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Case Report / Приказ болесника

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Wunderlich syndrome – a case report and brief literature review

Вундерлихов синдром – приказ случаја и кратак преглед литературе

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SUMMARY

Introduction Renal angiomyolipomas (AMLs) are neoplasms that can rarely rupture, causing hemorrhagic shock as the most serious complication. This pathological condition, (referring to AML) is classified as a benign tumor arising from the proliferation of epithelioid cells, consisting of fat tissue, blood vessels, and smooth muscle. Wunderlich syndrome describes a spontaneous, non-traumatic bleeding into the subcapsular or perirenal space. Most individuals with renal AML exhibit no symptoms and are often diagnosed incidentally, however, some may experience life-threatening complications such as rupture, hemorrhage, and circumstantial hypovolemic shock.

Case outline Description of the clinical presentation of AML with rupture in a female patient with a brief overview of other cases of AML in the literature. Female patient, 68 years old, admitted for examination due to sudden severe pain in the abdomen with propagation to the right lumbar region accompanied by nausea and fatigue. After a complete physical examination, an abdominal ultrasound, and a CT scan, surgery was performed during which the right kidney was removed alongside the hematoma and the kidney envelopes, which were sent for pathohistological analysis. The result of the histopathological analysis confirmed that it was AML.

Conclusion AMLs are benign neoplasms with potentially serious complications. The most serious complication of AML is rupture, leading to retroperitoneal hemorrhage, with tumor size being a significant risk factor. Considering the clinical importance of this potential complication, it is important to establish a swift and accurate radiological diagnosis, with the aim of timely therapeutic intervention and reduction of potential additional complications.

Keywords: renal angiomyolipoma; radiological diagnosis; rupture; management

САЖЕТАК

Увод Ренални ангиомиолипоми (АМЛ) су бенигне неоплазме са ниским ризиком од спонтане руптуре, који могу узроковати хеморагијски шок као најтежу компликацију. АМЛ се састоје од масног ткива, крвних судова и глатких мишића. Вундерлихов синдром се односи на спонтано, нетрауматско крварење из бубрега у субкапсуларни и/или перирenални простор. Већина пацијената са реналним АМЛ-ом је асимптоматска и дијагностикује се приликом рутинских прегледа, док неки могу доживети озбиљне компликације попут руптуре, крварења или хиповолемичног шока.

Приказ болесника Опис клиничке презентације АМЛ-а са руптуром код пацијенткиње уз кратак преглед других случајева АМЛ-а у литератури. Пацијенткиња, старости 68 година, примљена у хитну службу због изненадних, јаких болова у стомаку са ширењем у десни лумбални регион, који су праћени мучнином и слабошћу. Након клиничког прегледа, ултразвука абдомена и ЦТ прегледа абдомена и карлице, урађена је операција током које је одстрањен десни бубрег заједно са хематомом, који су послати на патохистолошку анализу. Резултат патохистолошке анализе потврдио је да се ради о АМЛ-у.

Закључак Ангиомиолипоми су бенигне неоплазме са потенцијално озбиљним компликацијама. Најтежа компликација АМЛ-а је руптура, што доводи до ретроперитонеалног крварења, при чему је величина тумора значајан фактор ризика. Обзиром на клиничку важност ове потенцијалне компликације, битно је успоставити брзу и тачну радиолошку дијагнозу, са циљем правовремене терапијске интервенције и смањења могућих додатних компликација.

Кључне речи: ренални ангиомиолипом; радиолошка дијагноза; руптуре; лечење

INTRODUCTION

Renal angiomyolipomas (AMLs) are neoplasms that can rarely rupture, causing hemorrhagic shock as the most serious complication. This pathological condition, (referring to AML) is classified as a benign tumor arising from the proliferation of epithelioid cells, consisting of fat

tissue, blood vessels, and smooth muscle. AMLs larger than 4 cm carry a notably increased risk of rupture; however, rupture can also occur in smaller tumors, while larger AMLs may remain stable. The median age for rupture of solitary renal AMLs is around 50 years. The overall incidence is approximately 0.13%, with a higher prevalence in females, which is thought to be related to hormonal influences. About 80% of renal AMLs occur sporadically, whereas the remaining cases are linked to tuberous sclerosis [1]. Wunderlich syndrome describes spontaneous, non-traumatic bleeding into the subcapsular or perirenal space. The appropriate management of this syndrome depends on confirming the diagnosis of perinephric hemorrhage and identifying its underlying cause [2]. Most individuals with renal AML exhibit no symptoms and are often diagnosed incidentally, however, some may experience life-threatening complications such as rupture, hemorrhage, or hypovolemic shock. Traditionally, a tumor size of 4 cm has been used as a threshold - initially proposed in 1986 - to differentiate between patients suitable for watchful waiting and those needing intervention. Nevertheless, this criterion is now subject to debate, with recent research suggesting that relying solely on size could result in unnecessary treatment in certain cases [3, 4].

This case report describes a female patient who experienced spontaneous perinephric bleeding due to a ruptured renal angiomyolipoma (AML), alongside a short review of this syndrome.

Patient consent was obtained in writing, and the case report adheres to ethical protocols in accordance with institutional guidelines and applicable regulations.

CASE REPORT

Patient, aged 68, was admitted to the emergency care unit of KBC "Dr Dragiša Mišović – Dedinje", Belgrade, in October of 2023, due to severe, sudden pain in the right lumbar region. The pain started 3 days before the clinical exam with localization in the left lower back. She was initially examined by a physiatrist, who determined that the pain was of spinal origin and

prescribed dexamethasone. Although she felt better for a short time, on the day of the examination, the pain reappeared, with propagation to the right, accompanied by vomiting and weakness. The following medical tests were performed:

General exam: TA 115/70 mmHg, pulse 64 bpm. Soft abdomen, painfully sensitive to deep palpation on the right.

Laboratory analyses: Leukocytes 20.4 × 10^9/L, Hemoglobin 128 g/L, Erythrocytes 4.28 × 10^12/L.

Abdominal ultrasound: The right kidney was enlarged, with a hyperechoic mass encompassing the interpolar region and the upper pole of the kidney, extending into the renal sinus. Within the mass there were irregular anechoic fields, with a total diameter of approximately 90 × 60 mm. Along the lateral contour of the kidney, there was an anechoic, dense, fluid collection with hyperechoic streaks, and inflamed perirenal fatty tissue, suggestive of a hematoma originating from a bleeding angiomyolipoma (Figure 1).

CT: Non-contrast and contrast-enhanced computed tomography (CT) scans of the abdomen and pelvis revealed significant changes in the right kidney. A heterodense formation, measuring about 85 × 60 mm with hypodense zones (of negative density - fat) was observed, likely corresponding to the previously described angiomyolipoma. Perirenal fluid was observed with density measuring 50–60 HU (density of fresh blood), 50 mm in diameter. Additionally. perirenal adipose tissue appeared inflamed, permeated with hyperechoic streaks. The fluid collection propagated medially into the para-aortic space in the upper retroperitoneum. Subhepatic free fluid with a layer thickness of approximately 12 mm was present. The right adrenal gland was not visualized, in addition to the described fluid collection. The left kidney had a CC diameter of 92 mm, with several focal changes of fat density in the parenchyma, reaching a diameter of up to 8 mm, characteristic of AML.

In conclusion, these findings support acute bleeding into the right perirenal space, most likely originating from a large angiomyolipoma situated in the superior portion of the right kidney (Figures 2, 3, 4).

After the completion of the diagnostic procedures, a surgery was performed in which the right kidney was removed alongside the hematoma and the kidney envelopes, which were then sent for pathohistological analysis. On the cross-section of the kidney, a tumorous change was observed in the upper half of the kidney parenchyma, spreading to the fatty tissue of the hilum, of a soft consistency and with areas of hemorrhage, with an approximate size of 96 × 80 mm. Pathohistological analysis revealed that it was an angiomyolipoma.

DISCUSSION

Angiomyolipomas (AMLs) are uncommon neoplasms first characterized by Morgan et al. in 1951. Although the majority of these tumors are found within the kidneys, they can also occur in other anatomical sites such as the liver, spleen, uterus, and fallopian tubes. Renal angiomyolipoma (AML) represents a benign neoplasm that presents sporadically and in association with genetic syndromes like tuberous sclerosis (TS) and lymphangioleiomyomatosis. Renal AMLs consist of mesenchymal elements of the kidney and are composed of varying amounts of mature adipose tissue, smooth muscle, and vessels with excessively thickened walls, which is why they are referred to as renal hamartomas [5, 6]. Angiomyolipomas (AMLs) are categorized into three main types: classic AML, fat-deficient AML, and epithelioid AML [7]. The most significant risk linked to angiomyolipomas (AML) is bleeding into the retroperitoneal space, which occurs when the tumor ruptures. Conventionally, the evaluation of risk has been primarily based on tumor dimensions, with tumors exceeding 4 cm regarded as having a greater propensity for aneurysm formation and

rupture. However, recent insights suggest that additional factors - such as aneurysm presence, pregnancy, trauma, coagulopathies, hormonal fluctuations, and comorbidities like tuberous sclerosis and lymphangioleiomyomatosis - also play significant roles in influencing rupture risk. There is evidence suggesting that genetic irregularities may serve as an early contributing factor in the intricate interplay of various risk elements associated with tumor rupture [8]. The development of aneurysms may play a significant role in the risk of tumor rupture, with the contribution largely dependent on the presence and size of these vascular dilations. Aneurysms are classified into two main categories based on their pathology: true aneurysms and pseudoaneurysms. True aneurysms, also known as primary aneurysms, involve all three layers of the arterial wall. In contrast, pseudoaneurysms - sometimes referred to as false aneurysms - are hematomas restricted by surrounding tissues. These often result from traumatic injuries or other breaches in the vessel wall [9,10]. Pregnancy is also a significant factor influencing both the progression and the potential rupture of these tumors. The hormonal and hemodynamic changes that occur during gestation can accelerate tumor growth and increase the likelihood of complications such as rupture [11]. Computed tomography (CT) remains the most frequently employed imaging technique for the diagnosis of AML. The classic form of AML typically manifests as a lesion with predominant fatty attenuation, easily identifiable by its characteristic low-density appearance. In contrast, fat-poor AMLs tend to exhibit attenuation levels that are similar to or higher than surrounding tissues, often showing uniform enhancement after contrast administration. Epithelioid AMLs, however, generally appear as hyperattenuating masses with a heterogeneous enhancement pattern, sometimes resembling multilocular cystic structures. This variability in imaging features reflects the diverse pathological compositions of these tumor subtypes [12]. Management of all AMLs must be based on a clear diagnosis combined with a thorough evaluation of the potential for tumor rupture. Reasons to initiate treatment encompass concerns of cancerous transformation as well as episodes of spontaneous bleeding,

risk of rupture, and risk for other complications. Treatment options range from drug therapy to embolization to surgical extirpation and depend on each individual case [8].

Recent studies published in leading urology and radiology journals have highlighted significant progress in understanding Wunderlich syndrome. Advances in imaging technologies, particularly contrast-enhanced computed tomography, have greatly enhanced the ability to diagnose spontaneous renal hemorrhages efficiently, often before hemodynamic instability occurs [8]. Additionally, the latest research focuses on minimally invasive interventions, such as superselective embolization, which effectively control bleeding while preserving renal function and minimizing recovery times [13]. Furthermore, recent reviews emphasize the importance of early detection of underlying vascular anomalies, like aneurysms or malformations, which are critical for preventing recurrence and planning targeted therapy [14]. The evolving understanding of AML has contributed to increased awareness and underscored the importance of developing personalized, minimally invasive treatment approaches aimed at enhancing patient outcomes, including prognosis and quality of life for those affected by Wunderlich syndrome.

Although they are non-cancerous lesions of the renal tissue, angiomyolipomas can, in rare cases, rupture and trigger hemorrhagic shock due to sudden bleeding. Described as a non-malignant growth, this lesion arises from the unchecked multiplication of epithelioid cells and is composed of a mixture of fat tissue, vascular structures, and smooth muscle fibers. Retroperitoneal hemorrhage, stemming from the rupture of the tumor, stands out as the primary complication linked to AML. The probability of this complication increases with larger tumor dimensions, making size a key predisposing factor. Taking into account the clinical importance and seriousness of this potential complication, it is important to establish an accurate radiological diagnosis in a timely manner with the aim of timely therapeutic intervention and

reducing the risk of potential additional complications.

Conflict of interest: None declared.

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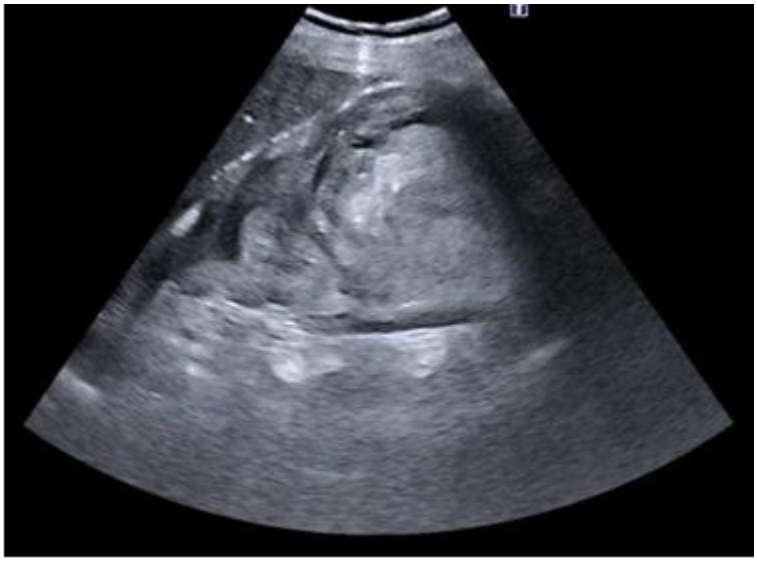

Figure 1. Abdominal ultrasound: the right kidney is enlarged, with a hyperechoic mass encompassing the interpolar region and the upper pole of the kidney, extending into the renal sinus; within the mass there are irregular anechoic fields; along the lateral contour of the kidney, there is an anechoic, dense, fluid collection with hyperechoic streaks, and inflamed perirenal fatty tissue, suggestive of a hematoma originating from a bleeding angiomyolipoma

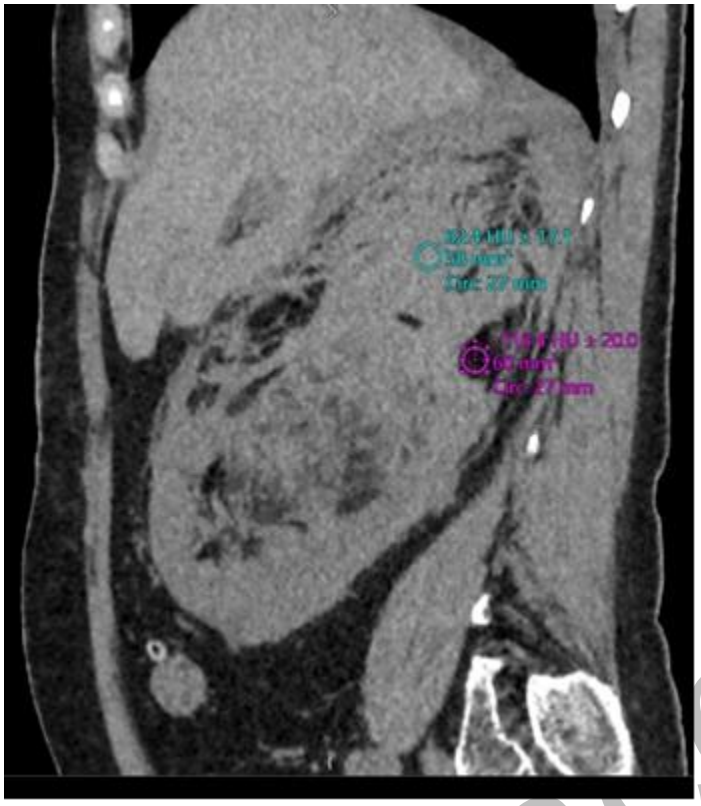

Figure 2. Sagittal unenhanced computed tomography imaging of the abdomen and pelvis showing altered right kidney with a mass situated within the interpolar region and at the upper pole of the kidney, measuring 175 × 110 × 65mm, with central fat density, alongside perirenal fluid with density measuring 50–60 HU (density of fresh blood), 50 mm in diameter

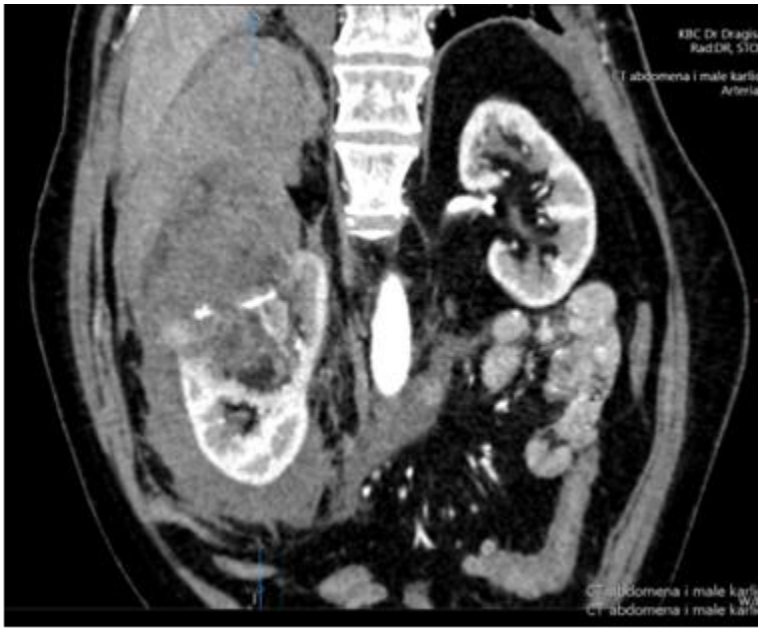

Figure 3. An intravenous-contrast-enhanced coronal computed tomography image shows perirenal hematoma of the right kidney and free fluid

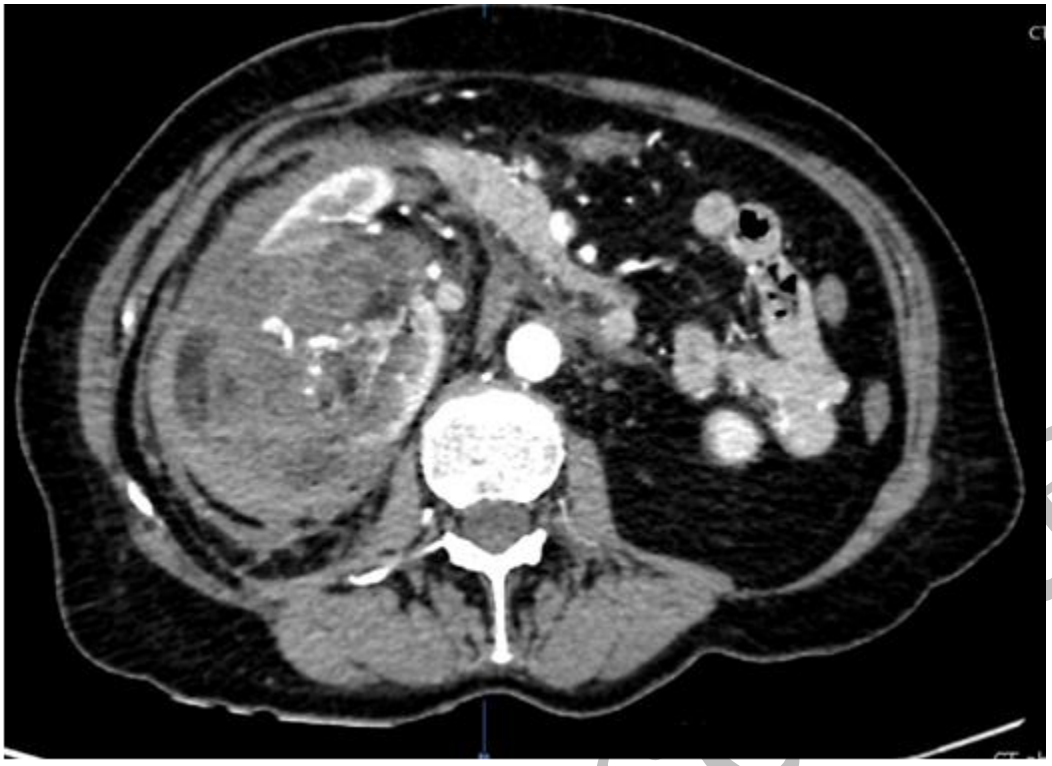

Figure 4. An intravenous-contrast-enhanced coronal computed tomography image revealed retroperitoneal hematoma, from the level of L2 vertebra

