Background: While hemorrhagic adrenal infarcts, usually bilateral, have been described in hematologic malignancies, usually PV, no hemorrhagic variant are exceedingly rare. Here we describe a single case of unilateral nonhemorrhagic adrenal infarct.
Case Presentation: 30-year- old male with past medical history of type 1 diabetes on insulin, presented with one day history of right upper quadrant (RUQ) abdominal pain. Initially pain was mild but slowly progressed to 10/10 intensity within the next several hours, felt like a sharp and stabbing pain with radiation up to the chest. Patient also reported to have difficulty breathing due to pain, along with nausea and vomiting. Laboratory results showed leukocytosis 11.2, H&H of 19.1/56.8, platelet count of 637, glucose of 283, alkaline phosphatase of 147. Abdominal ultrasonography reported nonspecific mild hepatomegaly and strongly suspected small thrombus within the IVC. Further work up included CTA study was negative for PE (pulmonary embolus), but MRI showed a filling defect in the suprarenal IVC along with right adrenal infarction. Patient was thermodynamically stable. Fortunately, cortisol was 21.3 and DHEA levels were 419, indicating no apparent effect on adrenal function. Patient underwent a bone marrow biopsy and was eventually diagnosed with PV. He was discharged in the stable condition with a close follow up with his endocrinologist for monitoring and repeat MRI for right adrenal infarct within the next few months.
Conclusion: This case illustrates the importance to screen patients with PV who presented with similar clinical presentation for possible adrenal insufficiency. It also highlights the importance of undertaking imaging for careful consideration, close follow up to prevent life threatening complications and even death.

Adrenal

ADRENAL CASE REPORTS

Adrenal Insufficiency in an Adolescent Boy With Type 1 Diabetes Mellitus - the Importance of Considering X-Linked Adrenoleukodystrophy

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Background: Primary adrenal insufficiency (PAI) is the rare, life-threatening failure of the adrenal glands to produce sufficient glucocorticoids and mineralocorticoids, presenting with fatigue, weakness, weight loss, hyperpigmentation, hypoglycemia, and hypotension. In adults, PAI is primarily autoimmune, occurring independently or in conjunction with thyroiditis and/or Type I diabetes mellitus (T1DM) as a component of Autoimmune Polyglandular Syndrome-2 (APS-2, Schmidt syndrome). APS-2 is rare (prevalence 1 in 20,000) and more common in females with a peak incidence in the third and fourth decades of life. In children, PAI is most often due to congenital adrenal hyperplasia (CAH), followed by autoimmune disease, but as many as 50% of cases of non-CAH pediatric PAI are not autoimmune. PAI is often the first documented clinical finding in boys with X-linked adrenoleukodystrophy (X-ALD), a rare disorder (prevalence 1 in 15,000) in which deficiency of a peroxisomal membrane protein leads to very long chain fatty acid (VLCFA) accumulation and progressive destruction of the adrenal cortex. A subset of boys with X-ALD develop cerebral ALD (cALD), characterized by progressive central demyelination, neurocognitive decline, and death. 70–80% of boys with X-ALD present with PAI prior to demonstrating neurologic symptoms. Diagnostic workup for X-ALD in pediatric patients presenting with PAI is crucial as timely intervention with hematopoietic stem cell transplant (HSCT) can stop progression of cerebral disease. **Clinical Case:** An eleven-year-old male was diagnosed with PAI after presenting with poor school performance, growth delay and skin hyperpigmentation. Medical history was significant for well controlled T1DM diagnosed at eight years old. Given his history of T1DM, his PAI was presumed to be autoimmune and further diagnostic testing was not performed. Eleven months later, a brain MRI performed for complaints of visual disturbance and chronic headaches revealed extensive demyelination with gadolinium enhancement consistent with cALD. Elevated VLCFAs and a mutation in the ABCD1 gene confirmed the diagnosis of X-ALD. Unfortunately, the extent of cerebral involvement was so severe that HSCT would not be of significant clinical benefit. In these situations, progressive neurological decline that leads to disability and ultimately death would be expected. **Clinical Lessons:** This case illustrates that VLCFA testing should be performed in all boys with PAI to rule out X-ALD. In boys, PAI should not be assumed to be autoimmune, even with co-existing autoimmune diseases since APS-2 (Schmidt syndrome) is rare and more likely to occur in adult women. PAI presents early and precedes neurologic symptoms in a majority of boys with cALD. Early identification of X-ALD through VLCFA testing prior to development of severe cALD is critical to allow early intervention with lifesaving HSCT.

Adrenal

ADRENAL CASE REPORTS

Adrenal Insufficiency Masquerading as Primary Hypothyroidism Following Immune Checkpoint Inhibitors Treatment

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Background: Immune checkpoint inhibitors (ICIs) are novel immunotherapy agents that have been used to treat multiple advanced cancer. Even though they confer potential clinical advantages by regulating immune reactions, they have been linked with serious immune-mediated adverse events. Here we present a case of a patient who was treated with ICIs, Nivolumab (programmed death-1 inhibitor) and Ipilimumab (cytotoxic T lymphocyte antigen-4 inhibitor), and subsequently developed two concurrent immune-related endocrine disorders. **Clinical Case:** An 83-year-old man with advanced renal cell carcinoma presented with generalized weakness. He had finished four cycles of immunotherapy with Nivolumab and Ipilimumab, and Ipilimumab was discontinued afterward. Two days after the fifth cycle of immunotherapy with Nivolumab,