DOI: https://doi.org/10.2298/SARH210410089A
UDC: 616.61-005.1

CASE REPORT / ПРИКАЗ БОЛЕСНИКА

A rare case of spontaneous perirenal hemorrhage – Wunderlich syndrome

Goran Aranđelović, Stefano Lai, Claudio Milani
St. John and Paul Hospital, Department of Urology, Venice, Italy

SUMMARY
Introduction Spontaneous perirenal hemorrhage or Wunderlich syndrome represents a rare entity in urological settings. The vast majority of the causes are represented by angiomyolipoma and renal cell carcinoma. In other cases, vascular abnormalities, polycystic kidneys, polyarthritis nodosa, or pyelonephritis could represent the cause of perirenal bleeding. The treatment depends on the clinical parameters at the presentation as well as on the possible presence of renal malignancies.
Our goal was to present a rare case of a healthy men who presented the idiopathic Wunderlich syndrome.
Case outline We present a case of a 50-year-old patient with spontaneous perirenal hemorrhage that was not due to any defined cause even after a six-months follow-up.
Conclusion In some rare cases of perirenal bleeding, the cause cannot be defined at the presentation, even with today's advanced radiological imaging methods. Thus, it is important to be aware of the fact that in those cases, a longer follow-up is needed, knowing that the presence of perirenal hematoma can always obscure the real diagnosis. Sometimes, even in cases where the proper follow-up has been done, the real cause of the bleeding remains unknown.
Keywords: Wunderlich syndrome; perirenal hematoma; angiomyolipoma

INTRODUCTION

Wunderlich [1] was the first who presented the spontaneous hematoma of the kidney in 1856. It refers to spontaneous non-traumatic renal bleeding into subcapsular and/or perirenal space. This condition may be caused by various pathologies, such as benign and malignant renal tumors, renal artery aneurysms, polyarteritis nodosa, polycystic kidneys, renal infections, or undiagnosed hematological conditions [2]. Some of the possible causes with their respective percentages are given in Table 1 [2, 3].

Table 1. Possible causes of Wunderlich's syndrome in percentage

Wunderlich's syndrome causes	%
AML	23
RCC	19
ACKD	8
Simple renal cyst	8
Sarcoma	4
Hematoma or hemorrhage only	38

AML – angiomyolipoma; RCC – renal cell carcinoma; ACKD – acquired cystic kidney disease

The treatment depends mainly on patient conditions and the determination of the cause of the hemorrhage. Since misdiagnosis is an emerging topic in modern medicine and there are disciplines that confirm an increasing alert on the risks of an omitted diagnosis or the consequences of incorrect treatment, we consider it important to present a rare case of spontaneous perirenal hematoma which was not due to any known or diagnosed cause [4, 5].

CASE REPORT

A 50-year-old male patient was admitted to the emergency room with acute abdominal and right flank pain and painful sensitivity to palpation. The patient did not report any history of trauma or drug use and the anamnesis did not reveal other illnesses. During clinical examination, the blood pressure of 120/80 mmHg and the heart rate of 82 beats per minute were recorded. The hemoglobin value was 13.8 g/dl, leukocytes 15,000/mm^3, platelet count amounted to 270 × 10^9/L, creatinine clearance was 0.88 mg/dl, coagulation parameters as prothrombin time, activated partial thromboplastin time, and international normalized ratio were in their respective normal ranges. Urine sediment showed only proteinuria. Ultrasound examination followed by computed tomography (CT) scan of the abdomen showed a very large right perirenal hematoma without showing a mass responsible for the hemorrhage. The patient was symptomatic and the flank pain was at that moment not very responsive to conservative therapy. An urgent arteriography was performed showing no acute vascular bleeding sites so that there was no need for arterial embolization. The patient was treated with intravenous antibiotic therapy and again with intravenous pain medications, this time with success. On the next day hemoglobin values decreased from 13.8 g/dl to 8.8 g/dl, so that blood transfusion was needed. Two units of blood were transfused so that the hemoglobin values increased and remained stable (11 g/dl)

Received • Примљено: April 10, 2021
Revised • Ревизија: October 9, 2021
Accepted • Прихваћено: October 14, 2021
Online first: November 4, 2021

Correspondence to:
Goran ARANĐELOVIĆ
Castello 6777
30122 Venice, Italy
gorana_4@yahoo.it

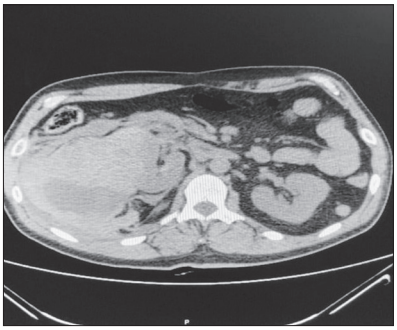

Figure 1. Abdominal computed tomography scan of the right retroperitoneal bleeding at the presentation

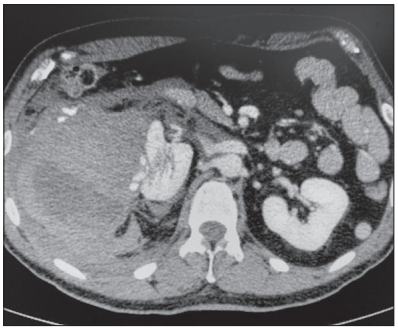

Figure 2. Abdominal computed tomography scan two days after the acute event

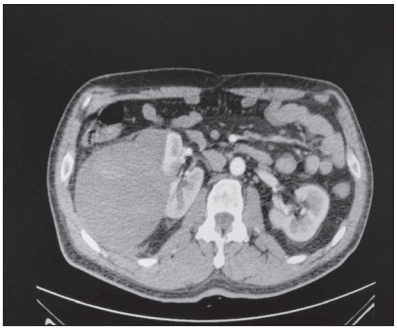

Figure 3. Computed tomography angiogram one month after the acute event

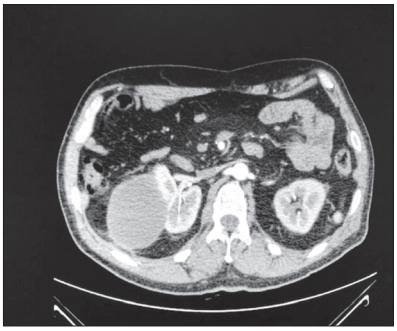

Figure 4. Computed tomography angiogram three months after the acute bleeding

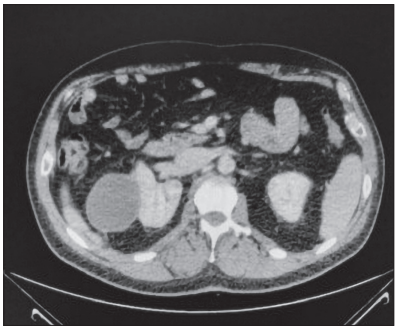

Figure 5. Computed tomography angiogram at six months follow-up

one day later. Two days after the acute event, the control CT scan showed a small increase of subcapsular hematoma, still without any sign of acute bleeding. After the close follow-up, the patient was dismissed asymptomatic and with stable hemoglobin levels on day 8 after the acute episode.

The control angio-CT scan after one, three, and six months from the acute event showed the important reduction of the perirenal hematoma without apparent cause for the previous bleeding.

Figures 1–5 show the CT scan of the right perirenal hematoma from the first presentation to the six-month follow-up: Figure 1 shows the bleeding at the presentation, Figure 2 shows CT scan two days after the acute event, Figure 3 shows the control angio-CT scan at 1 month after the acute event, Figure 4 shows the control angio-CT scan at three months after the acute bleeding and Figure 5 shows the control angio-CT scan at six months' follow-up.

All procedures involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. Written consent to publish all shown material was obtained from the patient.

DISCUSSION

Spontaneous renal bleeding to the subcapsular and perinephric space, known as Wunderlich syndrome (WS), could occur due to benign or malignant renal tumors, vascular lesions such as polyarteritis nodosa, anatomical lesions as renal cysts, renal infections, or nephritis [2]. Between 2003 and 2011 just 250 cases have been reported [6, 7]. Renal neoplasms are the most common cause of WS, accounting for 60–65% of all cases in which renal angiomyolipoma is the most common benign neoplasm responsible for WS [5, 7]. Renal cell carcinoma could also be the cause of WS, but in only 0.3–1.4% of cases [7]. This potentially life-threatening condition which could be associated with hypovolemic shock is also followed by other symptoms, such as acute lumboabdominal pain and palpable abdominal mass, symptoms forming the so-called Lenk's triad [2]. Although clinical guidelines for managing WS are not yet well established, the appropriate treatment for patients with WS depends on the right diagnosis that a bleeding has occurred, in which contrast-enhanced CT scan, as a method of choice with a sensitivity of 100%, has the most important role [7]. Although successful in the diagnosis of perirenal hemorrhage, CT scan has much lower sensitivity in identifying the renal neoplasm causing hemorrhage [8, 9].

If initial CT scan shows no mass responsible for the bleeding, angiography should be performed in order to reveal eventual vascular lesions and to perform embolization [10]. This could be very helpful especially in some rare cases such as segmental arterial mediolysis, as reported by Skeik et al. [11].

Thus, angiography and arterial embolization represent an important tool in diagnosis and therapy of spontaneous retroperitoneal hemorrhage. In some cases, as in the case of spontaneous retroperitoneal bleeding due to metastatic testicular germ tumor, as reported by Luis Eduardo et al. [12], it is not possible to dominate the bleeding just by using embolization. In those cases, an exploratory laparotomy with or without partial nephrectomy is needed [12]. A surgical intervention is necessary every time the hypovolemic shock caused by acute bleeding cannot be dominated by arterial embolization.

Thus, the main problem in WS is defining the source of bleeding in order to postulate the correct therapy.

In our case, it was not possible, in the acute phase, to define the cause of bleeding. Fortunately, the bleeding limited itself spontaneously and the patient was treated conservatively. This did not ensure that the bleeding source was not still present. The literature reports that if a CT scan, followed by angiography, does not reveal the bleeding source, a CT scan should be repeated later, as it is obvious that if the hemorrhage is massive, a possible renal cell carcinoma, angiomyolipoma, or other renal bleeding sites, such as renal cysts, could be seen just after the resorption of the hematoma [13]. That has also been proven, not just in cases of renal angiomyolipoma or clear cell carcinoma, but also in rare cases of renal sarcoma presenting WS [14]. Thus, we performed a CT scan at one, three, and six months after the acute phase. As seen from the presented figures, we were not able to elucidate the real cause of bleeding even six months after the acute event.

It is worth mentioning that hematologic issues can contribute to WS. As reported, some patients with end-stage renal disease are predisposed to bleeding diathesis in the setting of uremic platelet dysfunction, anemia, irregularities in von Willebrand factor, and impaired platelet – vessel wall interaction [15, 16].

All those factors were excluded in our case, given the young age, complete negative anamnesis and normal laboratory findings of the patient.

The patient was treated conservatively, which corresponds to previous findings on WS, stating that if the patient is hemodynamically stable in the acute phase, nephrectomy or partial nephrectomy should be deferred. A recent Korean study of 28 patients with WS stated that the definitive treatment of WS will depend on the clinical condition and the underlying cause, with possible therapeutic options including conservative therapy, angioembolization, nephron-sparing surgery, or radical nephrectomy [3, 17]. More interestingly, they found that five of 28 patients had no obvious cause of perirenal bleeding. This was also the case with our patient, given that nephrectomy or partial nephrectomy were not needed even later, as no malignant pathology could be observed.

In conclusion we can say that, although a vast majority of WS cases are represented by angiomyolipoma or by renal cell carcinoma, the cause sometimes remains unknown. In the present report we described a rare case of idiopathic WS whose cause could not be diagnosed even after six months of follow-up.

Conflict of interest: None declared.

REFERENCES

1. Wuendrlich RA. Handbuch der Pathologie und Therapie. Second edition. Stuttgart, Germany: Ebner and Seubert; 1856.
2. Albi G, Del Campo L, Taggaro D. Wunderlich's syndrome: Causes, diagnosis and radiological management. Clin Rad. 2002;57(9):840–5.
3. Kim JW, Kim JY, Ahn ST, Park TY, Oh MM, Moon DG, et al. Spontaneous perirenal hemorrhage (Wunderlich syndrome): An analysis of 28 cases. Am J Emerg Med. 2019;37(1):45–7.
4. Ferrara SD, Bajanowski T, Cecchi R, Boscolo-Berto R, Viel G. Bio-medicolegal scientific research in Europe: a comprehensive bibliometric overview. Int J Legal Med. 2011;125(3):393–402.
5. Viel G, Boscolo-Berto R, Cecchi R, Bajanowski T, Vieira ND, Ferrara SD. Bio-medicolegal scientific research in Europe. A country-based analysis. Int J Legal Med. 2011;125(5):717–25.
6. Zhang JQ, Fielding JR, Zou KH. Etiology of spontaneous perirenal hemorrhage: a meta-analysis. J Urol. 2002;167(4):1593–6.
7. Blakeley CJ, Thiagalingham N. Spontaneous retroperitoneal haemorrhage from a renal cyst: an unusual cause of hemorrhagic shock. Emerg Med J. 2003;20(4):388.
8. Katabathina VS, Katre R, Prasad SR, Surabhi VR, Shanbhogue AK, Sunnapwar A. Wunderlich syndrome: cross-sectional imaging review. J Comput Assist Tomogr. 2011;35(4):425–33.
9. Simkins A, Maiti A, Cherian SV. Wunderlich Syndrome. Am J Med. 2017;130(5):e217–e218.
10. Ho TH, Yang FC, Cheng KC. Wunderlich syndrome, spontaneous ruptured renal angiomyolipoma & tuberous sclerosis. QJM. 2019;112(4):283–4.
11. Skeik N, Olson SL, Hari G, Pavia ML. Segmental arterial mediolysis (SAM): Systematic review and analysis of 143 cases. Vasc Med. 2019;24(6):549–63.
12. Luis Eduardo SS, Arturo Enrique MG, Joel JÁ, Andrés MC, Laura Denisse RG. Bilateral wunderlich syndrome secondary to synchronous bilateral testicular germ cell tumor. A case report. Urol Case Rep. 2019;28:101028.
13. Parmar N, Langdon J, Kaliannan K, Mathur M, Guo Y, Mahalingam S. Wunderlich Syndrome: Wonder What It Is. Curr Probl Diagn Radiol. 2021;S0363-0188(21)00016-5.
14. Manikandan R, Mehra K, Dorairajan LN, Nachiappa Ganesh R, Sreenivasan SK, Kumar R. Case Report: Partial nephrectomy in primary renal sarcoma presenting as Wunderlich syndrome; a rare tumour with rare presentation managed atypically. F1000Res. 2019;8:423.
15. Chamarthi G, Koratala A. Wunderlich syndrome. Clin Case Rep. 2018;6(9):1901–2.
16. Swaminathan N, Sedhom R, Shahzad A, Azmaiparashvili Z. Post-partum occurrence of Wunderlich syndrome and microangiopathic haemolytic anaemia (MAHA): a case report. J Community Hosp Intern Med Perspect. 2021;11(2):277–9.
17. Choi HS, Kim CS, Ma SK, Kim SW, Bae EH. Wunderlich syndrome and regression of angiomyolipoma. Korean J Intern Med. 2020;35(6):1528–9.

Редак случај спонтане перирenалне хеморагије – Вундерлихов синдром

Горан Аранђеловић, Стефано Лаи, Клаудио Милани

Болница „Свети Јован и Павле", Клиника за урологију, Венеција, Италија

САЖЕТАК

Увод Спонтана перирenална хеморагија или Вундерлихов синдром представља редак ентитет у урологији. У највећем броју случајева узрок овог феномена представљају ангиомиолипоми и карциноми бубрега. Мање заступљени су узроци васкуларне природе, полицистични бубрези, нодозни полиартритис, пијелонефритис или идиопатски фактори. Третман ових болесника зависи од клиничких параметара на пријему, као и од присуства евентуалних малигних бубрежних обољења.

Наш циљ је био представити редак случај релативно младог болесника са Вундерлиховим синдромом који није био узрокован ниједном до данас знаних патологија.

Приказ болесника Приказујемо случај 50-годишњег болесника са спонтаним перирenалним крварењем чији узрок није откривен ни после шест месеци праћења од акутног крварења.

Закључак У случајевима перирenалног крварења, узрок не може увек бити откривен по пријему болесника, упркос данашњим врло развијеним радиолошким методама. У овим случајевима важно је имати на уму да је дуже праћење ових болесника, после акутног крварења, од суштинске важности за успостављање праве дијагнозе, с обзиром на то да перирenални хематом може маскирати присуство евентуалног малигнитета. У врло ретким случајевима узрок перирenалног крварења не може бити откривен ни после дужег праћења.

Кључне речи: Вундерлихов синдром; перирenална хеморагија; ангиомиолипом