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Wunderlich syndrome: Rare and unrecognized emergency

Hamedoun Larbi [a,*], Ilias Hassan [a], Amine Cherraqi [b], Rachida saouab [b], Alami Mohammed [a], Ameur Ahmed [a]

[a] Urology Department, Military Hospital of Instruction Mohamed V, Hay Ryad, 10100, RABAT, Morocco
[b] Radiology Department, Military Hospital of Instruction Mohamed V, Hay Ryad, 10100, RABAT, Morocco

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ABSTRACT

A real life-threatening emergency, Wunderlich syndrome (WS) is an interesting and unknown clinical condition characterised by intense beginning of spontaneous, non-traumatic renal haemorrhage in the sub-capsular and perirenal space, with a typical clinical and radiological presentation [1] that allow the diagnosis.

Although most cases are treated invasively either by surgery or embolization to control the bleeding, Fortunately our 66 year old patient admitted to the Mohammed V military hospital in Rabat benefited from a conservative treatment, which allowed him to avoid all the associated complications and to be discharged from hospital in less than 4 days.

1. Introduction

Wunderlich syndrome is an unusual entity characterized by acute abdominal pain caused by massive non-traumatic renal haemorrhage into the sub-capsular and peri-renal spaces. It tends to be lethal while perhaps not instantly perceived clinically and treated appropriately. The most often related etiologies incorporate benign and malignant renal tumors, for example renal angiomyolipoma and renal cell carcinoma, vascular diseases and renal infection. Kidney cysts, blood dyscrasias or anticoagulant and antiplatelet therapy are a portion of the more uncommon causes. We report the case of a 66-year-old patient who presented to the emergency department with acute abdominal pain mainly localized in the left flank associated with hypotension.

2. Case presentation

A 66-year-old man presented to the Emergency Department with acute abdominal pain since 48h.

He was a chronic smoker and with a medical history of high blood pressure, type 2 diabetes, chronic renal failure with haemodialysis and ischemic heart disease treated by stenting + Antiplatelet agents 10 days before.

The pain was severe, generalized in the whole abdomen, constant and pulsating in the left flank; it woke him from sleep. On physical examination the patient was afebrile, had heart rate 120 beats per minute and blood pressure 85/54 mm Hg, room air saturation 100%, and respiratory rate was 26 breaths/min. Systemic examination of the abdomen was soft with tenderness over the left lower quadrant, with a palpable mass over the renal angle. The patient's haemoglobin was 8.3 g/dl, white blood cells WBC 9100/µl, platelet count was normal 319000/µl (Fig. 1), creatinine 73 mg/dl, estimated glomerular filtration rate 8 ml/min/1.73 m^2 and Protein C-Reactive 84,8 mg/l. A cyto-bacteriological examination of the urine was sterile, confirmed by a culture, which showed no pathogen growth. Abdominal computed tomography scan with contrast agent was then performed and showed presence of a spontaneously hyper dense peri-renal, retroperitoneal collection of hematic density fusing downwards along the psoas muscle and the opposite contralateral kidney associated with fat infiltration suggestive of Wunderlich Syndrome (Fig. 2). The remainder of urinary tract was without anomaly.

Firstly, the patient had to be conditioned: empiric antibiotics and hydration were given intravenously adapted to his renal situation, a transfusion of 03 red blood cells per dialysis. A cardiologist's opinion was requested to reduce as much as possible the dose of anti-platelet agent without compromising his cardiac function. The patient remained hemodynamically and clinically stable which motivated us to manage him conservatively and discharged 03 days later. An abdominal scan was performed one month later, which showed Volume regression of retroperitoneal hematoma (Fig. 3). On half-year follow-up he had close total goal of the hematoma. Our patient had no other lesions

* Corresponding author.
E-mail addresses: larbi771@gmail.com (H. Larbi), ilyayanleh16@gmail.com (I. Hassan), Dr.amine.cherraqi@gmail.com (A. Cherraqi), rachida_sa@yahoo.fr (R. saouab), alamim1965@gmail.com (A. Mohammed), Ahmed.Ameur@yahoo.fr (A. Ahmed).

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Complete Blood Count		
	Value of our patient	Reference value
White Blood Cells	9,1*10^3/µl	4,0-10,0
Polynuclear Neutrophils	7,1*10^3/µl	1,5-7,5
Lymphocytes	0,9*10^3/µl	1,5-4,0
Monocytes	0,9*10^3/µl	0,1-0,8
polynucleaire eosinophiles	0,2*10^3/µl	0,0-0,5
Polynuclear Basophils	0,1*10^3/µl	0,0-0,1
Red Blood Cells	2,74*10^6/µl	3,90-5,50
Haemoglobin	8,3 g/dl	13,0-17,0
Hematocrit	24,3%	41,0-53,0
Mean Corpuscular Volume	88,6 fl	82,0-98,0
Mean Corpuscular Hemoglobin Content	30,3 pg	27,0-33,0
Mean Corpuscular Hemoglobin Content	34,2g/dl	32,0-36,0
Cell Distribution Index	15,1	-
Platelet	312*10^3/µl	150--450
Mean Platelet Volume	8,2 fl	-
Erythroblast	0,0%	-

Fig. 1. Complete blood count proving blood loss.

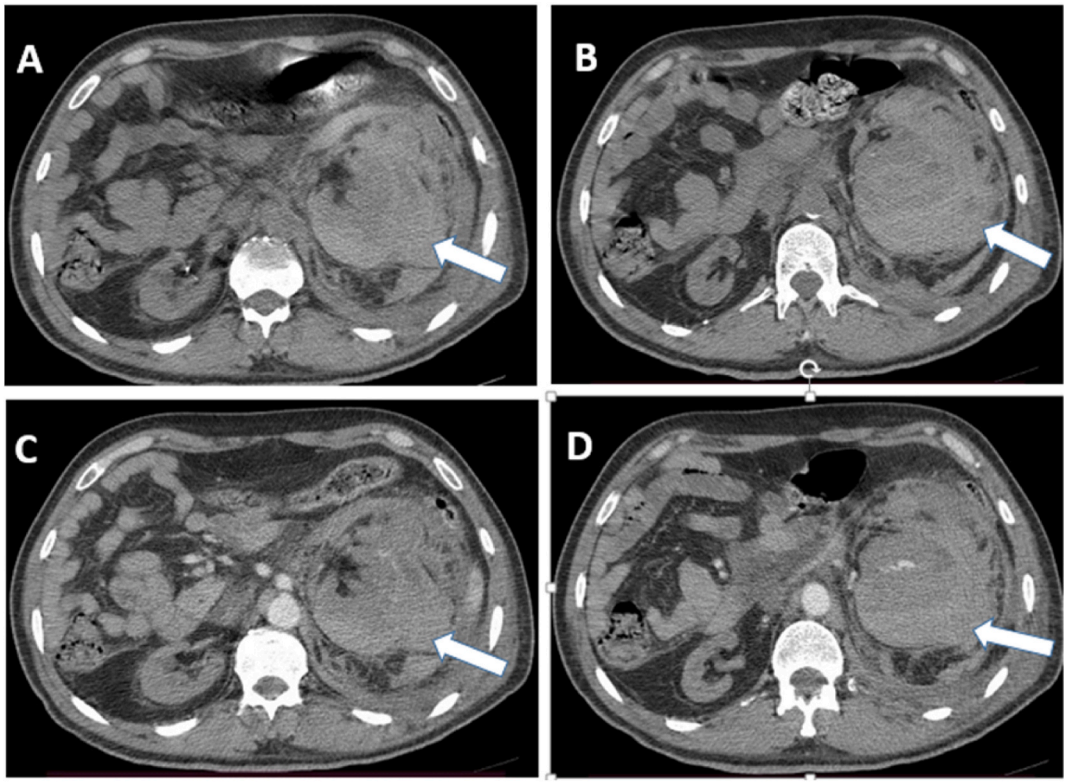

Fig. 2. Abdominal CT scan showing presence of a spontaneously hyper dense perirenal, retroperitoneal collection of hematic density. (A and B) with spontaneous contrast: C-. (C and D) after contrast injection: C+.

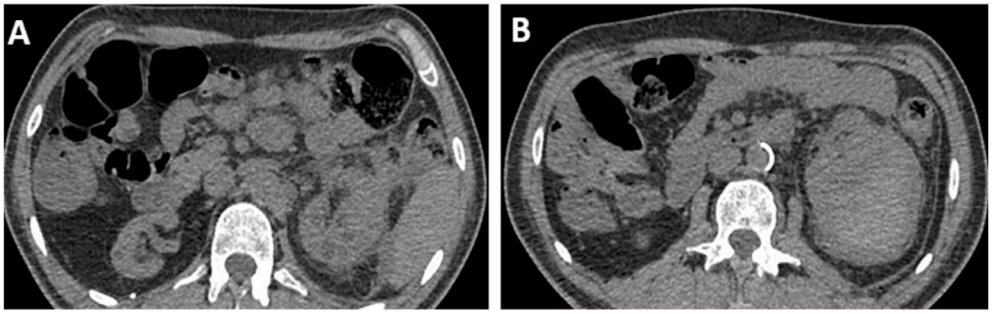

Fig. 3. (A et B): Abdominal CT scan showing regression of the retroperitoneal hematoma volume performed one month later.

associated with malignancy.

3. Discussion

Spontaneous renal hematoma was first described in 1700 by Bonet. Wunderlich gave his name to the syndrome in 1856.[1] Wunderlich syndrome, a rare but life-threatening entity, may affect 1–3% of long-term dialysis patients, which may be related to the anticoagulation received during dialysis.[2] It manifests as the classic Lenk triad[3]: sudden onset of low back pain, signs of collapse and lumbar mass. Our patient is an illustrative example of this clinical presentation.

It is important to know that early recognition and diagnosis of Wunderlish syndrome allows for proper management, which significantly reduces the mortality rate.

Angiomyolipoma is that the commonest benign neoplasm liable for WS, while renal cell carcinoma is that the commonest malignant tumor. It's frequently found in conjunction with hypertension (33–50%) and atherosclerosis (80–87%) or during treatment with anticoagulants or platelet aggregation inhibitors.[4] In some cases, no causes are often identified 5% (especially in haemodialysis patients).[3]

The more or less typical clinical presentations as well as the initial conditioning of the patient are the same whatever the aetiology, even in chronic haemodialysis patients. Afterwards, the decision to carry out a radical or conservative treatment will depend on the aetiology, especially tumour or persistent haemodynamic instability of the patient, which require an emergency nephrectomy.

Our patient had no associated lesions of the Urinary tract but had several predisposing conditions like haemodialysis, hypertension, diabetes and anti-platelet therapy all contributing to vascular fragility.

In case of clinical suspicion of WS, imaging aims to confirm the diagnosis by identifying renal haemorrhage in the subcapsular and/or perilesional space but also to detect the cause of bleeding when possible. Abdominal computed tomography scan may be a good choice of imaging modalities, because it's 100% sensitive to demonstrate the presence of perirenal and retroperitoneal haemorrhage,[3] which is characteristic of Wunderlich's syndrome the rationale why it had been requested in the first place for our patient. Ultrasound may be of interest in the follow-up assessment to demonstrate the resumption of the hematoma.

Management of WS is especially expectant, although endovascular or surgical interventions could also be required in cases of massive haemorrhage with persistent hemodynamic instability or suspicion of malignancy.[5] An early or immediate exploratory surgery with possible total or partial nephrectomy may be necessary if hemodynamic instability persists. Others prefer renal arteriography with embolization as a crucial diagnostic and therapeutic method.[4] Although there aren't yet evidence-based guidelines to favour either approach.[5]

However, as illustrated by our case and like many cases already reported in the literature, for patients in whom no underlying malignancy is identified on initial Abdominal computed tomography scan and hemodynamically stable, conservative management with follow-up imaging may be a reasonable option and may save the patient an unnecessary surgery.

4. Conclusion

Wunderlich's Syndrome is an emergency that ought to be immediately diagnosed and treated, with minimally invasive embolization, surgical exploration or conservative management with follow-up imaging as performed in our case. It is safer to keep this differential diagnosis in mind to evoke it in patients presenting with unexplained abdominal pain to emergency department.

Consent

Consent from the patient was obtained.

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Declaration of competing interest

None.

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