Abstract citation ID: bvac150.1301

Pediatric Endocrinology

PMON324

Crooke Cell Corticotroph Adenoma: A rare but Real Possibility in Children

Leena Mamilly, MD, Annie Drapeau, MD, MS, Patrick Walz, MD, Ralph Salloum, MD, and Rohan Henry, MD, MS

Background: Cushing disease (CD) is uncommon in the pediatric population and is usually caused by benign pituitary corticotroph microadenomas. In some adults with CD, Crooke cell adenoma (CCA), a more aggressive histologic variant of corticotroph adenomas, has been described. CCA has rarely been reported in children as the etiology of CD and its course is not well described.

Objectives: To report a pediatric case of CCA, the youngest to our knowledge

Case: An 11-year-old African American female presented to the endocrinology clinic with concerns for diabetes, excessive weight gain and linear growth arrest. Workup for possible etiology showed: midnight salivary cortisol 0.924 (normal < 0.112 μg/dL), ACTH 119(6-48 pg/mL), morning cortisol 8.5 (normal < 1.8 μg/dL) post 1 mg dexamethasone mg/dL, consistent with the diagnosis of CD. Magnetic resonance imaging (MRI) showed a 4 mm pituitary microadenoma in the left anterior side of the adenohypophysis for which she underwent endoscopic endonasal transsphenoidal resection. Histopathologic examination revealed pituitary adenomatous tissue consisting of a uniform population of basophilic cells with granular cytoplasm, arranged in trabeculae and small nests with bundles encircling the nuclei, findings pathognomonic of CCA. Given persistence of hypercortisolemia, another transsphenoidal gross total resection of residual adenomatous tissue was performed 4 months later. This led to resolution of CD as evidenced by weight loss and linear growth. Twelve months following the second surgery, the patient experienced rapid weight gain and a decelerated linear growth accompanied by pubertal arrest. Another pituitary MRI showed a recurrent left sided adenoma (8×8×7 mm). Biochemical investigation this time again showed elevated midnight salivary cortisol and 24- hour urine free cortisol (UFC). In addition, AM cortisol was unsuppressed following 1 mg overnight dexamethasone administration. In response, repeat transsphenoidal gross total resection was performed, however residual microscopic disease in the cavernous sinus wall was highly suspected intraoperatively. A spine MRI did not show disseminated disease. Despite this, hypercortisolemia persisted both clinically and biochemically. The patient subsequently underwent proton beam radiation therapy in combination with adjuvant adrenolytic therapy with mitotane. The patient responded to these interventions with normalization of 24-hour UFC, resumption of linear growth, and pubertal progression. The treatment course was complicated by the development of isolated TSH deficiency requiring thyroid hormone replacement.

Conclusion: CCA is an aggressive entity described in the adult literature as a cause of CD. The current report adds to the limited pediatric cases of this disease. CCA can be equally aggressive in children and refractory to surgical treatment. Here, we show that it may respond to proton beam radiation and adrenolytic therapy. Pediatric endocrinologists should be aware of the rare occurrence of CCA in children and tailor treatment accordingly using a multi-disciplinary approach.

Presentation: Monday, June 13, 2022 12:30 p.m. - 2:30 p.m.