Case Report

Type 1 diabetes in DKA with hypogonadotropic hypogonadism and pituitary apoplexy – A diagnostic dilemma

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ABSTRACT

A 17-year-old underweight boy came with a classical emergency of diabetic ketoacidosis associated with two additional clinical features; persistently low blood pressure despite adequate fluid resuscitation and hypogonadal features with Tanner stage 3 pubic hair, absent facial, and axillary hair along with a high-pitched voice. These findings triggered an endocrine workup which revealed hypogonadotropic hypogonadism. Suspecting primary pituitary pathology, an magnetic resonance imaging brain, was done which showed a well-defined hyperintense lesion in the pituitary suggestive of pituitary apoplexy. In the absence of headache, diplopia, and visual field defects, this incidental finding posed a dilemma regarding the diagnosis and management of diabetic ketoacidosis in the presence of apoplexy.

Key words: Delayed puberty, Diabetic ketoacidosis, Hypogonadotropic hypogonadism, Pituitary apoplexy

Pituitary apoplexy is a life-threatening clinical emergency that occurs from hemorrhage or ischemia of the pituitary gland, usually in the presence of non-functioning pituitary adenomas [1]. Common presenting symptoms are headache, visual impairment, and altered sensorium; however, 10–25% of patients can be asymptomatic [2]. The relative rarity and non-specific symptoms make it a diagnostic challenge, especially in the background of diabetic ketoacidosis with similar clinical presentations.

CASE REPORT

A 17-year-old male patient was brought to the emergency department with complaints of nausea, repeated episodes of vomiting, abdominal pain, and drowsiness. The patient was a recently diagnosed diabetic and he had skipped his insulin injections for the past 3 days.

On arrival, he was found to be dehydrated with a pulse rate of 98 beats/min, blood pressure of 80/60 mm of Hg, a saturation of 98% at room air, and a temperature of 99.7° Fahrenheit. He had raised blood glucose levels of 400 mg/dL and ketosis with urine ketone bodies being 2+. Arterial blood gas analysis showed high anion gap metabolic acidosis; hence, a diagnosis of diabetic ketoacidosis was confirmed. The patient was started on intravenous fluids. In spite of adequate fluid administration for over 24 h, he continued to have a blood pressure of 90/60 mm of Hg. On general physical examination, it was noted that he had a lean body habitus, absent facial, and axillary hair, along with a high-pitched voice (Fig. 1). He had Tanner stage 3 pubic hair and bilateral testicular volume was 10 mL. There was no anosmia or gynecomastia.

Routine blood work revealed mild hyponatremia (Sodium 131 mEq/L) with normal kidney and liver functions. Persistent hypotension and mild hyponatremia despite adequate fluid resuscitation alerted us to the possibility of cortisol deficiency. An endocrine consult was taken at this point and hormonal assays were sent.

Suspecting a pituitary pathology, magnetic resonance imaging (MRI) of the pituitary was done. The adenohypophysis showed a well-defined area of altered signal intensity appearing hyperintense on T1 and hypointense on T2 with gradient echo sequences and showed blooming suggestive of pituitary apoplexy (Fig. 2).

The patient was started on IV steroids. The history was revisited at this point. There were no complaints of headache or diplopia. The patient was reassessed and the central nervous system showed no deficits of higher mental function. The third nerve examination was normal. Bedside visual field testing by confrontation method was normal. Other cranial nerves showed no evidence of palsy. An ophthalmologist consult was sought, and visual field testing was done which was found to be normal.

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Figure 1: Absent axillary hair with lean body habitus

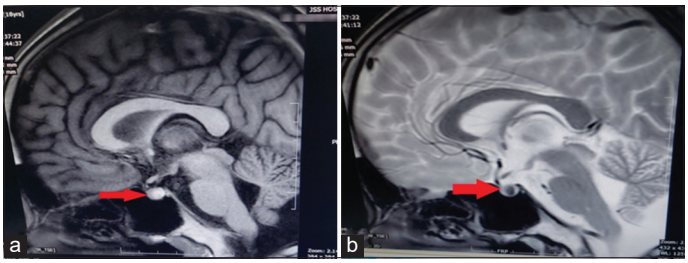

Figure 2: A lesion in the pituitary gland which appears hyperintense on T1 and hypointense on T2 suggestive of apoplexy

The hormonal assay revealed decreased testosterone (169.9 ng/dL), decreased follicle-stimulating hormone (1.4 IU/mL), and luteinizing hormone (4.7 IU/mL) suggestive of hypogonadotropic hypogonadism. Serum prolactin levels were normal (8.7 ng/mL), 8 am serum cortisol was 14.83 mcg/dL, adrenocorticotropic hormone (ACTH) (6.83 pmol/mL), and thyroid hormones were normal (thyroid stimulating hormone 2.5 micro IU/mL, free T4 7.4 mcg/dL). Serum cortisol was 14.3 mcg/dL which was inadequate for acute stress. An ACTH stimulation test was performed, which was normal.

The patient had an Hba1c of more than 15. His blood glucose levels were stabilized, and he was discharged after receiving counseling regarding compliance with insulin therapy. He is currently in follow-up with the endocrinology department and is receiving monthly testosterone injections.

DISCUSSION

Pituitary apoplexy is a rare life-threatening endocrine emergency. It has a slight male preponderance of 1.1–2.25 [3]. Around 21% of non-functioning pituitary adenomas can present as apoplexy and about 80% of adenomas can have apoplexy as their first presentation [4].

The causes of pituitary apoplexy can be varied. It occurs usually due to ischemia or hemorrhage into the pituitary gland compromising its blood supply. Other causes of apoplexy include systemic hypertension, anticoagulation therapy, coagulopathies, estrogen therapy, radiation therapy, major surgery, dopamine agonists, dynamic testing of pituitary functions especially thyroid hormone-releasing hormone, gonadotropin-releasing hormone, pregnancy, and head trauma [5].

The clinical presentation of apoplexy is also widely variable ranging from headache, most commonly, to visual field defects, and diplopia. The headache is often described as sudden and severe in nature usually retro-orbital in location but can also be bifrontal, suboccipital, or diffuse. The most common visual field defect has been found to be bitemporal hemianopia [6].

Around 20% of apoplexy cases can be asymptomatic [7]. Interestingly, in our patient, there was no history of headache, visual field defects, or diplopia. He presented with diabetic ketoacidosis in altered consciousness and this, further, confounded the presentation of apoplexy. Hyperglycemia has been known to alter the hemodynamics of pituitary circulation, but diabetic ketoacidosis leading to apoplexy has been rarely reported. It can be postulated that hyperglycemia leads to increased osmolality and dehydration causing a change in the microvasculature of the pituitary gland leading to apoplexy [8]. Other mechanisms include endothelial activation, impaired fibrinolysis, and platelet aggregation [9]. However, it can also be argued that the sudden physiological stress of apoplexy leads to a rise in the counter-regulatory hormones leading to hyperglycemia which may have precipitated diabetic ketoacidosis.

Regarding our patient, he had a history of poor compliance with insulin injections and his Hba1c was found to be more than 15. His hormonal assay also showed isolated hypogonadotropic hypogonadism with no raise in his counter-regulatory hormones. Thus, we may infer that his chronic hyperglycemia leads to pituitary apoplexy due to changes in the blood supply of the gland. He also appears to have a long-standing hypogonadism likely due to the compressive effect of a non-functioning pituitary adenoma.

MRI brain with contrast is the investigation of choice for pituitary apoplexy. CT can also be done, but it has lesser sensitivity as compared to MRI [10]. In a review of literature done by Couture *et al*., it was found that the most common abnormality in MRI was suprasellar and infrasellar mass with varying intensities in T1- and T2-weighted images according to the stages of hemorrhage. The compression of nearby structures was identified by T2-weighted images [11].

The management of acute apoplexy includes steroid administration and IV fluids. This is usually followed by maintenance steroid therapy, along with replacing the individual hormone deficits. Surgical decompression is recommended in patients who are clinically and neurologically unstable with a reduced level of consciousness or progressive visual deficit [12]. In our case, the patient had low testosterone levels, other pituitary hormones were normal, and the adrenocorticotropic hormone stimulation test was also normal. Hence, the patient received IV fluids and intravenous steroids in the acute phase followed by monthly testosterone injections in view of his isolated hypogonadotropic hypogonadism.

CONCLUSION

A high index of clinical suspicion is necessary during the management of diabetic ketoacidosis which can occasionally be complicated by other undiagnosed or suboptimally treated endocrine dysfunction. The latter needs prompt and appropriate treatment for the best outcome in diabetic ketoacidosis.

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