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CASE REPORTS

Wunderlich Syndrome in a Patient with Tuberous Sclerosis Complex Presenting with Hypovolemic Shock: Ruptured Renal Angiomyolipoma. A Case Report

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ABSTRACT

Background: Wunderlich syndrome is an uncommon but potentially fatal clinical entity defined by spontaneous, non-traumatic renal or perirenal hemorrhage. It is most frequently associated with renal angiomyolipoma, particularly in patients with tuberous sclerosis complex (TSC), where lesions are often bilateral, large, and highly vascularized, predisposing them to rupture.
Case Presentation: We report the case of a 30-year-old female with a known history of TSC who presented to the emergency department with acute right flank pain and signs of hypovolemic shock. Contrast-enhanced computed tomography (CECT) revealed a large right-sided perirenal and retroperitoneal hematoma with active contrast extravasation, originating from a ruptured renal angiomyolipoma. Multiple bilateral renal angiomyolipoma were also identified. The prompt use of advanced imaging facilitated rapid diagnosis and guided urgent management.
Discussion: This case underscores the pivotal role of CECT as the diagnostic modality of choice in suspected Wunderlich syndrome, allowing precise localization of bleeding, assessment of lesion characteristics, and evaluation of hemorrhage extent. Patients with TSC require close radiological surveillance due to the elevated risk of angiomyolipoma rupture and life-threatening hemorrhagic complications.
Conclusion: Wunderlich syndrome represents a rare but critical urological emergency, particularly in patients with TSC. Early recognition through contrast-enhanced imaging is essential for timely intervention and improved clinical outcomes. Proactive monitoring of renal angiomyolipomas in high-risk populations is imperative to prevent catastrophic events.
Keywords: Wunderlich syndrome, Renal angiomyolipoma; Retroperitoneal hemorrhage; Computed tomography;

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INTRODUCTION

Wunderlich syndrome refers to spontaneous, non-traumatic hemorrhage into the renal subcapsular or perirenal space and represents a rare but potentially fatal urologic emergency. (1,2)

Although uncommon in the general population, it's incidence is significantly increased in patients with tuberous sclerosis complex (TSC), primarily due to the presence of renal angiomyolipoma (AMLs). (3) Renal AMLs associated with TSC are frequently bilateral, large, and composed of abnormal vascular elements that predispose them to aneurysm formation and spontaneous rupture. (4,5) Massive hemorrhage may ensue, leading to hemodynamic instability and hypovolemic shock. (1,6) Early diagnosis is critical, as delayed recognition can result in severe morbidity or death. (3)

Contrast-enhanced computed tomography (CT) is the imaging modality of choice, allowing rapid identification of hemorrhage, assessment of its extent, and detection of the underlying etiology. (4,7)

We present a case of Wunderlich syndrome in a young woman with tuberous sclerosis complex, emphasizing the pivotal role of contrast-enhanced CT in diagnosis and management.

Case Presentation

A 30-year-old woman with a known diagnosis of tuberous sclerosis complex presented to the emergency department with sudden-onset severe right flank pain, accompanied by dizziness and syncopal episodes. On admission, the patient was hypotensive and tachycardic, consistent with hypovolemic shock.

Medical and Family History

The patient had been hospitalized three years earlier for a similar clinical presentation, at which time she was diagnosed with spontaneous retroperitoneal hemorrhage secondary to rupture of a large renal angiomyolipoma.

She remained clinically stable following discharge. Subsequent multidisciplinary evaluation confirmed the diagnosis of tuberous sclerosis complex. She had no history of seizures or cognitive impairment. Family history was significant for tuberous sclerosis complex in her mother.

Physical Examination

On physical examination, the patient appeared pale and diaphoretic, with pale mucosa. Multiple facial angiofibromas (Fig.1) were noted over the cheeks, perioral and nasal region.

Abdominal examination revealed marked right flank tenderness with a positive Giordano sign (costovertebral angle tenderness), suggestive of renal involvement. Neurological examination was unremarkable.

Vital signs revealed a heart rate of 120 beats per minute (sinus rhythm), temperature of 37 °C, and blood pressure of 85/55 mmHg. Laboratory evaluation demonstrated severe anemia (hemoglobin 7.6 g/dL, red blood cell count. 2.74 × 10^6/μL), white blood cell count. of 8.7 × 10^3/μL, creatinine level of 0.7 mg/dL, blood urea nitrogen of 11.6 mg/dL, C-reactive protein of 7 mg/dL, and lactate dehydrogenase of 928 U/L (Table 1).

Figure 1. Cutaneous Manifestation of Tuberous Sclerosis *(Facial angiofibroma involving the perioral and chin region, a characteristic cutaneous manifestation of tuberous sclerosis complex)*

Table 1. Laboratory values at presentation

Test (unit)	Observed values	Normal range
Hemoglobin (g/dL)	7.6	12.0-16.0
Red blood cell count ($\times 10^6$/μL)	2.74	3.9-5.2
White blood cell count ($\times 10^3$/μL)	8.7	4.0-10.0
Creatinine (mg/dL)	0.7	0.6-1.2
Blood urea nitrogen (mg/dL)	11.6	7-20
C-reactive protein (mg/dL)	7.0	<0.5
Lactate dehydrogenase (U/L)	928	140-280

g/dL = grams per deciliter; $\times 10^6$/μL = million cells per microliter; $\times 10^3$/μL = thousand cells per microliter; mg/dL = milligrams per deciliter; U/L = units per liter.

Imaging Findings

An urgent contrast-enhanced CT scan of the abdomen and pelvis was performed. Imaging demonstrated a large right-sided perirenal and retroperitoneal hematoma (Figures 1,2,3) arising from the right kidney, consistent with hemorrhage due to rupture of a renal angiomyolipoma. Diffuse peritoneal fat stranding and moderate free intraperitoneal fluid were also observed. Foci of active contrast extravasation were identified within the hematoma, indicating ongoing bleeding (Figure 3).

Figure 2. Contrast-enhanced abdominal CT (axial view) *[Large right perirenal and retroperitoneal hematoma (white arrow). Multiple renal angiomyolipomas (white asterisks) are identified as mixed fat- and soft-tissue-density lesions, causing distortion of the renal parenchyma. Associated peritoneal fat stranding (arrowhead) and free intraperitoneal fluid with high attenuation consistent with hemoperitoneum (black asterisk) are also noted.]*

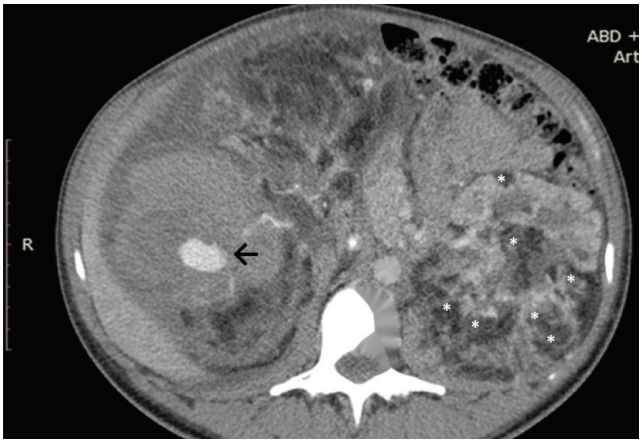

Figure 3. Contrast-enhanced abdominal CT (axial view). *[Hyperdense foci (black arrow) within the perirenal hematoma, showing active contrast extravasation consistent with ongoing hemorrhage. Multiple renal angiomyolipoma (asterisks).]*

Multiple large bilateral renal angiomyolipomas were present, appearing as heterogeneous lesions with mixed fat and soft-tissue density, resulting in distortion and partial loss of normal renal parenchymal architecture (Figure 4).

Figure 4. Contrast-enhanced abdominal CT (coronal view) *[The extent of the right perirenal hematoma (white arrows) and multiple renal angiomyolipomas with loss of normal renal architecture (asterisks).*

Comparison with prior imaging from 2021 revealed similar renal findings, supporting a chronic and progressive disease course. Brain magnetic resonance imaging performed in 2021 demonstrated multiple subcortical T2-hyperintense lesions consistent with cortical tubers (Figure 5), as well as multiple subependymal plaques (Figure 6), confirming the diagnosis of tuberous sclerosis complex.

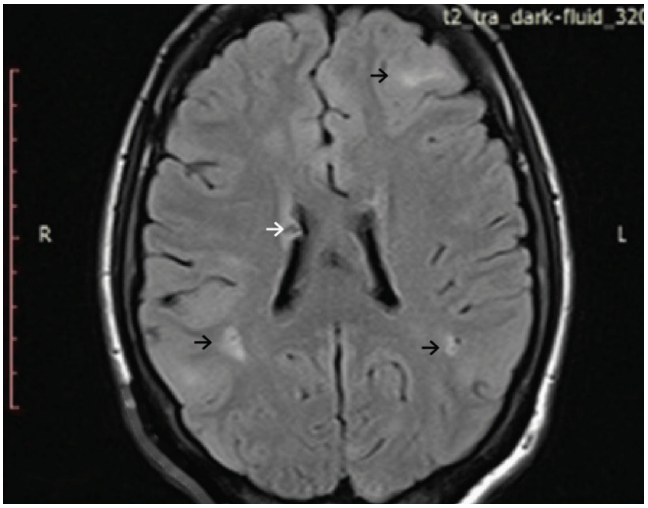

Figure 5. Brain MRI, T2-weighted axial image. *[Multiple subcortical T2-hyperintense lesions consistent with cortical tubers (black arrows), subependymal plaques lining the lateral ventricles (white arrow). Two pathognomonic CNS signs of tuberous sclerosis complex.]*

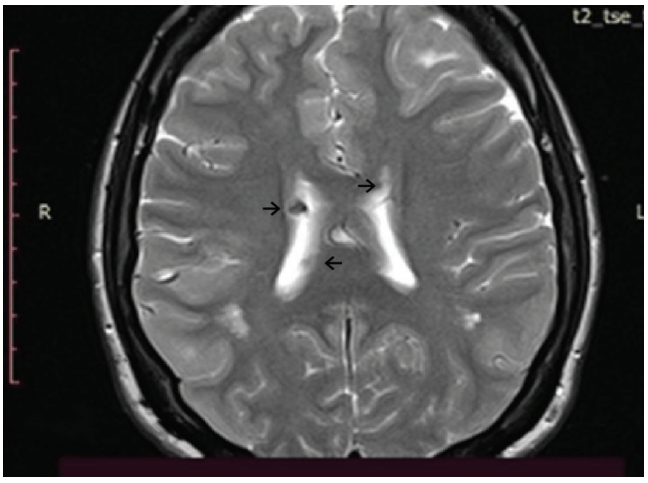

Figure 6. Brain MRI, T2-weighted axial image. *[Multiple subependymal plaques lining the lateral ventricles (black arrows).]*

DISCUSSION

Wunderlich syndrome is a rare but life-threatening complication, particularly in patients with tuberous sclerosis complex. (1,3) Renal angiomyolipomas associated with TSC are often large, bilateral, and highly vascular, significantly increasing the risk of spontaneous rupture and hemorrhage. (3,8) The classic clinical presentation includes acute flank pain, hypotension, and anemia, although the severity of symptoms depends on the extent of bleeding. (1,4)

Contrast-enhanced CT plays a central role in the diagnosis of Wunderlich syndrome, enabling rapid identification of hemorrhage, localization of the bleeding source, and evaluation of the underlying etiology (2,4). The detection of active contrast extravasation is particularly important, as it indicates ongoing hemorrhage and helps guide urgent therapeutic decision-making. (2,5)

Early imaging facilitates timely intervention, with selective arterial embolization currently considered the preferred minimally invasive treatment option, allowing effective hemorrhage control while preserving renal function. (4,7) Regular imaging surveillance is recommended in high-risk patients with tuberous sclerosis complex to prevent catastrophic hemorrhagic complications. (6,9)

CONCLUSIONS

Wunderlich syndrome represents a rare but potentially fatal emergency in patients with tuberous sclerosis complex. Prompt diagnosis using contrast-enhanced CT is essential for identifying the source and extent of hemorrhage and for guiding appropriate management. Regular monitoring of renal angiomyolipomas in high-risk patients is crucial to improving outcomes and preventing life-threatening complications.

Editorial Statements

Ethics Approval and Consent to Participate

This study involved the analysis of data from human participants. The research protocol was reviewed and approved by the Institutional Ethics Committee and was conducted in accordance with institutional regulations, the principles of the Declaration of Helsinki, and internationally accepted standards for biomedical research ethics.

Data Availability Statement

The datasets generated and/or analyzed during the current study are available from the corresponding author upon reasonable request.

Author Contributions (CRediT): Conceptualization & Methodology: AH; Data Curation, Investigation and Writing – Original Draft: ET, EM: Formal Analysis: EV, ML: Writing – Review & Editing: LL, ET: Supervision: AH. All authors have read and approved the final version of the manuscript.

Artificial Intelligence (AI) Statement

The authors used a generative artificial intelligence tool solely for language editing and grammar improvement.

All scientific content, data analysis, interpretation, and conclusions were independently developed, verified, and approved by the authors.

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