Wünderlich Syndrome: A Rare Syndrome Caused by a Polyarteritis Nodosa

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Abstract

Wünderlich syndrome (WS) is a rare condition, in which spontaneous nontraumatic renal hemorrhage occurs into the subcapsular and perirenal spaces and was described for the first time by Bonet in 1700. The etiology of the entity is very diverse, including benign or malignant neoplasms, vascular diseases, infectious and inflammatory kidney diseases, and could be combined with some diseases. Clinically the syndrome is characterized by the classic Lenk’s triad. We describe a case of WS in a 37-years-old woman with loss of strength in the right superior limb for three months. After admission in the hospital her physical examination showed up pallor, tachycardia and evolved with diffuse abdominal pain. Following all medical conducts the patient do not resist.

Keywords

Wünderlich syndrome, Retroperitoneal hemorrhage, Computed tomography

Introduction

Many causes of retroperitoneal hemorrhage have been described. The origin may be caused by any retroperitoneal structure (renal, vascular, pâncreas, adrenal, etc.). Spontaneous retroperitoneal hemorrhage (SRH) of non-traumatic originated from renal parenchymal rupture was first described by Bonet in 1700. And in 1856 Carl Wünderlich made the clinical description. The causes vary a lot: benign or malignant tumors, vascular diseases, inflammatory process, cystic rupture, infectious. These illness can lead to a retroperitoneal hemorrhage or bleeding into urinary tract, which could be a life-threatening. The symptoms may start rapid or stealthy. It is classically associated with symptoms and signals named by Lenk’s triad. The appropriate treatment depends the accurate diagnoses made by radiological imaging and clinical information.

Case Report

A 37-years-old single woman who presented to emergency department with loss of strength in the right superior limb for three months and associated with loss of sensitivity on feet and legs. She was admitted to the hospital for investigation. In her physical examination was pallor, tachycardia and evolved with diffuse abdominal pain. Following all medical conducts the patient do not resist. The computed tomography showed following images (Figure 1, Figure 2 and Figure 3).

Discussion

Carl Reinhold August Wunderlich, a German physician, in 1856, made the clinical description of SRH of non-traumatic originated from renal parenchymal rupture called by ‘spontaneous renal capsule apoplexy’ [1,2].
The most common causes of the WS are renal neo-
plasms. Angiomyolipomas a mesenchymal neoplasm
composed by adipose tissue, smooth muscle and dys-
morphic blood vessels is the most common cause of the
WS. Renal cell carcinoma is in second place and is in first
between the malignant tumors that causes WS [3-5,9-
13].

Vasculitis has different types and have their classifi-
cation according to the size and type of vessels involved
and the existence or the lack of associated fibrinoid ne-
crosis and/or granulomas [14].

Table 1: Causes of spontaneous retroperitoneal hemorrhage.

| Tumors               | Vascular                      |
|----------------------|-------------------------------|
| Angiomyolipoma       | Renal artery aneurysm         |
| Renal cell carcinoma | Renal artery rupture          |
| Pheochromocytoma     | Thrombosis                    |
| Myeloma              | Renal infarction              |
| Adenoma              | Abdominal aortic aneurysm ruptura |
| Hemangiomia          | Aretrovenous fistula/malformation |
| Metastasis           | Polyarteritis nodosa          |
| Carcinoma            | Behçet syndrome              |
| Retroperitoneal tumors |                             |
| Pancreas             |                               |

Table 2: Causes of spontaneous retroperitoneal hemorrhage

| Systemic                           | Others                        |
|------------------------------------|-------------------------------|
| Thrombocytopenic purpura           | Renal cystic rupture          |
| Leukemia                           | Renal calculi                 |
| Hemophilia                         | Nephrosclerosis               |
| Polycythemia                       | Pyelonephritis                |
| Anticoagulant therapy              | Renal tuberculosis            |
|                                    | Renal cortical abscesso       |
|                                    | Nephritis                     |

Spontaneous retroperitoneal hemorrhage may
have several origins as well as low back pain. The
causes could be tumoral, non-tumoral, vascular, in-
flammatory or systemic (some of these are exposes
on Table 1 and Table 2) and [1-11].
Polyarteritis nodosa (PAN) is a rare form of primary systemic vasculitis with heterogeneous presentations. PAN is an autoimmune disease characterized by necrotizing arteritis of small- and medium-sized muscular arteries associated with aneurysmal nodules, condition first described by Adolph Kussmaul and Rudolph Maier in 1866, and could affect multiple organs [15-19].

Most cases of PAN, primary cases, are idiopathic. But several subgroups have already been defined by their aetiologic association, secondary PAN. It is often notice in hepatitis B and hepatitis C infections, malignancies such as hairy cell leukemia, familial mediterranean fever (FMF), connective tissue disease related and deficiency of adenosine deaminase [20-26].

PAN is not associate with antineutrophil cytoplasmic antibodies (ANCA) [20,21,27]. No specific serologic diagnostic tests have been developed.

Renal involvement is common. Some authors plead approximately 60% of patients with PAN have renal involvement. Rarely complicated by acute kidney injury (AKI) in patients with renal involvement, but has high morbidity. PAN is rare in patients with acute abdomen, on the other hand, acute abdomen is frequent in cases of PAN [4,15-19].

The American College of Rheumatology proposed classification criteria for PAN in 1990 [28].

The diagnosis is made by histology in most cases. Imaging studies in vasculitis’s are important to determine disease extension and activity. MRI, CT, and CT angiography are the noninvasive techniques to delineate vasculitis lesions.

The clinical picture vary hugely depending on the degree of the bleeding and duration. Acute lumbo-abdominal pain, nausea, vomiting, hematuria, hemodynamic instability, anemia and hypovolemic shock may be present [1]. SRH is clinically characterized by the Lenk’s triad: Acute flank pain, hemodynamic compromise and flank palpable mass.

WS is not common but is life-threatening entity. SRH is a diagnostically challenging condition and need to be management rapidly and radiological imaging plays an important role in establishing SRH and causes.

Confirmation of the diagnosis by identifying renal hemorrhage in the subcapsular and/or perirenal space, detecting the possible cause and identify any underlying pathological process that may result in a predisposition to hemorrhage for the bleeding is the aim of the imaging in a suspect case of WS [4,29].

Ultrasonograpy may be the first tool to detect perinephric hematoma, but it has been consider to be only modest useful in identifying renal hemorrhage and in differentiating the renal mass and clotted blood, therefore identification of etiology may not be possible. To document the evolution of hematoma a follow-up with ultrasound is advantageous, because it’s cheap and accessible. Color and spectral Doppler may help in the detection of some vascular and non-vascular pathologies responsible for WS. On US, acute hematoma appears as an isoechoic to hypechoic collection; subacute and chronic hematomas appear as hypechoic to anechoic [3,4,12].

Computed tomography (CT) with multiplanar display of datasets allows high-resolution imaging of the renal parenchyma, vasculature, and the collecting system, is the investigative modality of choice in diagnosing WS, its etiology, related complications and severity [1,9,11,30,31]. To demonstration perirenal hemorrhage CT is the method of choice (sensitivity of 100%) but has sensitivity about 57% in identifying renal neoplasm when performed at the time of hemorrhage and may miss rare cases of segmental arterial medio lysis [1,5,9,12,30] Sequential dynamic contrast image sets should be obtained as corticomedullary, nephrographic and excretory phase images [4]. When CT give few information’s it has been suggested that angiography should be done as it can reveal vascular lesions not shown by CT. Renal angiography helps in diagnosing vasculitis, the most common vascular cause of perinephric hemorrhage [5,11,31,32]. If angiography and CT fail because of distortion of renal architecture caused by multiple renal cysts and hypovascularity of some tumors [32], CT could be repeat. Large-volume hematomas could mask hidden neoplasms or the cause of perirenal bleeding remains unclear, therefore follow-up CT is advocate [15,33]. Renal arteriography with selective embolization locates the bleeding site and allows bleeding control avoiding surgery.

If CT failed in initial examination or contrast-enhanced CT is contraindicated, a magnetic resonance (MR) may be performed to reveal the cause of bleeding [3,4,30].

Treatment options depend of the aetiology, the amount of bleeding and how fast the diagnostic are done. Some cases could be follow closely. Minimally invasive procedures as artery embolization is another option. Although, sometimes, nephrectomy could be required.

WS could be a life threatening, and determining its etiology of is often a challenge. Nevertheless, when Lenk’s triad is present the patient should be managed aggressively. Accurate diagnosis order a combination of clinical and radiological imaging information’s. All imaging radiological methods have their role. CT, angiography and MR perform an important function in the diagnosis and determine the cause of the syndrome.

Acknowledgement

Funding: None.
Conflict of Interest: None.

All the authors made substantial contribution to the preparation of this manuscript and approved the final version for submission.

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