

Lupus vasculitis complicated by a retroperitoneal hematoma: A case report

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Abstract

Context: Vasculitis associated with systemic lupus erythematosus (SLE) is a rare but potentially serious complication that may involve blood vessels of various calibers. Spontaneous retroperitoneal hematoma represents an exceptional manifestation of this vascular involvement, and its diagnosis relies largely on imaging.

Case Presentation: A 36-year-old female patient presented to the emergency with afebrile right-sided low back pain associated with marked pallor. An abdominal CT angiography revealed multiple confluent right-sided retroperitoneal hematomas associated with a pseudoaneurysm arising from the ipsilateral renal artery. The patient subsequently underwent conventional arteriography, which demonstrated a thin and irregular appearance of the ipsilateral renal artery, consistent with large-vessel vasculitis.

Conclusion: This case highlights the crucial role of imaging, particularly CT angioscanner, in the diagnosis of rare vascular complications of systemic lupus erythematosus. Early identification of a vasculitis-related retroperitoneal hematoma allows prompt therapeutic management and improves patient outcomes.

Keywords: Lupus Vasculitis; Retroperitoneal hematoma; Hemorrhage; CT scan; Interventional radiology; Embolization

1. Introduction

Systemic lupus erythematosus (SLE) is a chronic autoimmune disease with polymorphic clinical manifestations, which may affect numerous organs and systems. Among its complications, lupus vasculitis represents a relatively rare but potentially serious involvement, related to inflammation of the vascular wall that may affect vessels of different calibers. It is responsible for varied clinical manifestations, sometimes severe, that may be life-threatening.

Spontaneous retroperitoneal hematoma represents an unusual and often serious clinical entity, most commonly associated with trauma, anticoagulant therapy, or tumoral pathology. Its occurrence in the setting of lupus vasculitis remains exceptional and poses a diagnostic challenge due to the nonspecific nature of clinical and biological signs.

Our report is one of the few to detail this presentation, involving a 36-year-old woman with no particular medical history, who presented with retroperitoneal bleeding secondary to lupus-related renal artery vasculitis.

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2. Case presentation

A 36-year-old woman with no medical or surgical history presented to the emergency department with right-sided low back pain evolving for 3 days. On the clinical examination, the patient was conscious, with pale conjunctivae, afebrile at 36.8 °C, and her vital parameters were within normal limits, with a blood pressure of 11/7 mm/Hg and a heart rate of 82 beats/min. Abdominal examination revealed abdominal tenderness of the right flank and right iliac fossa, with no other associated signs.

Routine laboratory tests showed normochromic normocytic anemia with a hemoglobin level of 3.9 g/dL (normal value: 12–16 g/dL), kidney failure with a urea level of 3.3 g/L (normal value: 0.15–0.45 g/L), and a creatinine level of 111 mg/L (normal value: 6–11 mg/L), as well as hyponatremia at 128 mmol/L (normal value: 135–145 mmol/L).

An abdominal angioscanner was performed, revealing multiple right peri- and pararenal hematomas extending along the psoas muscle down to the level of the right iliac fossa, associated with a large pseudoaneurysm appearing to arise from the posterior segmental branch of the right renal artery, which was thin and irregular. A moderate intraperitoneal effusion due to transudation was also noted. (Figure 1) (Figure 2).

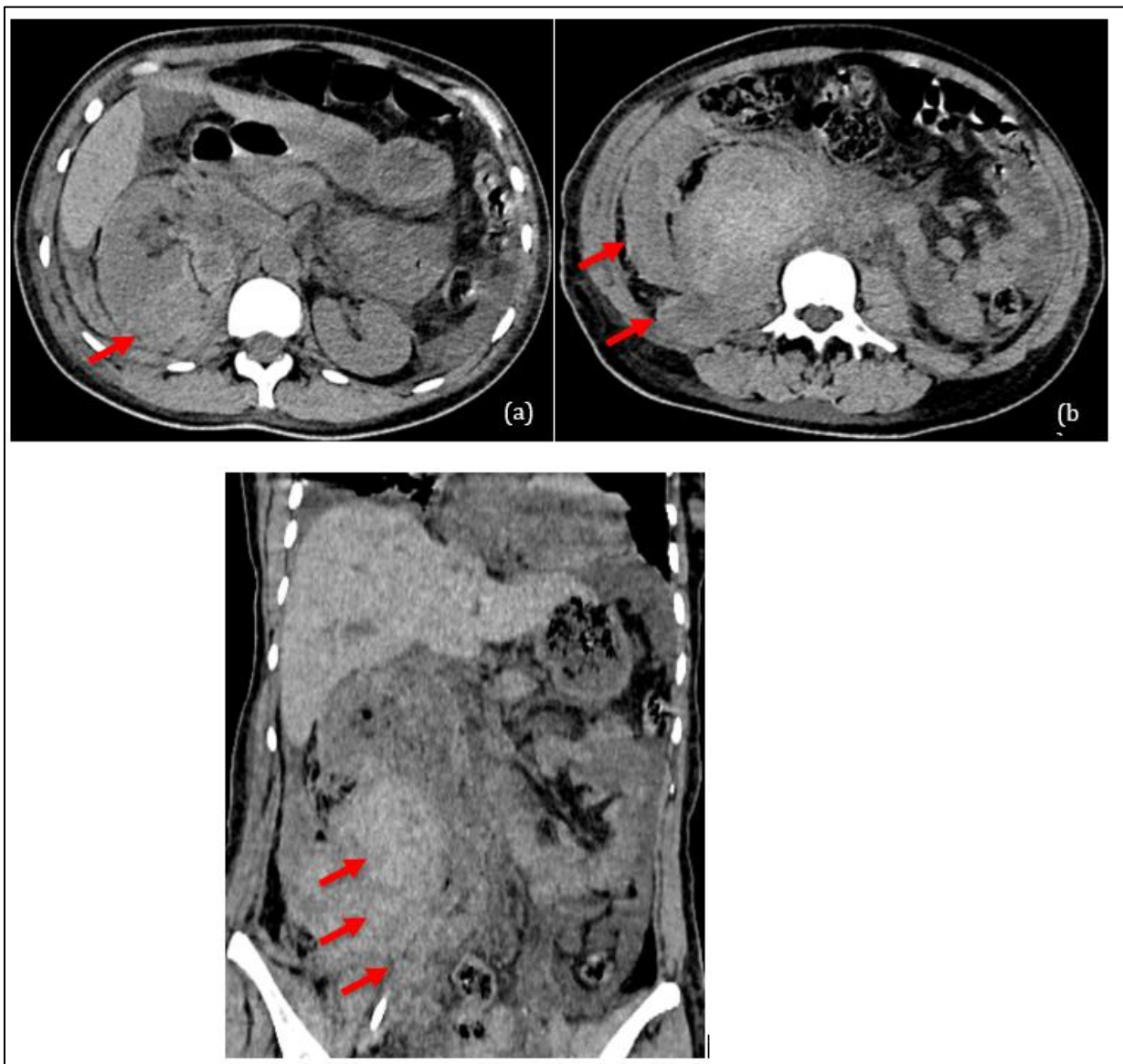


Figure 1 Spontaneous contrast abdominal scan in axial (a, b) and coronal (c) sections, showing the presence of multiple right peri- and pararenal hematomas extending the psoas muscle down to the right iliac

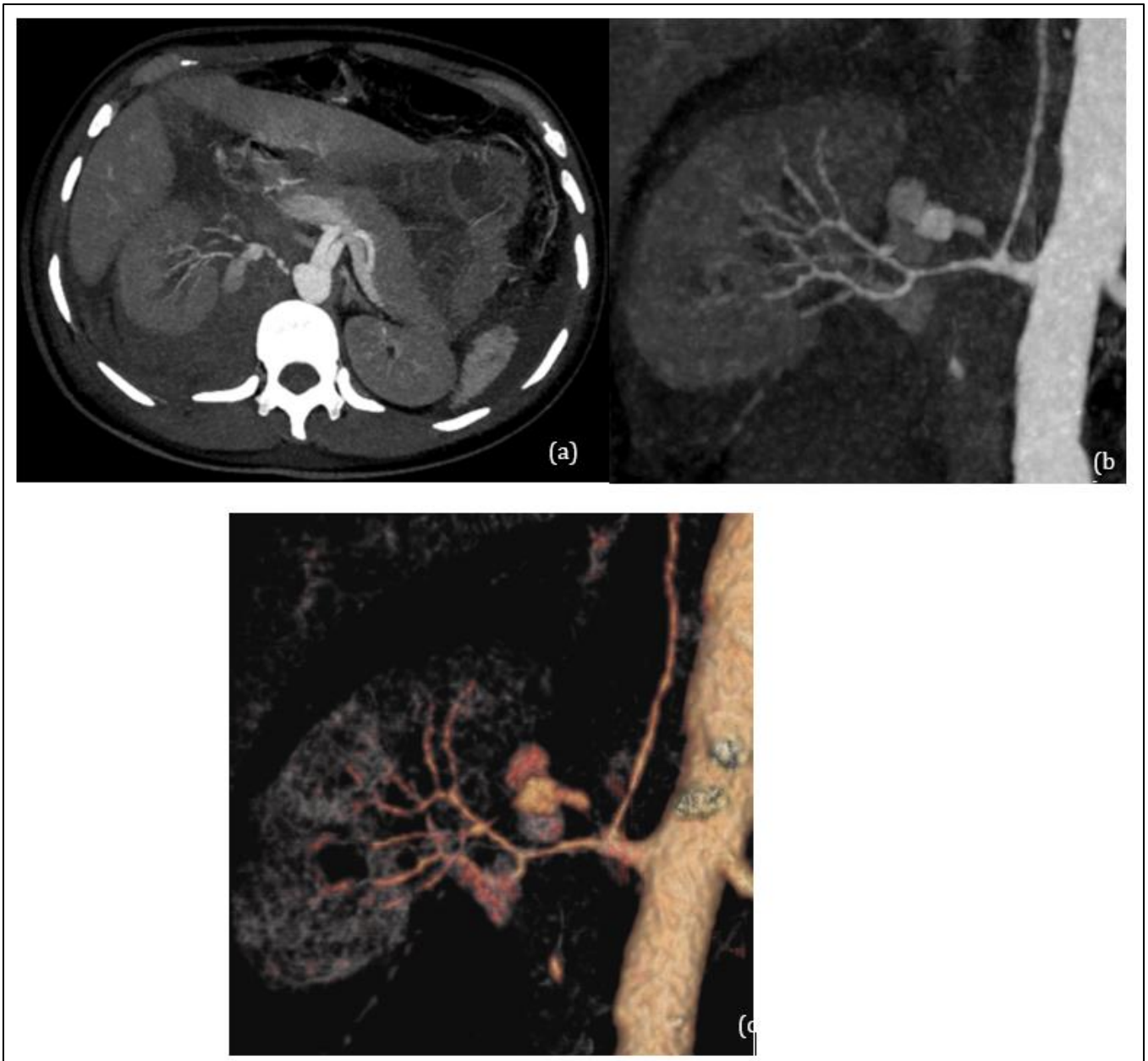


Figure 2 Abdominal angiography in axial (a), coronal (b), and 3D reconstructions (c), showing a large pseudoaneurysm of the posterior segmental branch of the right renal artery

The patient underwent arteriography, which showed a thin and irregular appearance of the posterior segmental branch of the right renal artery (Figure 3).

The right pseudoaneurysm visible on the abdominal angiography was not detected on the angiography performed 24 hours later.

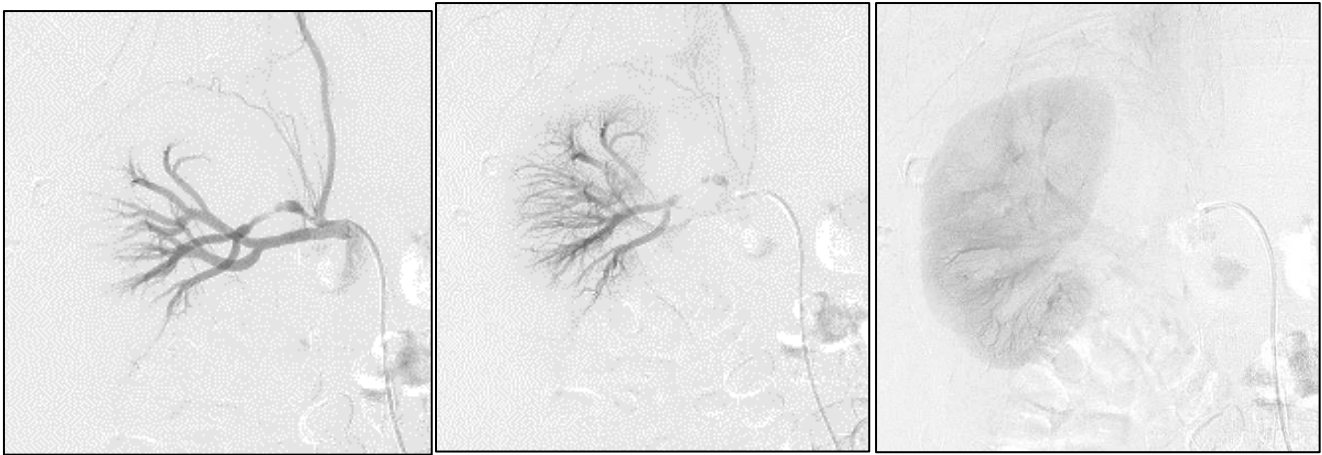


Figure 3 Arteriography of the right renal artery: thin and irregular appearance causing stenoses and dilatations of the posterior segmental branch of the right renal artery, without extravasation of iodinated contrast medium and without identification of a pseudoaneurysm

Three days later, the patient developed a generalized skin rash followed by skin detachment involving more than 90% of the body surface area and a positive Nikolsky sign. A systemic disease was suspected, and an immunological workup revealed positive antinuclear antibodies (ANA), negative anti-DNA antibodies, negative ANCA, and positive anti-ENA (extractable nuclear antigens) antibodies. The diagnosis of Lyell-like lupus was retained.

The clinical course rapidly worsened with a hemorrhagic syndrome due to thrombocytopenia at $20000/\mu\text{L}$, admission to the intensive care unit, and death despite resuscitation measures.

3. Discussion

Vasculitis associated with systemic lupus erythematosus (SLE) is a relatively rare but severe complication, with a prevalence ranging between 11 and 36% according to reported series, predominantly affecting small vessels and more rarely medium- and large-caliber vessels [1–3]. Visceral forms, although less frequent, are associated with increased morbidity and mortality [2,4].

Spontaneous retroperitoneal hematoma is an infrequent clinical entity, most often related to trauma, anticoagulant therapy, aneurysm rupture, or neoplastic disease. Its occurrence in the context of lupus vasculitis is exceptional and has been reported only as isolated cases or short series in the literature [5–7].

The proposed pathophysiological mechanisms include weakening of the vascular wall due to inflammation, fibrinoid necrosis, as well as hemostatic disorders frequently associated with SLE [3,8]. Several large series have highlighted the predominance of cutaneous involvement in lupus vasculitis, whereas visceral involvement mainly affects the peripheral nervous system, the gastrointestinal tract, and more rarely the muscular and retroperitoneal territories [1,4].

In the study by Drenkard et al., involving 540 patients with SLE followed over 10 years, visceral vasculitis accounted for only 12.3% of cases but was significantly associated with increased mortality [1]. The visceral manifestations reported mainly included multiple mononeuritis, digital necrosis, vasculitis of the large arteries of the limbs, as well as rare mesenteric and coronary involvement [1]. Although retroperitoneal hematoma was not explicitly described in this series, the presence of medium-caliber vessel vasculitis with necrosis and wall fragility constitutes a pathophysiological mechanism compatible with the occurrence of deep hemorrhagic complications, such as that observed in our case. Similarly, Ramos-Casals et al. showed that medium-vessel vasculitis was responsible for more severe forms requiring aggressive management [4].

The diagnosis of retroperitoneal hematoma is often delayed due to the nonspecific nature of the clinical signs, dominated by abdominal or lumbar pain, acute anemia, and sometimes hemodynamic instability. In this context, abdominal CT angiography is the reference examination [9–11]. It allows confirmation of the diagnosis, assessment of the extent of the hematoma, detection of signs of active bleeding, and guidance of the etiology by excluding traumatic, tumoral, or dysplastic vascular causes.

In patients with SLE, imaging also plays a key role in identifying indirect arguments in favor of vasculitis, such as vascular wall thickening, segmental abnormalities of medium-caliber vessels, or association with other signs of systemic activity [2,12]. MRI may be useful as a complementary tool for follow-up or when CT is contraindicated, particularly in cases of renal failure, although its role remains secondary in the acute setting due to lower spatial resolution [10].

Therapeutic management is mainly based on intensive immunosuppressive treatment, combining high-dose corticosteroids and, in severe forms, cyclophosphamide or another immunosuppressive agent [2,13]. Interventional or surgical treatment is reserved for situations of hemodynamic instability or persistent active bleeding. Several authors emphasize the value of early diagnosis based on imaging and immunological workup, which is decisive for prognosis [9,11].

Our observation falls within these rare cases of severe hemorrhagic lupus vasculitis. It highlights the crucial role of the radiologist in the rapid recognition of this complication, enabling appropriate multidisciplinary management and a favorable outcome.

4. Conclusion

Lupus vasculitis complicated by spontaneous retroperitoneal hematoma is an exceptionally rare but life-threatening condition. This observation underlines the major contribution of imaging, particularly CT angiography, in the early diagnosis of hemorrhagic vascular complications and in guiding therapeutic management. Prompt radiological recognition associated with multidisciplinary care may improve prognosis and help optimize the management of severe visceral forms of systemic lupus erythematosus.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest to be disclosed

Statement of informed consent

Informed consent was obtained from all individual participants included in the study

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