Spontaneous subcapsular kidney haemorrhage – the first symptom of renal cell carcinoma

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Abstract

Spontaneous subcapsular kidney haemorrhage is a rare clinical problem of both malignant and non-malignant kidney disease, also known as Wunderlich syndrome. Spontaneous subcapsular haemorrhage in the kidney happens mostly in patients with angiomyolipoma and is less common in renal carcinoma. We present a patient in whom the first symptom of renal carcinoma involved haematoma caused by spontaneous rupture of a neoplastic tumour.

Key words: kidney, spontaneous subcapsular kidney haemorrhage, renal cell carcinoma.

Introduction

Spontaneous kidney rupture involves disruption of continuity of the renal parenchyma or collecting system, with no reported trauma in clinical history. In the year 1700 Bonet *et al*. first reported spontaneous retroperitoneal kidney haemorrhage (SRH). The first description of the clinical picture of spontaneous kidney rupture with subcapsular haemorrhage and/or bleeding into the retroperitoneal space was reported by Wunderlich in 1856 [1]. Renal rupture followed by formation of subcapsular, retroperitoneal haemorrhage and intraperitoneal bleeding is most common in patients with chronic renal insufficiency in the course of systematic diseases. Organ rupture may happen in chronically dialyzed patients. As the literature suggests, spontaneous renal rupture happens mostly in patients with angiomyolipoma (AML) and is less common in renal carcinoma. Lumbar pain is the most common symptom of spontaneous kidney rupture. A shock caused by massive retroperitoneal or intraperitoneal haemorrhage may develop in isolated cases [2].

We present a patient in whom the first symptom of renal carcinoma involved haematoma caused by spontaneous rupture of a neoplastic tumour.

Case report

A male patient aged 63 was admitted to the urology department because of subcapsular haematoma of the right kidney. Imaging diagnostic methods were performed in the outpatient clinic department since the patient had complained of right-sided lumbar pain which had appeared

spontaneously without any known history of trauma. Abdominal ultrasound examination in this patient showed irregular kidney limits with a liquid space around. The collecting system of this kidney was not enlarged. The left kidney appeared normal. Computed tomography (CT) examination showed hyperdense masses measuring mean +60 H.U. (Hounsfield units), co-existing with liquid spaces in the right retroperitoneal space (Figure 1). They surrounded the right kidney, had regular external limits and were probably surrounded by the renal capsule. Adjacent linear infiltration of the neighbouring adipose tissue was observed. The right kidney was compressed by the above-mentioned abnormal masses, while the left kidney appeared normal in CT imaging. Function of both kidneys was preserved. No peritoneal or extraperitoneal enlarged lymph nodes were seen. Laboratory results did not show any pathologies in blood morphology or the coagulation system. The patient was qualified for a surgical procedure. The right retroperitoneal space was reached *via* latero-posterior lumbar access. A greatly enlarged kidney with an adipose capsule of a yellowish-bluish colour was found. Numerous planar adhesions with the adjacent tissues were seen. The adipose capsule of the abnormal kidney was incised along its lateral side – the space between renal parenchyma and the capsule was occupied by masses of partially haemolyzed haematoma. In the superior renal pole a solid tumour mass measuring 5 cm with irregular external limits was found. The mass being suspected of tumour, a decision to remove the organ was made. The kidney was totally removed with its adipose capsule. Histological examination confirmed the presence of a tumour of 5 cm, with necrotic characteristics inside the tumour – clear cell carcinoma grade II, with diffused necrosis and haemorrhage.

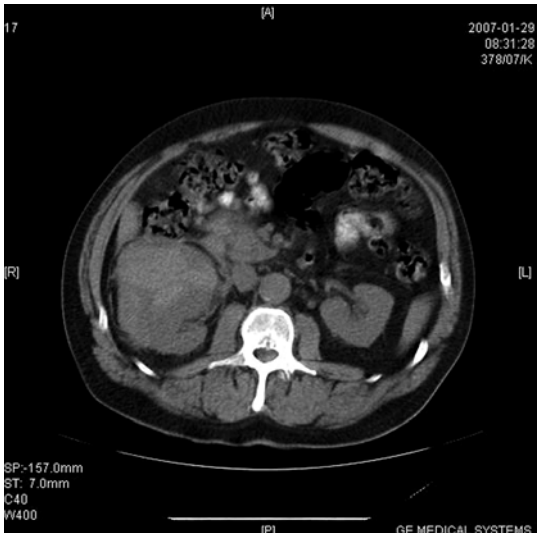

Figure 1. Computed tomography scan – perirenal masses of haematoma manifesting density of around + 60 H.U. (Hounsfield units). They surrounded the right kidney, had regular external limits and were probably surrounded by the renal capsule. The right kidney was compressed by the above-mentioned abnormal masses; the left kidney appeared normal in CT imaging. Masses of the haematoma precluded an unequivocal evaluation of the CT pattern

The post-operational period was uncomplicated and the patient was discharged home in a good state on the 8th day.

Discussion

In 1856 Wunderlich first described the clinical picture of spontaneous subcapsular renal bleeding without any trauma history. From 1933 to 1985 a few general articles reviewing 283 patients with spontaneous perirenal haemorrhage were published. The most common causes included bleeding in renal neoplasms (63%), vascular abnormalities (26%) and infections (10%). These results are similar to those published in 2004 in a meta-analysis reviewing 165 cases described in the literature. Renal tumours were responsible for spontaneous bleeding in 61.5% of the cases. In 11 cases no cause of bleeding was defined. Benign tumours accounted for 31.5% of the neoplasms and included: angiomyolipoma (AML) (29.1%), myelolipoma (1.2%), adenoma (1.2%) and a single case of oncocytoma. Malignant tumours comprised 29.7% of the neoplasms, of which renal cell carcinoma (RCC) was the cause of bleeding in 26.1% of cases, metastases in 2.4%, and in a single case sarcoma induced the bleeding. Both an older analysis conducted by Cinnaman *et al.* and the results presented by Zhang *et al*. in 2004 confirmed that the most common cause of spontaneous haemorrhage involves the benign renal tumour called angiomyolipoma. According to Cinnaman *et al.*, angiomyolipoma may provide the cause of bleeding in 33%, and Zhang *et al.* confirmed that the tumour may result in the development of haematoma in 29.1% of cases. Vascular disease leads to haematomas in about 17% of cases, and includes most often polyarteritis nodosa, aneurysms, arteriovenous abnormalities, portal hypertension, and Wegener's granulomatosis [2-4].

From 1985 to 1999, 47 papers were published in English-language literature, describing 165 cases of spontaneous perirenal haematomas. In Europe an average of around 5 and in the USA of around 4 articles on SRH were published annually [3].

Renal clear cell carcinoma (RCC) represents one of the most frequent malignant tumours in the kidneys. The clinical presentation of renal cell carcinoma is usually variable. Spontaneous renal haemorrhage as the presenting symptom of renal

cancer is rare. The incidence of spontaneous rupture of RCC is 0.3-0.6%. In an article written by Skinner *et al.*, at the Massachusetts General Hospital the complication was noted in only one of 329 patients [5]. As indicated by relevant literature, angiomyolipoma is one of the most frequent causes of a spontaneous renal haemorrhage. The tumour is formed from mature adipocytes, smooth muscles and blood vessels, which are devoid of elastic fibres. Lack of the fibres in vascular walls causes them to be susceptible to microtraumas (distension) and this may explain the propensity of the tumours to bleeding [6-8].

The predominant symptom described in the literature which led to further diagnostics was lumbar pain of various levels of intensity. Micro- or macro-haematuria or shock symptoms appeared much less often.

Ultrasonography (US) is the basic imaging method of such pathologies. As a generally accessible, reproducible and non-invasive procedure, it enables the imaging of abnormal masses which surround the ruptured kidney, but it fails to clearly define the primary cause of the rupture. According to relevant literature, computed tomography (CT) manifests 100% sensitivity in the detection of haematoma in the retroperitoneal space and, as compared to US, it is more sensitive and specific in the recognition of the causes of haematoma. Computed tomography, however, enables full imaging of perirenal or subcapsular haematoma. Specificity and sensitivity of diagnosis in cases of renal pathologies in kidneys with haematoma are 0.57 and 0.82, respectively, for CT, and 0.11 and 0.33 for US. In cases of pronounced bleeding from a ruptured kidney, the interpretation of CT images may be hampered by the presence of extravasated blood, which may mask the actual cause of bleeding [3, 7, 8].

Characteristic radiological features of angiomyolipomas with a size less than 4 cm and insignificant clinical haemorrhage require nephron-sparing surgery. In cases of significant bleeding from the ruptured angiomyolipoma type tumour, embolisation of the bleeding vessel may precede further surgical treatment. In the case of renal carcinoma, there is no relation described in the literature between the tumour size and the advancement of renal haemorrhage. As indicated by the data available in the literature, a nephrectomy is performed in around 68% of patients with spontaneous perirenal haematoma. The selection of such a procedure reflects the conviction of numerous urologists that the cause of haematoma development may involve an undiagnosed neoplasm. In around 10.3% of patients, drainage of the haematoma provided the treatment of choice. In 9.7% of patients in whom imaging studies led to no suspicion of neoplastic lesions, a conservative procedure was implemented, involving monitoring the patient's general condition and evolution of the haematoma at its stage of resorption. In 4.8% of cases, exposure of the kidney and evacuation of the haematoma were followed by a partial resection of the kidney. The selection of such a procedure may reflect relative indications (patients with impoverished function of the other kidney) or may represent the procedure of choice due to the tumour size, its location or condition of draining lymph nodes. According to Pummer *et al.*, in a proportion of patients embolisation of the acutely bleeding vessel can be conducted and at a later stage, when the condition of the patient is stable, the kidney may be exposed, the haematoma evacuated and, if needed, nephrectomy can be performed. As indicated by the literature, embolisation is performed in around 4.2% of patients with SRH. If the decision is taken to precede surgery with a conservative treatment, it should be kept in mind that in cases of renal rupture in the course of RCC protracted bleeding to the retroperitoneal space may result in dissemination of the neoplastic disease [3, 7, 9-11].

In the presented case, urological diagnostic methods were started because of the right lumbar pain. Both US and CT examinations confirmed the presence of subcapsular haematoma of an unknown origin. Having found intraoperatively a tumour mass measuring more than 4 cm and located in the superior pole of the right kidney, the decision of radical organ removal was taken. Histological examination later confirmed the presence of clear cell carcinoma which had caused the bleeding.

In conclusion, spontaneous perirenal bleeding seldom represents the first sign of cancer (renal cell carcinoma). Renal cell carcinoma represents the second most frequent cause of SRH, after tumours of angiomyolipoma (AML) type. Ultrasonography and CT are useful in diagnosing perirenal haematoma but they do not always provide the unequivocal cause of the bleeding.

The unclear aetiology of the haematoma (a suspicion of renal tumour) should prompt the therapist to determine the cause of its development using surgery. In cases of intraoperative suspicion of a tumour, one of the therapeutic options is a nephrectomy, provided that the other kidney manifests normal function. In selected cases, the NSS (nephron-sparing surgery) approach or emergency embolisation of bleeding blood vessels can be implemented before the postponed surgery is conducted (provided that tumour presence has been confirmed).

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