

# Acute Adrenal Crisis: A Medical Emergency

## *Insuficiência Suprarrenal Aguda: Uma Emergência Médica*

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### ABSTRACT

Acute adrenal crisis is a rare but severe condition. Clinical symptoms are diverse and non-specific, therefore, the diagnosis of this condition relies heavily on a high level of clinical suspicion and should be considered, if relevant, regardless of the patient's medical history. Timely treatment is crucial to the survival of patients with this condition.

The case of a woman with a history of adrenal insufficiency due to non-classic 21-hydroxylase deficiency exemplifies the importance of recognizing warning signs that may indicate an acute adrenal crisis.

This condition is life-threatening and can be initially assessed in the context of primary healthcare, making it essential for family physicians to be aware of this condition and how to prevent it and recognize it promptly. The failure to adjust chronic corticosteroid therapy in the face of respiratory infection was likely the triggering factor for the acute adrenal crisis.

**KEYWORDS:** Adrenal Hyperplasia, Congenital; Adrenal Insufficiency; Emergencies

### RESUMO

A insuficiência suprarrenal aguda é uma doença rara, mas grave. Os sintomas clínicos são diversos e inespecíficos, assim, o diagnóstico depende de um nível elevado de suspeição clínica e deve ser considerado, independentemente dos antecedentes médicos do utente. O tratamento rápido é crucial para o prognóstico.

O caso clínico descreve um quadro de insuficiência suprarrenal aguda numa utente com diagnóstico prévio de insuficiência suprarrenal crónica. Destaca-se a importância de identificar rapidamente sinais de alerta que possam sugerir uma crise suprarrenal aguda, que requer avaliação e referência hospitalar imediata. É uma con-

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dição potencialmente ameaçadora de vida que pode ser inicialmente avaliada no contexto de cuidados de saúde primários, tornando essencial que os médicos de família tenham conhecimento desta doença, saibam prevenir e reconhecê-la atempadamente. A falha no ajuste da corticoterapia crônica perante uma infecção respiratória foi provavelmente o fator desencadeador da crise no caso descrito.

**PALAVRAS-CHAVE:** Emergências; Hiperplasia Suprarrenal Congênita; Insuficiência Suprarrenal

## INTRODUCTION

Adrenal insufficiency is a disease caused by a disruption in the synthesis and secretion of cortisol by the adrenal gland. It can be of primary etiology, due to dysfunction or destruction of the adrenal cortex, with the main cause in developed countries being autoimmunity, resulting in a deficiency of glucocorticoids and mineralocorticoids, or of secondary etiology, with the most frequent cause being abrupt cessation of prolonged corticosteroid therapy, in this case, associated with deficiency of glucocorticoids.<sup>1,2</sup>

Among the causes of primary adrenal insufficiency are autoimmune pathology (Addison's disease; type I polyglandular syndrome, type II polyglandular syndrome); infectious (tuberculosis, fungal infection, cytomegalovirus, human immunodeficiency virus); vascular (bilateral adrenal hemorrhage, sepsis, coagulopathy, thrombosis); infiltrative (metastatic carcinoma, lymphoma, sarcoidosis, amyloidosis, hemochromatosis); congenital (congenital adrenal hyperplasia); and iatrogenic (adrenalectomy).<sup>3-6</sup>

Regarding 21-hydroxylase deficiency, the most common cause of congenital adrenal hyperplasia, it belongs to the group of autosomal recessive diseases, with global or partial impairment of glucocorticoid synthesis (resulting in increased ACTH and adrenal gland hypertrophy), with increased androgen levels.<sup>5-7</sup> Thus, the severity of its symptoms varies according to enzymatic activity, with manifestations occurring either early in the neonatal period (classic salt-wasting or virilizing form) or later in childhood or adulthood (non-classic form).<sup>8</sup> Clinical manifestations of the classic form may include salt-wasting crisis in the first weeks of life, with sepsis-like symptoms, as well as virilization and genital ambiguity in females, with clitoral hyperplasia or labial fusion. On the other hand, the much more common, non-classic form is characterized by a milder and progressive symptomatology with early adrenarche, acne, hirsutism, and fertility alterations. Diagnosis is suggested by increased levels of 17-hydroxyprogesterone and confirmation through a genetic test of the *CYP21A2* gene.<sup>5-8</sup>

Regarding treatment, patients with the classic form

require glucocorticoid and mineralocorticoid replacement to prevent the occurrence of potentially fatal adrenal crisis, while those with the non-classic form rarely require chronic treatment with steroids, but may require glucocorticoids in acute conditions.<sup>5-7</sup>

The decompensation of adrenal insufficiency can be triggered by multiple factors, including sudden discontinuation of corticosteroids or an acute stress situation without adjustment of therapy, which can lead to an adrenal crisis. The clinical symptoms of this condition are diverse and nonspecific, ranging from intense asthenia, postural hypotension, abdominal pain, nausea, vomiting, or fever, but can also progress to hypovolemic shock, with neurological dysfunction and coma.<sup>9,10</sup> Therefore, it is important to consider that the diagnosis depends on a high level of clinical suspicion, and in the case of an emergency, suspicion should be raised, regardless of whether the patient has this known condition, as it may be their inaugural presentation.

## CASE REPORT

Female, 53 years old, autonomous and cognitively intact. Her medical history included non-classical 21-hydroxylase deficiency adrenal insufficiency diagnosed in adolescence, dyslipidemia and osteoporosis. Her routine medication included prednisolone 5 mg once daily, rosuvastatin 10 mg once daily and alendronic acid 70 mg + colecalciferol 5600 IU weekly. No drug allergies were known and the National Vaccination Program was up to date.

She required a medical appointment at the primary care unit due to myalgia, asthenia and mucopurulent cough, evolving for four days, associated with the onset of nausea, vomiting, and abdominal pain. She denied recent pharmacological changes, recent trips, contact with animals, consumption of supplements, herbal products, unfiltered water, wild mushrooms or unpasteurized products. On physical examination, she was conscious, cooperative and oriented. Regarding vital signs, she was afebrile, tachypneic, hypotensive (blood pressure 89/63 mmHg), tachycardic (123 bpm), with peripheral oxygen saturation of 91%, in room air. She was dehydrated and non-jaundiced. She was also

hypoglycemic (61 mg/dL) with ketotic breath. Cardiac and pulmonary auscultation were normal, and abdominal palpation revealed no signs of peritoneal irritation but tenderness in the lower quadrants. Lower limbs had no edema or signs of deep venous thrombosis. The neurological examination revealed no abnormalities. Therefore, due to clinical instability, the patient was referred to the hospital Emergency Department (ED).

In the ED, she presented with respiratory alkalosis on blood gas analysis, as well as analytically, with acute kidney injury (urea 65 mg/dL, creatinine 1.19 mg/dL) and liver injury (AST 185 U/L, ALT 153 U/L, GGT 86 U/L, ALP 51 U/L), associated with a marked elevation of inflammatory parameters (CRP 266 mg/L). Urine analysis showed glucosuria (>500 mg/dL) and ketone bodies (>160 mg/dL), despite the absence of a previous diagnosis of diabetes and no history of SGLT2 inhibitor use. This finding may have been related to transient stress-induced hyperglycemia or acute tubular dysfunction during the adrenal crisis. Additionally, respiratory virus panel testing was positive for Influenza A and the thoracoabdominopelvic computed tomography scan revealed diffuse thickening of the bronchial walls in both lung fields, suggestive of inflammatory findings, and discal atelectasis in both lower lobes.

Thus, it was concluded that the patient had an acute adrenal crisis in the context of a viral respiratory infection. Steroid therapy and fluid replacement were initiated immediately after the first clinical evaluation in the ED, given the patient's clinical instability. No cortisol sample was collected before steroid administration, which represents a limitation for diagnostic confirmation.

During the hospitalization, there were no complications, and over the course of days, there was a progressive reduction of corticosteroid therapy. The hepatic and renal alterations also showed progressive improvement, and the patient was discharged after a week of hospitalization.

## CONCLUSION

Acute adrenal crisis is a rare but serious condition caused by decreased production of cortisol in the adrenal gland, which can present as a medical emergency. It is crucial to recognize the symptoms and provide immediate treatment to ensure the patient's survival. This condition is more common in patients with primary adrenal insufficiency who do not follow their prescribed corticosteroid therapy or adjust it appro-

priately during severe illness, acute infections, or gastrointestinal disorders.

As demonstrated in the presented clinical case, failure to adjust corticosteroids in response to a respiratory infection was enough to trigger an adrenal crisis, requiring urgent treatment with high-dose corticosteroids and intravenous fluid therapy.<sup>9,10</sup>

Thus, this clinical case highlights the importance of recognizing and valuing warning signs that may indicate serious illness, such as acute adrenal crisis, and it is crucial for timely hospital referral. Additionally, it is imperative to adjust corticosteroid doses promptly during acute illness in patients with primary adrenal insufficiency to prevent clinical deterioration and fatal decompensation. Furthermore, patients with primary adrenal insufficiency should be literate about their condition to manage it better and seek medical attention early when necessary.

## CONTRIBUTORSHIP STATEMENT/ DECLARAÇÃO DE CONTRIBUIÇÃO

BMP - Conducting the research, conceiving the original idea, writing the article, discussing the results, and contributing to the final manuscript.

AD, CCR - Resources, writing the article, contributing to the design and implementation of the research, writing the article, discussing the results and contributing to the final manuscript.

CR - Planning and supervising the work, writing the article, discussing the results and contributing to the final manuscript.

DA - Writing of the article, supervision, writing of the article, discussion of the results and contribution to the final manuscript.

MP - Resources, planning and supervision of the work, writing of the article, discussion of the results and contribution to the final manuscript.

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BMP - Realização da pesquisa, concepção da ideia original, redação do artigo, discussão do resultado e contribuição para o manuscrito final.

AD, CCR - Recursos, redação do artigo, contribuição para a concepção e implementação da pesquisa, redação do artigo, discussão do resultado e contribuição para o manuscrito final.

CR - Planejamento e supervisão do trabalho, redação do artigo, discussão do resultado e contribuição para o manuscrito final.

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