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A rare case of spontaneous perirenal hemorrhage – Wunderlich syndrome

Редак случај спонтане перинеалне хеморагије – Вундерлихов синдром

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SUMMARY

Introduction Spontaneous perirenal hemorrhage or Wunderlich syndrome represents a rare entity in the urological settings. The vast majority of the causes are represented by angiomyolipoma and renal cell carcinoma. In other cases, the vascular abnormalities, polycystic kidneys, polyarthritis nodosa or pyelonephritis could represent the cause of perirenal bleeding. The treatment depends on clinical parameters at the presentation as on the presence of eventual renal malignancies. Our goal was to present a rare case of a healthy men who presented the idiopathic Wunderlich syndrome.

Case outline We present the case of 50 years old patients with spontaneous perirenal hemorrhage which was not due to any defined cause even after sixth months follow-up.

Conclusion In some rare cases of perirenal bleeding the cause cannot be defined at the presentation, even with today's very developed radiologic imaging and methods. Thus, it is important to be aware of the fact that in those cases, the longer follow up is needed, knowing that the presence of perirenal hematoma can always obscure the real diagnosis. Sometimes, even in the cases where the proper follow-up has been done, the real cause of the bleeding remains unknown.

Keywords: Wunderlich syndrome; perirenal hematoma; angiomyolipoma

САЖЕТАК

Увод Спонтана перинеална 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, поличистични бубрези, нодозни полиартеритис, пиелонефритис или идиопатски фактори. Третман ових пацијента зависи од клиничких параметара на пријему као и од присуства евентуалних малигних бубрежних обољења. Нас циљ је био представити редак случај релативно младог пацијента са Вундерлиховим синдромом који није био узрокован ниједним до данас знаних патологија.

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

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

Кључне речи: Вундерлихов синдром; перинеална хеморагија; ангиомиолипом

INTRODUCTION

Wunderlich was the first who presented the spontaneous hematoma of the kidney in 1856 [1]. It refers to spontaneous non-traumatic renal bleeding into subcapsular and/or perirenal space. This condition may be caused by various pathologies 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 the respective percentages are given in Table 1 [2, 3].

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

CASE REPORT

A 50-year-old male patient was admitted to emergency room with acute abdominal and right flank pain and painfulness to palpation. He did not report any history of trauma, the anamnesis was silent for other illnesses and he did not reported any drug assumption. During clinical examination the blood pressure of 120/80 mm Hg and the heart rate of 82 per minute were recorded. The hemoglobin value was 13.8 g/dl, leukocytes 15000/mm3 platelet count 270 × 1 10^9/L, creatinine clearance 0.88 mg/dl, coagulation parameters as prothrombin time (PT), activated partial thromboplastin time (aPTT) and international normalized ratio (INR) were in the normal range. Urine sediment shown just proteinuria. Ultrasound examination followed by CT scan of abdomen showed a huge 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. The next day the hemoglobin values went 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 day later, the hemoglobin values remained stable (11g/dl). 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 Hb levels on day 8 after the acute episode.

The control Angio-CT scan after 1, 3 and 6 months from the acute event showed the important reduction of the perirenal hematoma without apparent cause for the previous bleeding.

The figure from 1 to 5 show the CT scan of the right perirenal hematoma from the first presentation to 6 months follow-up: figure 1 showing the bleeding at the presentation, figure 2 showing CT scan after 2 days from the acute event, figure 3 showing the control Angio-CT scan at 1 month after the acute event, figure 4 showing the control Angio-CT scan at three months after the acute bleeding and figure 5 showing the control Angio-CT scan at 6 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

The spontaneous renal bleeding to the subcapsular and perinephric space, known as Wunderlich syndrome (WS) could be due to benign or malignant renal tumors, vascular lesions as polyarteritis nodosa, anatomical lesions as renal cysts, renal infections or nephritis [2]. Between 2003 and 2011 just 250 known cases have been reported [6, 7]. Renal neoplasms are the most common cause of WS, accounting for 60-65 % of all cases of which renal angiomyolipoma is the most common benign neoplasm responsible for WS [5, 7]. The renal cell carcinoma could also be the cause for WS but just in 0.3-1.4 % of cases [7]. This, potentially life-threatening condition which could be associated with hypovolemic shock and is also followed by other symptoms such as acute lumbo-abdominal pain and palpable abdominal mass, symptoms forming so called Lenk's triad [2]. Although the clinical guidelines for management of WS are not yet well established, the appropriate treatment for the patient with WS depends on the right diagnosis that a bleeding has occured in which the most important role has contrast-enhanced CT scan as a method of choice with sensitivity of 100 % [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 perform the embolization [10]. This

could be very helpful especially in some rare cases as segmental arterial mediolysis as reported from Skeik group in 2019 [11].

Thus, angiography and arterial embolization represent the 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 from Salgado et all, it is not possible to dominate the bleeding just with the 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 Wunderlich syndrome is to define the source of bleeding in order to postulate the right therapy.

In our case, it was not possible, in acute phase, to define the cause of bleeding. Fortunately, the bleeding limited itself spontaneously and the patient was treated in conservative manner. This did not make as sure that the bleeding site was not present. The literature reports that if the CT scan, followed by angiography, do not reveal the bleeding source, a CT scan should be repeated at the time distance, because it is obvious that if the hemorrhage is huge, an eventual renal cell carcinoma or angiomyolipoma or other eventual renal bleeding sites as renal cysts, could be seen just after the resorption of the hematoma has occurred [13]. That was also proved, not just in cases of renal angiomyolipoma or clear cell carcinoma, but also in rare cases of renal sarcoma presenting the Wunderlich syndrome [14]. Thus, we performed CT scan at 1, 3, and 6 months from the acute phase. As seen from the presented images, we were not able to give the answer about the real cause of bleeding even at the time distance of 6 months after the acute event.

It is worth to mention that hematologic issues can contribute to Wunderlich syndrome. As reported, some patient 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 Wunderlich syndrome, stating that if in acute phase the patients is hemodynamically stable, nephrectomy or partial nephrectomy should be deferred. A recent Korean study of 28 patient with WS stated that the definitive treatment for WS will depend on the clinical condition and underlying cause of the patient with possible therapeutic options including conservative therapy, angioembolization, nephron-sparing surgery, or radical nephrectomy [3, 17]. More interestingly they found that 5 of 28 patients had no obvious cause of perirenal bleeding. This was also the case with our patient, given that the nephrectomy or partial nephrectomy were not needed even later, as no malignant pathology could be documented.

In conclusion we can say that, although a vast majority of Wunderlich syndrome cases are represented by angiomyolipoma or by renal cell carcinoma, sometimes the cause remains unknown. We described a rare case of idiopathic Wunderlich syndrome where the cause could not be diagnosed even after 6 months follow-up.

Conflict of interest: None declared.

REFERENCES

1. Wuendrlich RA, Handbuch der Pathologie und Therapie, 2nd ed. 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; 57:840-845. doi:10.1053/crad.2002.0981. PMDI: 12384111.
3. Kim JW, Kim JY, Ahn,ST et al. Spontaneous perirenal hemorrhage (Wunderlich syndrome): An analysis of 28 cases. Am J Emerg Med. 2019; 37(1):45-4. doi: 10.1016/j.ajem.2018.04.045. PMID: 29779678
4. Ferrara SD, Bajanowski T, Cecchi R et al. Bio-medicolegal scientific research in Europe: a comprehensive bibliometric overview. Int J Legal Med 2011: 125(3):393-402. doi: 10.1007/s00414-010-0538-1. PMID: 21191611
5. Viel G, Boscolo-Berto R, Cecchi R et al. Bio-medicolegal scientific research in Europe. A country-based analysis J Legal Med. 2011: 125(5):717-25. doi: 10.1007/s00414-011-0576-3. PMID: 21594612
6. Zhang JQ, Feilding JR, Zou KH. Etiology of spontaneous perirenal hemorrhage: A meta-analysis. J Urol 2002; 167:1593-1596. doi: 10.1097/00005392-200204000-00006. PMID: 11912370.6.
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. doi: 10.1136/emj.20.4.388. PMID: 12835370.
8. Katabathina VS, Katre R, Prasad SR et al. Wunderlich syndrome: cross-sectional imaging review. J Comput Assist Tomogr. 2011; 35(4):425-33. doi: 10.1097/RCT.0b013e3182203c5e. PMID: 21765296.
9. Simkins A, Maiti A, Cherian SV. Wunderlich Syndrome. Am J Med. 2017; 130(5):e217-e218. doi: 10.1016/j.amjmed.2016.11.031. PMID: 28159184.
10. Ho TH, Yang FC, Cheng KC. Wunderlich syndrome, spontaneous ruptured renal angiomyolipoma & tuberous sclerosis. QJM. 2019; 112(4):283-284. doi: 10.1093/qjmed/hcz004. PMID: 30629244.
11. Skeik N, Olson SL, Hari G et al.. Segmental arterial mediolysis (SAM): Systematic review and analysis of 143 cases. Vasc Med. 2019; 24(6):549-563. doi: 10.1177/1358863X19873410. PMID: 31793853.
12. Salgado SL, Martinez GAE, Juarez AJ. Bilateral wunderlich syndrome secondary to synchronous bilateral testicular germ cell tumor. A case report. Urol case Rep. 2019; 28:101028. doi: 10.1016/j.eucr.2019.101028. PMID: 31641608.
13. Parmar N, Langdon J, Kaliannan K et al. Wunderlich Syndrome: Wonder What It Is. Curr Probl Diagn Radiol. 2021; 9:S0363-0188(21)00016-5. doi: 10.1067/j.cpradiol.2020.12.002. PMID: 33483188.
14. Ramanitharan M, Ketan M, Lalgudi ND. Case Report: Partial nephrectomy in primary renal sarcoma presenting as Wunderlich syndrome; a rare tumour with rare presentation managed atypically. F1000Res. 2019; 10;8:423. doi: 10.12688/f1000research.18698.1. PMID: 31354947.
15. Gajapathiraju C, Abhilash K. Wunderlich syndrome. Clin Case Rep. 2018; 6(9):1901-1902. doi: 10.1002/ccr3.1738. PMID: 30214791.
16. Neeraja S, Ramy S, Anum S, et al. Post-partum occurrence of Wunderlich syndrome and microangiopathic haemolytic anaemia (MAHA): a case report. J Community Hosp Intern Med Perspect. 2021. 23;11(2):277-279. doi:10.1080/20009666.2021.1883812. PMID: 33889338.
17. Choi HS, Kim CS, Ma SK et al. Wunderlich syndrome and regression of angiomyolipoma. Korean J Intern Med. 2020; 35(6):1528-1529. doi: 10.3904/kjim.2020.053. PMID: 32229794.

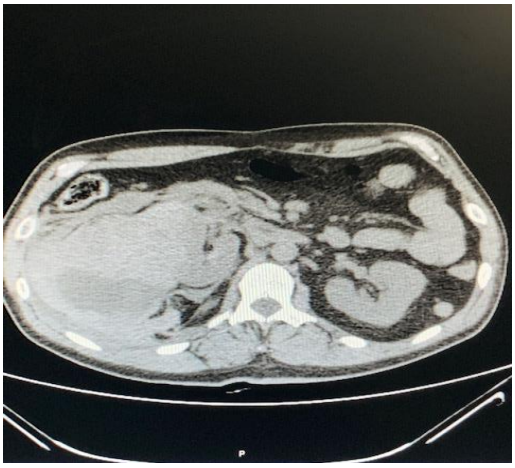

Figure 1. Abdominal computed tomography scan of the right retroperitoneal bleeding at the presentation

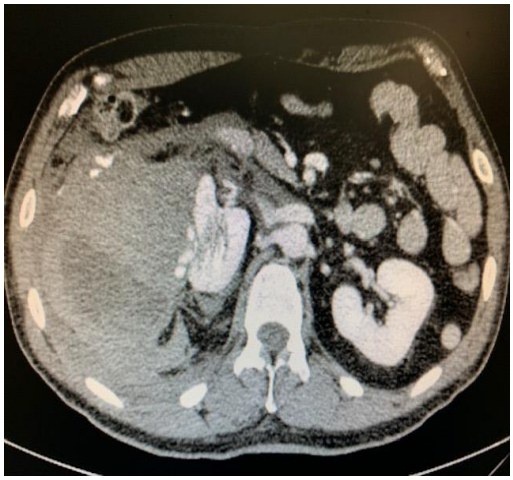

Figure 2. Abdominal computed tomography scan two days after the acute event

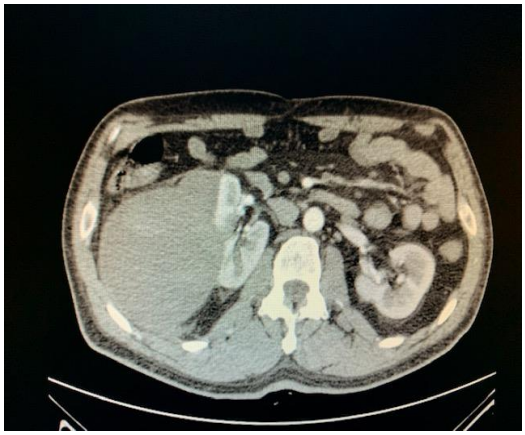

Figure 3. Computed tomography angiogram one month after the acute event

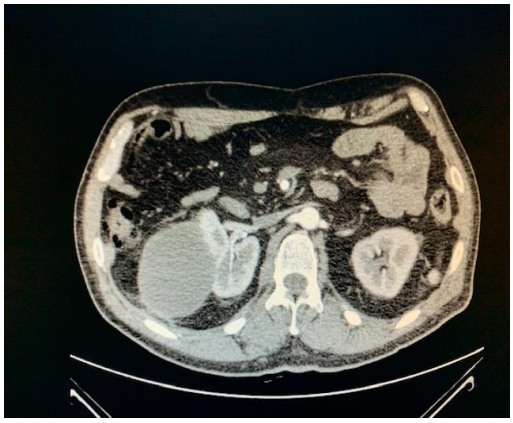

Figure 4. Computed tomography angiogram three months after the acute bleeding

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

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

