SPONTANEOUS BLEEDING FROM THE KIDNEYS TO THE RETROPERITONEUM (WUNDERLICH-SYNDROME)

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Abstract: Between years 2002 to 2013 we have treated 12 patients (8 M and 4 F) aged 19-83 years (mean 51.16 years old) with spontaneous retroperitoneal hemorrhage from the kidney. After initial examination and determination of the treatment plan we made laboratory tests, including blood and urine tests followed by Ultrasound and CT. In 10 (83.3%) patients, the bleeding from the kidney to the retroperitoneum was accompanied by acute hemorrhagic shock with the need for blood transfusion. The minimum hemoglobin values range was 6.7–11.1 g/l (average 9.05 g/l). Hematoma volume was estimated in the range of 500–2000 ml (average 1316 ml). Four (33.3%) patients were treated conservatively, the other 8 (66.7%) underwent nephrectomy for worsening hemorrhagic shock. Four (33.3%) patients had a tumor of the kidney, of which three were renal cell carcinomas and one was angiomylolipoma, two had renal cysts, one chronic glomerulonephritis and in the four patients that did not undergo surgery the reason of the spontaneous rupture of the kidney was unknown, but probably due to anticoagulant treatment. Bleeding from the kidneys to the retroperitoneum may be accompanied by acute hemorrhagic shock requiring blood transfusion. Surgical treatment is indicated, depending on the primary diagnosis, damage of the kidney and the extent of the bleeding. In the case of less bleeding into the retroperitoneum, a conservative approach may be chosen.

Key words: Wunderlich-syndrome – renal rupture – retroperitoneal bleeding – renal tumor

Introduction

Spontaneous rupture of the kidney with extensive bleeding into the kidney capsule or retroperitoneal space is a life-threatening condition, which was first described by Wunderlich in 1856 [1]. The most common causes of non-traumatic rupture of the kidneys include benign and malignant renal tumor (angiomyolipoma (AML) and renal cell carcinoma (RCC), functional disorders of coagulation and anticoagulation, anatomical and vascular abnormalities (aneurysms, fistulas, vasculitis, venous thrombosis), renal cysts and inflammation as well as other pathological conditions that affect the kidneys [2]. Wunderlich-syndrome should be distinguished from Herlyn–Werner–Wunderlich- (HWW) syndrome, which involves a rare developmental anomaly of the uterus (Uterus didelphys) with hemivagina and ipsilateral renal agenesis with a spectrum of anomalies of the Müllarian and Wolffian ducts [3].

Materials and Methods

In the period 2002–2013 we have treated 12 patients (8 M and 4 F) aged 19–83 years (mean 51.2 yrs.) with spontaneous retroperitoneal bleeding from the kidneys into the renal capsule and to the perirenal and pararenal retroperitoneal space. During examination a treatment plan was established which included laboratory tests, including blood tests, internal examination and urine tests. All the patients underwent USG and CT. File characteristics of

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patients are in Table 1. The follow-up period of conservatively treated patients ranged from 10–36 month, on average 26 month. At the end of the follow-up period of the non-surgically treated patients additional laboratory tests were performed along with USG and if applicable CT or DMSA renal scan. Patient history was evaluated for presence of secondary hypertension.

Results

In 10 (83.3%) patients bleeding from the kidney were accompanied by acute hemorrhagic shock, requiring infusion therapy and transfusion. Minimum hemoglobin value ranged from 6.7–11.1 g/l (average 9.05 g/l). Retroperitoneal hematoma volume estimate calculated from an ultrasound examination or CT ranged from 500–2000 ml (mean 1,316 l). Prior to kidney rupture five (41.6%) patients were on anticoagulants (Warfarin) and antithrombotic treatment for other cardiovascular causes. Four (33.3%) patients were treated without surgery and retroperitoneal drainage, another 8 (66.7%) patients were according to the primary diagnosis, extent of rupture, extent of bleeding and continued development of hemorrhagic shock treated by surgery, while in all nephrectomy was performed. Four (33.3%) patients had a tumor of the kidney, of which three RCC and the other AML, two hemorrhagic renal cyst, one chronic glomerulonephritis and no pathological changes in the kidney were histologically documented in the other patients who were treated conservatively by infusions and appropriate treatment. In one 62 year old patient after conservative management of spontaneous rupture of the kidney after treatment with warfarin for two years a shrunken kidney with loss of its function was seen. At follow-up in the other conservatively treated patients morphological and functional healing of the kidney occurred.

Discussion

Spontaneous rupture of the kidney with massive bleeding into the retroperitoneal space is a rare acute medical condition. In the literature are mostly reported sporadic case reports or small groups of patients [4]. Meta-analysis of patients with Wunderlich-syndrome (n 165) showed that the most common conditions were kidney tumors in 61.5% patients (31.5% benign most AML, RCC 29.7%), vascular causes (17%), anticoagulant treatment and hematological diseases (polycythemia) in 12.7%, idiopathic or unknown causes in 6.7% and in 2.4% infections [5]. The spontaneous rupture of the kidney associated with bleeding were also seen in patients with the diagnosis of polycystic kidney, renal cysts, vasculitis, hypertension, during treatment with aspirin or Dikumarol derivates, but there was also bleeding from the adrenal gland [6]. The literature review in Pubmed described also other etiological causes. As can be seen from Table 1, in our cohort compared with other data in the literature, only in four (33.3%) patients the kidney tumors were seen, of which only one with AML. We observed the most frequently in patients with retroperitoneal hemorrhage idiopathic of the retroperitoneal rupture of the kidney and in a significant proportion of those receiving antithrombotic or anticoagulant treatment for cardiovascular causes. Treating of patients with Wunderlich-syndrome can be done in two important ways. If the extent of the rupture and bleeding is minimal, does not jeopardize patient with significant blood loss and hemorrhagic shock, hematoma location is outside the vascular stem and the pyeloureteral junction, or no risk of formation of secondary hypertension and hydronephrosis due to fibrosis or impaired organizing hematoma, the patients can be
followed conservatively without surgery or external drainage around the kidneys. The aim of conservative treatment is to allow the kidney to recover spontaneously and fully restore its function. Otherwise, surgical intervention is necessary, which is usually as in our study, dependant on the primary cause and extent of damage to the kidneys, usually results in transperitoneal nephrectomy. Currently were described kidney-sparing procedures either robotically assisted resection of the kidney [7] or selective embolization of bleeding blood vessels [8]. Renal angiography in selected patients not only helped identify the bleeding vessel, but also enabled active control of bleeding by selective embolization, avoiding urgent nephrectomy [9]. These procedures can be used in stabilized patients especially in the early stages of bleeding, if a large hematoma is not present and elective nephrectomy in not planned. Patients on antiplatelet therapy with acetylsalicylic acid may suffer from repeated spontaneous hemorrhages. In such cases reexamination of the coagulation status is important [10]. In both conservatively treated patients as well as patients with kidney restorative surgery follow-up is always necessary to check the function of the kidneys for the emergence of secondary changes due to the damage (hypertension, hydronephrosis).

**Conclusion**

Bleedings from the kidneys of various origins may be accompanied by the same symptoms, usually even acute hemorrhagic shock. It is a condition requiring immediate emergency care. Surgical treatment is indicated, depending on the primary diagnosis and the extent of bleeding. A conservative approach may be chosen if less bleeding into the retroperitoneum is present. The most common surgical procedure in our group of patients was nephrectomy.

**Table 1**

Data of patients with Wunderlich-syndrome who were treated at the Department of Urology at Košice in the years 2002–2013.

<table>
<thead>
<tr>
<th>Patient</th>
<th>M / F</th>
<th>Age</th>
<th>Year</th>
<th>Histology</th>
<th>AC</th>
<th>Th</th>
<th>Losses</th>
</tr>
</thead>
<tbody>
<tr>
<td>1/BA</td>
<td>M</td>
<td>41</td>
<td>2004</td>
<td>CH-RCC</td>
<td>–</td>
<td>NE</td>
<td>500</td>
</tr>
<tr>
<td>2/KL</td>
<td>M</td>
<td>47</td>
<td>2007</td>
<td>P-RCC</td>
<td>–</td>
<td>NE</td>
<td>1000</td>
</tr>
<tr>
<td>3/SM</td>
<td>F</td>
<td>63</td>
<td>2009</td>
<td>CC-RCC</td>
<td>–</td>
<td>NE</td>
<td>1000</td>
</tr>
<tr>
<td>4/KA</td>
<td>F</td>
<td>65</td>
<td>2010</td>
<td>AML</td>
<td>–</td>
<td>NE</td>
<td>2000</td>
</tr>
<tr>
<td>5/BI</td>
<td>M</td>
<td>62</td>
<td>2010</td>
<td>0</td>
<td>+</td>
<td>K</td>
<td>500</td>
</tr>
<tr>
<td>6/TI</td>
<td>M</td>
<td>19</td>
<td>2010</td>
<td>0</td>
<td>+</td>
<td>K</td>
<td>1000</td>
</tr>
<tr>
<td>No.</td>
<td>Name</td>
<td>Sex</td>
<td>Age</td>
<td>Year</td>
<td>Diagnosis</td>
<td>Status</td>
<td>Date</td>
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<tr>
<td>7/HE</td>
<td>6</td>
<td>F</td>
<td>65</td>
<td>2010</td>
<td>N</td>
<td>+</td>
<td>NE</td>
</tr>
<tr>
<td>8/KM</td>
<td>2</td>
<td>M</td>
<td>27</td>
<td>2011</td>
<td>Cyst</td>
<td>−</td>
<td>NE</td>
</tr>
<tr>
<td>9/KM</td>
<td>3</td>
<td>M</td>
<td>27</td>
<td>2011</td>
<td>Cyst</td>
<td>−</td>
<td>NE</td>
</tr>
<tr>
<td>10/JL</td>
<td>0</td>
<td>M</td>
<td>66</td>
<td>2011</td>
<td>0</td>
<td>+</td>
<td>K</td>
</tr>
<tr>
<td>11/DJ</td>
<td>5</td>
<td>M</td>
<td>49</td>
<td>2011</td>
<td>GN</td>
<td>−</td>
<td>NE</td>
</tr>
<tr>
<td>12/BM</td>
<td>5</td>
<td>F</td>
<td>83</td>
<td>2012</td>
<td>0</td>
<td>+</td>
<td>K</td>
</tr>
</tbody>
</table>

Legend: M – male, F – female, AC – anticoagulant and antiplatelet therapy is indicated from cardiovascular causes, CH-RCC, chromophobe renal cell carcinoma, P-RCC-papillary renal cell carcinoma, CC RCC RCC of clear cell, AML – angiomyolipoma, 0 – histological examination N – normal – histologically parenchyma without pathological changes, GN – glomerulonephritis, renal cyst cyst-verified histopathological examination and medical history before rupture, Hb – the lowest measured hemoglobin. Trans. – transfusion; Note-Aff. r.: Non-functioning kidney after 2 years found 99 mTc DMSA scan types.

Fig. 1. Rupture of the kidney with AML in 65 y. patient with hematoma enclosed by the left half of the retroperitoneal space
Fig. 2. Hemorrhage into the cyst with destruction of the right kidney in a 27 y. patient

Fig. 3. Spontaneous rupture of the right kidney in 62 y. patient on a warfarin therapy with a subsequent loss of renal function after 2 y.
Acknowledgement

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References