOLIPOMA

ULOGA APARATA ZA INTRAOPERATIVNU AUTOTRANSFUZIJU U LEČENJU VUNDERLIHOVOG SINDROMA UZROKOVANOG BUBREŽNIM ANGIOMIOLIPOMOM

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Summary

Introduction. Wunderlich syndrome is defined as acute spontaneous hemorrhage into the subcapsular and perirenal spaces in the absence of trauma. The most common causes of Wunderlich syndrome are various renal neoplasms, polycystic and vascular kidney diseases. Wunderlich syndrome is common in patients who use anticoagulant therapy or suffer from various types of congenital coagulopathies. The clinical diagnosis is usually confirmed by computer tomography of the abdomen and requires urgent surgical treatment or renal artery embolization. **Case Report.** A 45-year-old woman was admitted to the Emergency Center due to acute pain in the left abdomen, associated with arterial hypotension. A computer tomography of the abdomen was performed using intravenous contrast, which showed a ruptured tumor of the left kidney with a massive retroperitoneal hematoma. The tumor was 7 cm in diameter and according to the computer tomography data it corresponded to an angiomyolipoma. The patient underwent emergency surgery. Intraoperative cell salvage was used with additional blood replacement. Nephrectomy and drainage of the massive retroperitoneal hematoma were performed. Pathohistological analysis of the surgically removed kidney confirmed a renal angiomyolipoma. **Conclusion.** Wunderlich syndrome caused by the rupture of angiomyolipoma is still a life-threatening condition that should be considered in the differential diagnosis of sudden abdominal pain, especially in women of reproductive age. In hypotensive patients, available radiological methods should be used to make an accurate diagnosis, before taking the patient to the operating room.

Key words: Angiomyolipoma; Kidney Neoplasms; Retroperitoneal Space; Hemorrhage; Signs and Symptoms; Diagnosis; Blood Transfusion, Autologous; Nephrectomy

Sažetak

Uvod. Vunderlihov sindrom je definisan kao akutno nastalo spontano krvarenje u supkapsularnom i perirenalnom prostoru u odsustvu traume. Najčešći uzroci Vunderlihovog sindroma su različiti tumori bubrega, policistična i vaskularne bolesti bubrega. Vunderlihov sindrom je češći kod pacijenata koji koriste antikoagulacionu terapiju ili boluju od različitih vrsta kongenitalnih koagulopatija. Klinički postavljena dijagnoza se obično potvrđuje komjuterizovanom tomografijom abdomena i zahteva hitno hirurško lečenje ili embolizaciju renalne arterije. **Prikaz slučaja.** Žena starosti 45 godina je primljena u Urgentni centar zbog akutnog bola u levoj polovini trbuha koji je praćen arterijskom hipotenzijom. Načinjena je kompjuterska tomografija abdomena sa intravenskim kontrastom koji je pokazao postojanje rupturiranog tumora levog bubrega sa masivnim retroperitonealnim hematomom. Tumor je bio prečnika 7 cm i prema tomografskim karakteristikama je odgovarao angiomiolipomu. Pacijentkinja je hitno operisana. Primenjen je aparat za intraoperativnu autotransfuziju uz dodatnu nadoknadu krvi. Urađena je nefrektomija i drenaža masivnog retroperitonealnog hematoma. Patohistološkom analizom hirurški odstranjenog bubrega potvrđeno je da se radi o angiomiolipomu bubrega. **Zaključak.** Vunderlihov sindrom uzrokovan rupturom angiomiolipoma i dalje predstavlja životno ugrožavajuće stanje koje treba uzeti u obzir u diferencijalnoj dijagnozi iznenadno nastalog abdominalnog bola, posebno kod žena u reproduktivnom periodu života. Kod hipotenzivnih pacijenata treba koristiti dostupne radiološke metode radi postavljanja tačne dijagnoze, pre uvođenja pacijenta u operacionu salu.

Ključne reči: angiomiolipom; tumori bubrega; retroperitonealni prostor; krvarenja; znaci i simptomi; dijagnoza; autologna transfuzija; nefrektomija

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Abbreviations
WS – Wunderlich syndrome
CT – computed tomography

Introduction

Wunderlich syndrome (WS) is defined as acute spontaneous hemorrhage into the subcapsular and perirenal kidney spaces in the absence of trauma. This syndrome was first described in 1856. The WS is characterized by Lenk's triad: acute flank or abdominal pain, a palpable flank mass, and hypovolemic shock [1]. Nontraumatic retroperitoneal hemorrhage is commonly caused by rupture of an abdominal aortic aneurysm, adrenal bleeding, or coagulopathy. The most common causes of WS are various renal neoplasms, especially angiomyolipoma and renal cell carcinoma, but also cystic and vascular kidney diseases. The WS is common in patients who use anticoagulant therapy or suffer from various types of congenital coagulopathies.

Clinically established diagnosis is usually confirmed by computed tomography (CT) of the abdomen and requires urgent surgical treatment or renal artery embolization. The choice of treatment depends on the severity of the clinical presentation as well as the cause itself. Surgical treatment includes total or partial nephrectomy. Total nephrectomy is the treatment of choice in cases when bleeding is caused by a spontaneous rupture of renal cell carcinoma or angiomyolipoma larger than 5 cm in diameter.

Case Report

A 45-year-old woman was admitted to the Emergency Center due to acute pain in the left hemiabdomen, associated with arterial hypotension. The patient was communicative during admission. Sudden pain in the left abdomen occurred about an hour before admission. She complained of weakness and dizziness and denied injuries to the body before the pain started. During the physical examination, there was no evidence of injuries. The patient had no chronic diseases. She underwent an umbilical hernia surgery two weeks before admission and had no other prior surgeries.

Abdominal CT was performed immediately using intravenous contrast. It showed a ruptured left kidney tumor with massive retroperitoneal hematoma. The largest diameter of the tumor was 7 cm and according to CT data it corresponded to an angiomyolipoma. Hematoma dimensions were 80 x 80 x 170 mm (anterior-posterior x latero-lateral x cranio-caudal), and density 50 - 60 Hounsfield units. The described hematoma was compressing the pancreas, stomach, small intestine and vascular pedicles of the kidney and spleen **(Figures 1, 2 and 3)**. The laboratory tests showed significant anemia. Due to hemodynamic instability, the patient underwent emergency surgery. Since the CT showed a renal angiomyolipoma and the patient lost a significant amount of blood, intraoperative cell salvage was per-

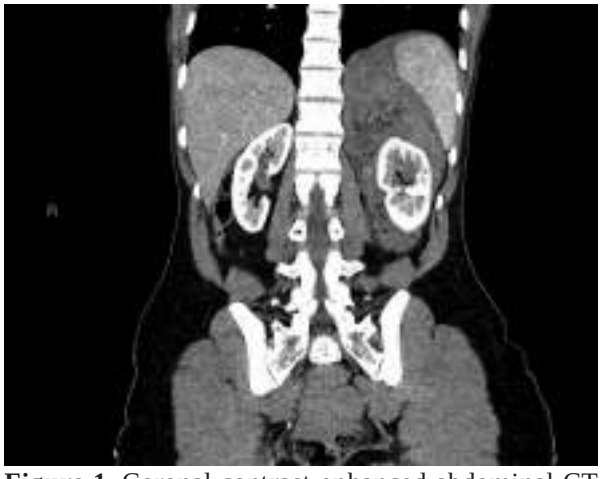

Figure 1. Coronal contrast-enhanced abdominal CT showing a massive left perirenal hematoma
Slika 1. *Koronalni kontrasni snimak kompjuterizovane tomografije abdomena pokazuje masivni levi perirenalni hematom*

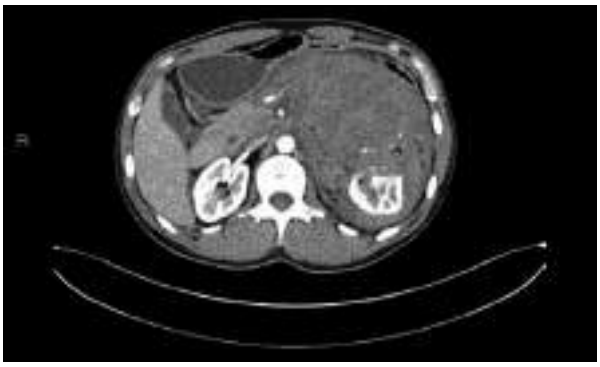

Figure 2. Abdominal CT – adipose tissue in the tumor indicates an angiomyolipoma
Slika 2. *Kompjuterizovana tomografija abdomena – adipozna komponenta tumora ukazuje na angiomiolipom*

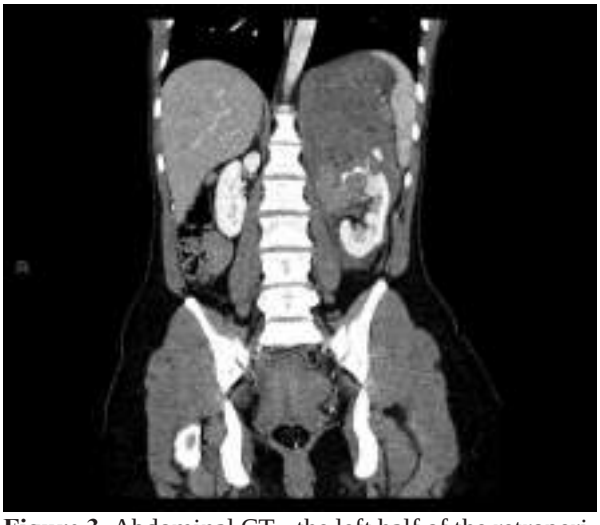

Figure 3. Abdominal CT - the left half of the retroperitoneum is filled with a massive hematoma spreading from the tumor mass in the upper pole of the left kidney
Slika 3. *Kompjuterizovana tomografija abdomena: leva polovina retroperitoneuma je ispunjena masivnim hematomom koji se pruža od gornjeg pola levog bubrega*

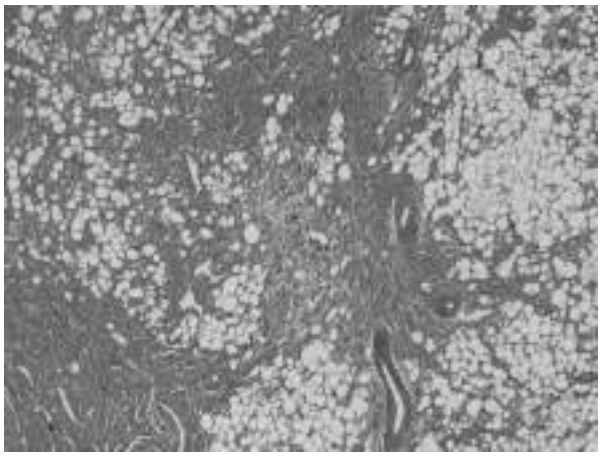

Figure 4. Angiomyolipoma (HE staining, 25x)
Slika 4. *Angiomiolipom HE bojenje, 25x*

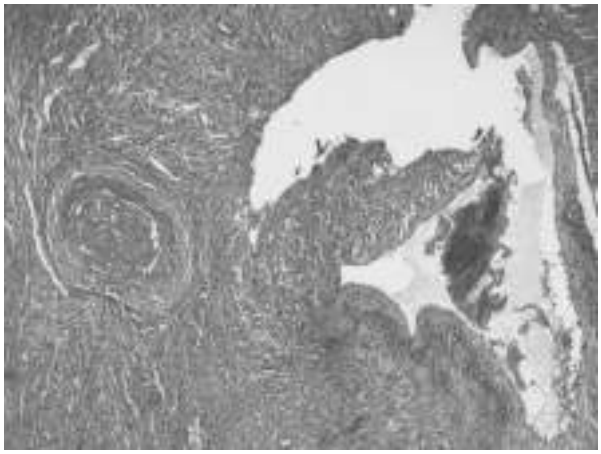

Figure 5. Angiomyolipoma, focal necrosis (HE stain, 50x)
Slika 5. *Angiomiolipom, fokalna nekroza, HE bojenje, 50x*

formed with additional blood replacement by resuspended erythrocytes and blood plasma. Transperitoneal approach to the left kidney was performed with a left subcostal incision. The renal artery and vein were ligated and cut, after which a nephrectomy and drainage of the massive retroperitoneal hematoma were performed. The total blood loss was about 2500 ml. By using a cell salvage device, 250 ml of red cells concentrate was saved and returned to circulation. Postoperatively, the patient was transferred to the intensive care unit. The recovery proceeded without complications. The patient was discharged on the tenth postoperative day and was followed up postoperatively; she had no complaints and the renal function was regular.

Pathohistological analysis of the surgically removed kidney confirmed a renal angiomyolipoma with the following immunohistochemical characteristics: HMB45+, MelanA+, STAT6-, CD34+, ER-, PR-, Calponin+, SMA+/-, p63-, GATA3-, RCC- **(Figures 4 and 5)**.

Discussion

The WS is a type of non-traumatic retroperitoneal hemorrhage. Massive retroperitoneal hemorrhage originating from the kidneys is a life-threatening condition that mainly requires urgent surgical treatment. The main risk factors for spontaneous rupture of angiomyolipoma are the size of the tumor, quantity and size of aneurysms in the tumor, pregnancy, as well as tuberous sclerosis.

In cases of spontaneous rupture of renal tumors, especially in patients who had not previously been diagnosed with renal tumors, radiological diagnosis plays a key role. The radiological method of choice is abdominal CT, even though at the time of hemorrhage its sensitivity is less than 60% [2].

Total nephrectomy is a preferable method of treatment if the cause of bleeding is a spontaneous rupture of renal cell carcinoma or angiomyolipoma larger than 5 cm in diameter. On the other hand, both traumatic and spontaneous retroperitoneal hemorrhage of renal origin can be treated conservatively, if the mechanism of occurrence and bleeding dynamic allows it (estimated by CT or magnetic resonance imaging). In cases of spontaneous rupture of a kidney tumor, radiological studies sometimes do not show the exact location of bleeding or the pathohistological type of the tumor, so surgical treatment is also a diagnostic procedure. The surgically removed tissue examined by a pathologist sets the final diagnosis and determines the further course of treatment and follow up of the patient. Cubillana et al. found that conservative management of WS represents a safe and acceptable treatment option in patients without malignant kidney tumors [3]. Sotosek et al. have successfully conservatively treated WS caused by spontaneous rupture of bilateral renal angiomyolipomas [4]. On the other hand, surgical approach is often necessary for tumors larger than 4 cm in diameter [5].

In our case, we performed a surgical treatment with total nephrectomy, due to the hemodynamic instability of the patient, who could not be treated conservatively. We decided to perform a total nephrectomy instead of a partial nephrectomy due to the size of the tumor, the extension of the tumor in the hilus of the kidney, as well as the existence of a functional contralateral kidney without pathological changes. Taking into account the massive bleeding as well as the findings of CT, which described a rupture of an angiomyolipoma and not a renal cell carcinoma, an intraoperative cell salvage device was used in order to save as much blood cells as possible. Cell salvage devices have been used for more than forty years in surgical treatment of various diseases [6]. A review of the existing literature did not reveal any previous intraoperative use of a cell salvage device in the management of WS. The patient also received fresh frozen blood plasma and resuspended erythrocytes intraoperatively.

Although angiomyolipoma is the most common benign kidney tumor, its prevalence is less than 0.15% in the general population. It is known that it is more common in women [7]. In most cases, patients with angiomyolipoma do not have symptoms and the tumor is mostly discovered incidentally using various radiological methods, primarily during examination of the abdomen and retroperitoneum with ultrasound, CT or magnetic resonance [8]. Low prevalence of this tumor in the population is the main reason why its rupture is

a relatively rare cause of sudden pain in the abdomen or retroperitoneum.

Conclusion

Wunderlich syndrome caused by rupture of angiomyolipoma is still a life-threatening condition that should be considered in the differential diagnosis of sudden abdominal pain. Spontaneous rupture of angiomyolipoma deserves its place in the differential diagnosis of sudden pain, especially in the population of fertile women. Available radiological methods should be used to make an accurate diagnosis in hypotensive patients before taking the patient to the operating room.

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