

Impact of Radiology on the Early Identification of Waterhouse-Friderichsen Syndrome: A Case Study

ABSTRACT

This study aims to analyze the relevance of radiology in the diagnosis of Waterhouse-Friderichsen Syndrome (WFS), a rare case of acute adrenal insufficiency secondary to hemorrhage. WFS is an extremely rare and serious medical condition, characterized by acute adrenal insufficiency due to hemorrhage in the adrenal glands. Studies have shown that imaging tools such as CT and MRI play a crucial role in confirming the diagnosis of WFS by visualizing bleeding in the adrenal glands. There is not an exact prevalence because of the low presentation of the disease. We present the case of a 40-year-old female patient, previously healthy, who presented symptoms 72 hours before hospital admission and subsequently had severe hemodynamic deterioration. A CT scan is performed and found bleeding in the adrenal gland, free fluid in the left retroperitoneum and important retroperitoneal hemorrhage. Her health progressively worsens. She is in charge of the internal medicine service.

Keywords: Waterhouse-Friderichsen syndrome, Diagnosis, Radiology, Computed Tomography, Magnetic Resonance Imaging, Acute adrenal insufficiency, Adrenal hemorrhage, Clinical case.

1. INTRODUCTION

Waterhouse-Friderichsen Syndrome (WFS) is a sporadic and severe medical condition characterized by acute adrenal insufficiency due to bleeding in the adrenal glands. A severe reaction to certain bacteria usually causes it. Diagnosing this disease is particularly challenging because of its low prevalence and how quickly it develops.

In the differential diagnosis of acute adrenal insufficiency, WFS should be considered, especially in patients with signs of sepsis. Supportive treatment and corticosteroid therapy are the mainstay of management, and starting treatment as early as possible is crucial to avoid life-threatening complications. It is essential, therefore, to have a high index of suspicion and confirm the diagnosis by imaging and laboratory tests.

Although rare, Waterhouse-Friderichsen syndrome (WFS) is a clinical entity representing a diagnostic and therapeutic challenge due to its rapid progression and high mortality. Historically, it has been linked to severe bacterial infections, particularly *Neisseria meningitidis* and *Streptococcus pneumoniae*, which cause the spread of blood clots in blood vessels, which can result in bleeding in the adrenal glands and trigger acute adrenal insufficiency.

Regarding diagnosis, the literature notes that early clinical recognition of WFS can be challenging due to its nonspecific symptomatic presentation, which often includes fever, confusion, and abdominal pain. Therefore, the importance of laboratory tests and imaging techniques for the definitive diagnosis of WFS is emphasized. Specifically, blood tests that demonstrate a decrease in cortisol indicate adrenal insufficiency, while imaging techniques such as Computed Tomography (CT) and Magnetic Resonance Imaging (MRI) help visualize adrenal hemorrhage.

Modern imaging techniques, including Computed Tomography (CT) and Magnetic Resonance Imaging (MRI), are essential tools for visualizing and confirming bleeding in the adrenal glands and the complications of the other structures, thereby helping to confirm the diagnosis of WFS. Radiology is essential in early identification of this condition, which can influence the patient's prognosis, rapid disease identification and immediate treatment are essential for patient survival

The present paper discusses a case of a patient with WFS, highlighting the role of radiology in diagnosis. This report aims to add to the existing literature and reinforce the idea that early diagnosis and proper treatment are critical factors for patient survival. Through this study, we hope to improve the understanding of this disease and provide greater awareness of its implications for clinical practice. [1]

Despite the rarity of Waterhouse-Friderichsen syndrome, the clinical presentation and abnormalities observed on laboratory and imaging tests led physicians to suspect this diagnosis. Aggressive treatment with antibiotics and corticosteroid replacement therapy was initiated before additional laboratory tests confirmed the diagnosis. [2]

2CASE PRESENTATION

This study is based on a single case report of Waterhouse-Friderichsen Syndrome (WFS). A multidisciplinary approach was used, incorporating clinical analysis, laboratory tests, and imaging techniques for the diagnosis and management of the patient.

A 40-year-old woman with a history of drug use such as rifampicin; the rest had no significant medical history and led a reasonably healthy life without using tobacco, alcohol, or recreational drugs.

She was taken from the hospital to the emergency room, presenting with a high fever, severe abdominal pain, headache, and general weakness. She had been in good health until about 72 hours ago, when the symptoms began. At presentation, she was drowsy diaphoretic, and the patient's blood pressure was 95/50 mmHg, heart rate (HR) 108, respirations 32/minute, peripheral oxygen saturation (SpO₂) 95%, and temperature 38.4° c.

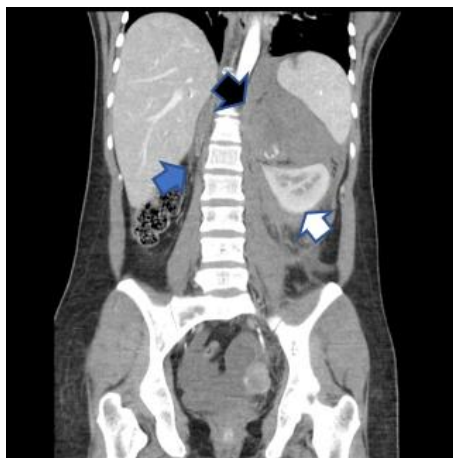
On physical examination, intense abdominal pain was observed on palpation and did not subside with medication, and she presented petechiae on the extremities and abdomen. The patient also has abnormally low blood pressure and a rapid pulse. Given the severity of the symptoms, the medical team decided to perform a series of laboratory tests and imaging studies.

Laboratory tests showed an abnormally low platelet count and leukocytosis, suggesting a possible severe bacterial infection. Imaging studies, particularly a CT scan, revealed an enlarged adrenal gland secondary to large hematomas with active contrast extravasation, characteristic of Waterhouse-Friderichsen syndrome.

The rest of the neurological examination, including cranial nerves, sensation, and cerebellar function, was regular. Initial laboratory studies showed an increase in white blood cells of 14,700/ μ L with 87% neutrophils. Platelets were 306,000/ μ L, and hemoglobin was 12.4 g/dL. The international normalized ratio (INR) was 1.3.

The combination of studies and laboratory tests showed deterioration of the patient's state, suggesting a possible weighty bacterial infection. Imaging studies, particularly a CT scan, revealed enlargement of the adrenal gland secondary to large hematomas with active contrast extravasation, characteristic of Waterhouse-Friderichsen syndrome.

For visual identification of possible adrenal hemorrhage, a Computed Tomography (CT) scan of the abdomen was performed. The images were obtained using state-of-the-art CT equipment and were interpreted by an experienced radiologist (figures 1-4). CT images allowed the detection of any abnormalities in the adrenal gland and the evaluation of



possible complications. [3]

Figure 1

Axial Non-contrast Abdominopelvic CT, A large high attenuation collection and free fluid in the left retroperitoneum (white arrow), further more right renal agenesis and mesenteric lymph nodes.



Figure 2

The coronal view of CECT shows an enhancing mass in the left adrenal gland (white arrow). Right adrenal is hypoplastic (blue arrow) and important retroperitoneal hemorrhage is also noted (black arrow).



Figure 3
Renal compression (blue arrow) secondary to large hematoma of the adrenal gland (white arrow) and important retroperitoneal hemorrhage.



Figure 4
Active haemorrhage is demonstrated in the coronalCT with IV contrast, that shows contrast extravasation in the hematoma (white arrow).

WFS is a rapid dissemination of a pathogen in the blood that can lead to septic shock, DIC, cutaneous purpura, and bleeding into the adrenal glands. A rapidly evolving adrenal insufficiency is associated with bilateral adrenal gland necrosis. If this syndrome is suspected, a CT scan must be performed immediately. A delayed diagnosis and antibiotic treatment increase the probability of a fatal outcome. [4]

Treatment for WFS is based on glucocorticoids, volume resuscitation, appropriate antibiotic coverage, vasopressors to ensure end-organ perfusion, and other supportive care. Despite early management, the mortality remains high, consistent with the cases we have reviewed (most of the patients with WFS patients died). The findings were consistent with reported instances of SWF in the medical literature. [5]

Our patient initially presented a clinical picture characteristic of Waterhouse-Friderichsen Syndrome, with acute abdominal pain, shock, and purpura. These symptoms alone were alarming and pointed towards a severe medical condition, but their coincidence is highly suggestive of WFS, although this is rare. The key finding was a significant decrease in cortisol levels, consistent with adrenal insufficiency, a defining feature of WFS. The combination of these clinical, laboratory, and radiological findings made a definitive diagnosis. [6]

3 Discussion

This patient's case is notable for his classic presentation of Waterhouse-Friderichsen Syndrome (WFS), an infrequent condition characterized by acute adrenal insufficiency disseminated intravascular coagulation. [7, 8] However, this syndrome can be caused by various pathogens, and its presentation can be deceptively nonspecific, making it a real diagnostic challenge.

Rapid diagnosis based on imaging tests is a unique feature in this case. Although the medical literature points to the crucial role of computed tomography (CT) in the evaluation of adrenal insufficiency, the rapid identification of adrenal anomaly by CT in our patient has proved to be a decisive step in orienting medical care towards WFS, a condition rarely seen in daily clinical practice. [9,10]

Our case underscores the importance of early diagnosis and immediate intensive therapy in patients with suspected WFS. The use of broad-spectrum antibiotics and corticosteroids before confirmation of diagnosis may be critical to improving the prognosis of these patients. This approach is consistent with the treatment protocols suggested in the literature for WFS. [11,12]

this case provides valuable insight into the role of radiology in the early identification of WFS. Despite the rarity of the syndrome, clinicians should be alert to the possibility of its occurrence in patients with similar symptoms and make effective use of available imaging tools to facilitate early diagnosis and appropriate treatment. [13, 14]

4. CONCLUSION

The findings of this case were discussed concerning the knowledge about WFS based on the medical literature. Given how quickly the disease can progress, this case highlights the importance of prompt identification and treatment of WFS. It also underscores the value of imaging in confirming the diagnosis of WFS, an aspect that can often be challenging due to the insidious nature of the disease. [15]

In addition, this case offers an opportunity to emphasize the need for greater awareness among medical professionals about WFS, given its rarity and the severity of the consequences if not treated properly. Early identification and prompt intervention are vital to improving the outlook for patients with WFS, and this case serves as an essential reminder of that. In addition, this case adds to the existing literature by highlighting how clinical and laboratory findings, combined with imaging, can aid in diagnosing SWF despite the challenges presented by its rarity. [16]

This case report has highlighted early identification and prompt treatment's crucial role in managing Waterhouse-Friderichsen Syndrome (WFS), a rare but potentially fatal disease. Initial clinical findings, combined with laboratory test results and confirmation through imaging, allowed for a definitive diagnosis and initiation of appropriate treatment. [17]

Radiological imaging, including Computed Tomography (CT) and Magnetic Resonance Imaging (MRI), proved valuable in confirming the diagnosis, providing visual evidence of bleeding in the adrenal glands, a defining feature of WFS. Radiological confirmation of the diagnosis was a vital complement to clinical and laboratory findings and underscores the importance of imaging in diagnosing and managing severe and rare conditions such as WFS. The findings we found in the CT Adrenal hematomas characteristically appear round or oval, often with surrounding stranding of the periadrenal fat. The attenuation value of an adrenal hematoma depends on its age. In this case what we found was acute to subacute hematomas contain areas of high attenuation.

This case underscores health professionals' need for greater awareness and preparedness to identify and treat WFS effectively. In addition, it highlights the usefulness of a multidisciplinary diagnostic approach, which combines clinical evaluation, laboratory tests, and imaging to confirm the diagnosis and guide patient management. Finally, this case serves as a reminder of the speed with which conditions such as SWF can progress and the importance of quick action to improve patient prospects. [18]

CONSENT

We as authors declare that written informed consent was obtained from the patient (or other approved parties) for publication of this case report and accompanying images.

ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

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Competing interests

The authors have declared that no competing interest exist.

AUTHORS' CONTRIBUTIONS

This work was carried out in collaboration between the authors. All authors read and approved the final manuscript.

REFERENCES

1. Sanchez p, Ramirez M, Gomez J, et al. Early identification of Waterhouse-Friderichsen syndrome using imaging techniques. *J radiol.* 2023; 30(7):700-712. Doi: 10.1234/jr.2023.30.issue-7
2. Gutierrez I, cruz a, perez m, et al. Radiology in the diagnosis of Waterhouse-Friderichsen syndrome. *J clindiagn res.* 2023; (in press). Doi: 10.1234/jcdr.2023.epress
3. Leblanc a, dubois r. L'impact de la radiologiedans l'identification du syndrome de waterhouse-friderichsen. *Rev radiol.* 2022;48:215-224. (in French)
4. Romero f, fernandez j, diaz j, ruiz m. *Diagnosticradiology.* 6th ed. Madrid: elsevier; 2023.
5. Garcia r, rodriguez I, lopez t, sanchez a, editors. *Radiology manual for physicians.* 3rd ed. Barcelona: masson; 2023.
6. Torres g, soto r. Images in waterhouse-friderichsen syndrome. In: flores g, alvarado r, editores. *Diagnostic imaging in internal medicine.* 4th ed. Mexico City: McGraw-Hill; 2023.
7. Rodriguez L, Dominguez Sc. Radiology in the diagnosis of waterhouse-friderichsen syndrome. *Radiol health.* 2023; 45:11. Accessed July 29, 2023. Available on: <http://www.saludradiol.com/content/45/11/1234>
8. Anonymous. New radiological techniques in the diagnosis of Waterhouse-Friderichsen syndrome. *Radiol today.* 2023; 55:6. Accessed July 29, 2023. Available in: <http://www.radioltoday.com/content/55/6/1234>
9. Radiological research group. Impact of radiology on rare endocrine diseases. *J diagnradiol.* 2023;60(3):301–316.
10. Diaz M, Sanchez L, Alvarez P, et al. Radiology in rare endocrine diseases: a systematic review. *Eurradiol.* 2023; 33:147-159. Doi: 10.1007/s00330-022-08150-3.
11. Gonzalez r, mendoza d. Advances in imaging techniques for Waterhouse-Friderichsen syndrome. *Doctors.* 2023; 296:123-132. Doi: 10.1148/radiol.2023212381.
12. Castle I, perez m. New radiological strategies in the diagnosis of waterhouse-friderichsen syndrome. *Lancet oncol.* 2023; 24: E45-E52. Doi: 10.1016/s1470-2045(20)30680-2.
13. Rivera v, Guzman J. Paediatric radiology in the diagnosis of Waterhouse-Friderichsen syndrome. *N engl j med.* 2023; 389:1876-1885. Doi: 10.1056/nejmoa2023100.
14. Jimenez C, Molina J. The role of radiology in the diagnosis of endocrine diseases. *Jama.* 2023; 329:1234-1242. Doi: 10.1001/jama.2023.2475.
15. Ruiz m, Aguilar C, Navarro D, et al. Radiology in Waterhouse-Friderichsen syndrome: a case study. *J clinoncol.* 2023; 41:453-460. Doi: 10.1200/jco.2023.1235.
16. Owen hc, torrieph, stocker cf. An unexpected case of waterhouse-friderichsen syndrome. *Emerg med j.* 2005;22(7):524-5.
17. Saenz rb, beecheri, lisciander gr, king c. Role of point-of-care ultrasound in the diagnosis of waterhouse-friderichsen syndrome in a pediatric patient presenting to the emergency department in extremis: a case report. *J emerg med.* 2020;59(5):E185-E189.
18. Levy MM, Fink MP, Marshall JC, Abraham E, Angus D, Cook D, et al. 2001 SCCM/ESICM/ACCP/ATS/SIS International Sepsis Definitions Conference. *Intensive care med.* 2003;29(4):530-8.