Neuroendocrinology and Pituitary

CASE REPORTS IN SECRETORY PITUITARY PATHOLOGIES, THEIR TREATMENTS AND OUTCOMES

A Case of Pseudotumor Cerebri After Removal and Recurrence of ACTH-Producing Tumor

Karla Bermudez Saint Andre, MD, Steven Petak, MD, JD, MACE, FACP, Laila Tabatabai, MD.
Houston Methodist Hospital, Houston, TX, USA.

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Introduction: Pseudotumor cerebri also known as idiopathic intracranial hypertension (IIH) is a condition of elevated cerebrospinal fluid pressure which as a result causes headaches and vision problems. Several case reports and studies have reported a relation of IIH with Cushing's disease (CD) in adults and children, particularly after surgical or medical therapy. We describe a very uncommon presentation of persistent intracranial hypertension (ICH) in a patient with recurrence of cushing's disease after her initial surgical resection.

Case presentation:38-year-old African-American female with a BMI of 45, type 2 diabetes, HTN and history of Cushing's syndrome from an ACTH-producing pituitary macroadenoma. She initially presented with increasing weight gain and features of Cushing's syndrome including hypertension, hyperglycemia, truncal obesity and moon facies. Imaging studies showed a 2 cm intrasellar mass with suprasellar extension without compression of the optic chiasm. Hormonal evaluation confirmed Cushing's syndrome from an ACTH-producing pituitary macroadenoma. Patient underwent initial transsphenoidal hypophysectomy (TSR) and was tapered down to physiologic doses of glucocorticoids.

Post-operatively she started complaining of significant headaches and transient vision loss. She followed up with neuro-ophthalmology and was diagnosed with papilledema that was not present in the pre-operative examination. An LP (lumbar puncture) was recommended to assess for ICH, however patient declined the procedure. A year after, a second surgery had to be performed for recurrent pituitary adenoma. Unfortunately, a repeat MRI pituitary shortly after her second surgery revealed recurrent pituitary macroadenoma of 2.2 cm. Patient continued with headaches and underwent an LP with an opening pressure of 28 cm H20 (ICH > 26 cm H2O). She underwent a third TSR with follow up MRI showing gross total resection of the previously seen pituitary mass. For her ICH she was started on acetazolamide but was not able to tolerate due to paresthesias and metallic taste. Her symptoms have improved after her last resection and last MRI brain shows no residual tumor. She is currently on furosemide and focusing on weight loss.

Conclusion: Our patient's presentation is an interesting and unusual case because we believe she had both pseudotumor cerebri (IIH) and real tumor cerebri from the complications of her ACTH-secreting macroadenoma. The cause of IIH after treatment of cushing's disease is believed to be mostly due to steroid withdrawal after surgical resection or medical treatment comprising hormonal control of cerebrospinal fluid production and absorption. In our patient we suppose that the persistent weight gain caused by the recurrence of her CD could also contribute to her IIH. The treatment in general is the same with physiologic doses of corticosteroids, diuretics and weight loss.

Adrenal

ADRENAL CASE REPORTS I

Pheochromocytoma Masquerading as Acute Coronary Syndrome. To Cath or Not to Cath?

Krish Khatri, MD[1], Richard Allen Haas, MD[2].
[1]UNIV OF MASSACHUSETTS, WORCESTER, MA, USA,
[2]UMass Memorial Healthcare, Shrewsbury, MA, USA.

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Background

Pheochromocytoma had been known as the "Great Mimic" as it can present with signs and symptoms consistent with numerous differentials [1]. Cardiac ischemia is one in particular which creates a diagnostic dilemma and poses significant risk for misdiagnosis.

Clinical Case

62-year-old female with recent onset hypertension and type II diabetes(HbA1C of 6.9%) presented to the ER with new episodes of nausea, vomiting, and substernal chest pain. During the preceding four months she also experienced episodic headaches, palpitations, and flushing.

Initial vital signs were significant for blood pressure 157/81 mm Hg. Physical exam was unremarkable. An Electrocardiogram showed normal sinus rhythm with right axis deviation and ST segment depressions in the inferior leads II, III and aVF.

Labs revealed a troponin peak at 2.95 (<0.04 ng/mL) and d-dimer of 403 (<400 ng/mL). Serum chemistry, TSH, and complete blood count were within normal limits. A CT Chest with angiography was negative for pulmonary emboli. However, it did note a heterogenous 7.6 cm right adrenal mass. At this juncture there was clinical concern for pheochromocytoma and serum free metanephrines was ordered. Prior to receiving medical treatment for her pheochromocytoma, she underwent cardiac catheterization which showed no evidence of coronary artery disease. The procedure was uncomplicated. However, post-procedurally she did develop worsening paroxysms and severe hypertensive episodes with heart rate up to 140 beats per minute and systolic blood pressure up to 220 mm Hg. She was subsequently started on alpha blockage with phenoxybenzamine 10mg twice daily to which she responded favorably. Her initial serum free metanephrine was 6087(< 57 pg/mL) and free normetanephrine 2489 (<148 pg/mL).

Conclusions

This case highlights the importance of maintaining a high index of suspicion for pheochromocytoma for all patients with acute chest pain and hypertension. Her untreated pheochromocytoma could have been fatal during or immediately after the cardiac catheterization. Given the suspicion for pheochromocytoma in this case, it would have been most appropriate to have begun alpha blockade after blood was drawn for metanephrines and before attempting any invasive procedures. Pheochromocytoma should be included in the differential diagnosis of acute coronary syndrome because it can mimic an ischemic episode.

References: