AACE Clinical Case Rep. 7 (2021) 228–229

Contents lists available at ScienceDirect

AACE Clinical Case Reports

journal homepage: www.aaceclinicalcasereports.com

Visual Vignette

Acute Unilateral Nonhemorrhagic Adrenal Infarction in Pregnancy

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ARTICLE INFO

Article history:
Available online 13 January 2021

Case Presentation

A 24-year-old woman, 30 weeks pregnant, presented to an emergency room with severe, sharp, constant abdominal pain in the left upper quadrant, which radiated to the back and was associated with nausea and vomiting. She appeared diaphoretic, with dry oral mucous membranes. Abdominal examination showed a gravid uterus, which was tender upon light palpation in the left upper area, with voluntary guarding. Genitourinary examination revealed a closed cervical os, with no blood in the vaginal vault. Abdominal magnetic resonance imaging revealed a T2-hypointense left adrenal gland (Fig. 1) with loss of normal hyperintensity on diffusion-weighted imaging (Fig. 2). Edematous changes were identified within the left perinephric and anterior pararenal spaces (Fig. 3). Other than moderate hepatomegaly, the rest of the abdominal and pelvic organs, vasculature, and abdominal wall were normal.

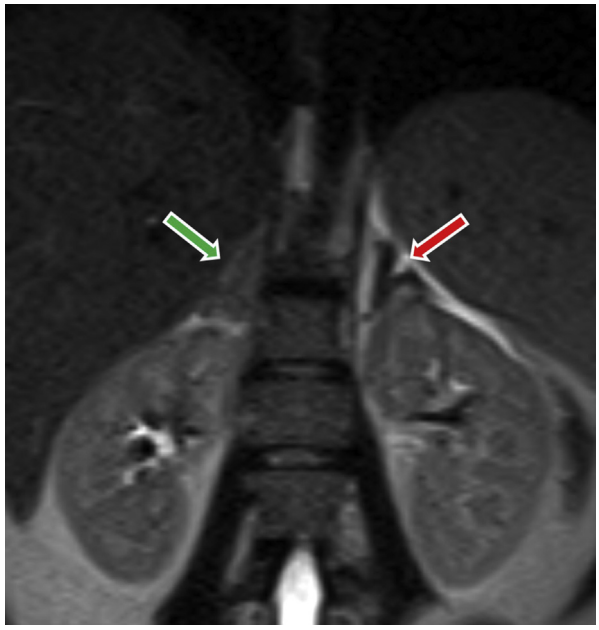

Fig. 1.

What is the diagnosis?

Answer

The diagnosis was left adrenal gland nonhemorrhagic infarction with necrosis. The patient was admitted for hydration and pain control. Her morning serum cortisol level was equivocal at 10.3 μg/dL, with a midnormal adrenocorticotropic hormone (ACTH) level of 38.9 pg/mL. ACTH stimulation led to a suboptimal cortisol response, with a serum cortisol level of 9.1 μg/dL at baseline, 10.7 μg/dL at 30 minutes, and 11.0 μg/dL at 60 minutes. The patient was started on a stress dose of intravenous hydrocortisone. Evaluation for thrombophilia was conducted, and the patient was initiated on therapeutic weight-based enoxaparin. Investigations for etiology included normal factor V Leiden, C and S protein activity, antithrombin III antigen, antiphospholipid screening, C3 and C4 complements, anti-double stranded DNA antibody, antineutrophil cytoplasmic antibody, cryoglobulin, homocysteine, and factor II mutation. Factor VIII activity increased to >200%, which was within the expected range during pregnancy. The patient was heterozygous for *MTHFR C677T* and *A1298C* mutations. Hydrocortisone was tapered, and the patient was discharged on a physiologic dose in addition to enoxaparin. On follow-up, after not receiving hydrocortisone for 24 hours, her morning serum cortisol level was 16.1 μg/dL, with a concurrent ACTH level of 129 pg/mL. The patient was

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https://doi.org/10.1016/j.aace.2020.12.015

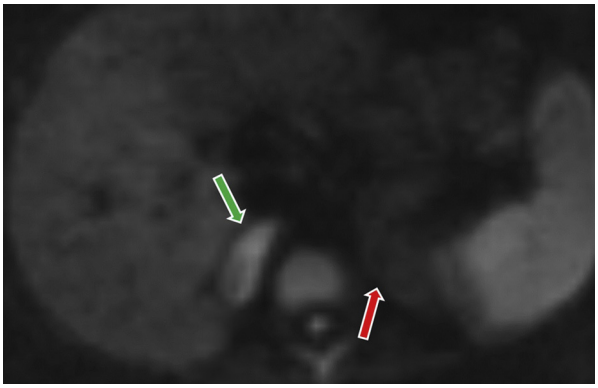

Fig. 2.

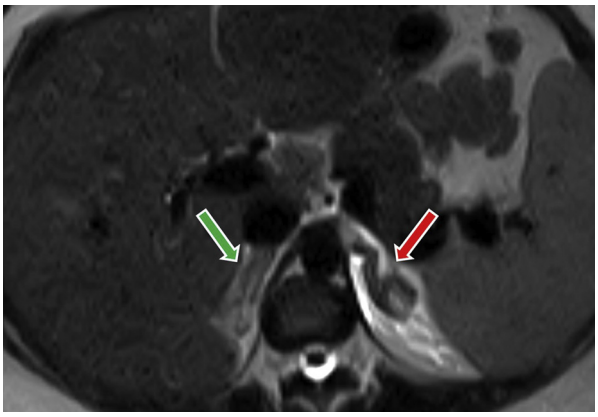

Fig. 3.

asked to discontinue hydrocortisone as she was doing clinically well. At 36 weeks of gestation, the patient was admitted because of dizziness and documented to have orthostatic hypotension. Her morning serum cortisol level was 11.9 μg/dL, with an ACTH level of 155 pg/mL. ACTH stimulation led to a serum cortisol level of 8.4 μg/dL at the baseline, at 16.3 μg/dL at 30 minutes, and 17.3 μg/dL at 60 minutes. She was resumed on a physiologic dose of corticosteroids, given her clinical presentation, but the patient was not able to pick up the medication and did not resume taking it. At 39 weeks of gestation, she delivered a viable female infant via a normal vaginal delivery. She was given a stress dose of corticosteroids during labor and discharged for home on a physiologic dose of prednisone once a day because of the patient's preference over having to take hydrocortisone twice a day, with plans to retest for adrenal gland function on follow-up, which should occur soon, to determine if prednisone can be safely discontinued.

Adrenal infarction is a very rare cause of abdominal pain during pregnancy.[1] Pregnancy is considered an independent risk factor for inducing a thrombophilic state.[1] A common underlying etiology is antiphospholipid syndrome, in which the infarction is usually bilateral and can be life-threatening if not recognized and treated promptly.[2–4] A study has evaluated the long-term effects of antiphospholipid syndrome-induced adrenal infarction and has shown favorable outcomes in those who survived its acute phase, with 1 obvious complication being adrenal insufficiency.[4] Interpretation of serum cortisol levels and relying on the results of ACTH-stimulation test in pregnant patients can be challenging because of increased corticosteroid-binding globulin production in the liver.[5] Urine or salivary cortisol levels were not measured in our patient. We decided to proceed with initiating hydrocortisone based on our clinical judgment at the time of the patient's acute presentation, realizing that the ACTH level obtained prior to the ACTH-stimulation test was not high and the contralateral adrenal gland appeared normal on imaging. The plan was to reconsider the need for continuing corticosteroids at follow-up. *MTHFR* gene mutations have also been linked to adrenal infarction.[1] Magnetic resonance imaging is the preferred imaging modality because it has high sensitivity in detecting adrenal infarction and can also be used to assess an underlying adrenal hemorrhage, if present.[2] The anatomy of the adrenal gland makes it prone to infarction in a thrombotic state because venous drainage is done via only 1 vein. Any clotting in that vein can cause blood stasis and lead to gland necrosis. To this end, patients with unilateral adrenal infarction benefit from anticoagulation, which protects the other gland from venous clotting and infarction. However, clinicians must be aware of the increased risk of bilateral adrenal hemorrhage with therapeutic doses of anticoagulants.

Disclosure

The authors have no multiplicity of interest to disclose. The views expressed in this article are those of the authors and do not reflect the official policy or position of the University of Florida.

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