evaluation of short stature [111.1cm (<3 percentile)]. He was born at 37 weeks of gestational age with birth weight of 3230g by cesarean section and had a port wine nevus on the right side of the face since birth. He had a history of epilepsy first occurring at 6 months of age when he was diagnosed with SWS. There were two more attacks of seizure till 32 months of age. Brain magnetic resonance imaging revealed leptomeningeal angioma, choroidal hemangioma, and diffuse brain atrophy. He was diagnosed with glaucoma and had been managed with surgery and medication. There was no family history of SWS or any other brain anomaly. His mid-parental height was 167.7cm. All blood tests were normal including complete blood count, chemistry, and thyroid function test. Hand x-ray showed delayed bone age. Cocktail test was performed for the evaluation of short stature. As a result, he was diagnosed with complete growth hormone deficiency (peak GH on L-dopa test: 2.46 ng/mL, peak GH on glucagon test: 3.71 ng/mL). The recombinant growth hormone therapy was started at the age of 8 years and 1 month. He showed good response to GH treatment. His height became 125.8cm at the age of 9 years and 5 months (height velocity 9.2 cm/year), and 134.3cm at the age of 10 years and 6 months.
Conclusion: We experienced a case with Sturge-Weber syndrome and complete growth hormone deficiency which was successfully managed by recombinant growth hormone therapy. It may be better to consider the possibility of GH deficiency even if there are certain conditions that affect the growth itself.

Neuroendocrinology and Pituitary

CASE REPORTS IN UNUSUAL PATHOLOGIES IN THE PITUITARY

Significant Response to Temozolomide in Two Aggressively Growing Pituitary Adenomas

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Introduction: Aggressive atypical pituitary tumors are characterized by invasive growth, recurrence and resistance to standard therapies. We present two female patients with pituitary adenomas in whom multiple other therapies had failed, who presented with significant response to temozolomide. **Case presentations:** In patient #1 (w, 78y), the diagnosis of macroprolactinoma had been made in a community hospital and dopaminagonistic treatment with bromocriptin had been initiated. After failure to achieve significant tumor reduction under this treatment and persisting visual field disturbances, first transnasal-transphenoidal surgery (TSS) was performed in 07/2011, followed by cabergoline exposure in increasing dose due to failure to control prolactin levels. Repeat TSS and stereotactic radiosurgery were performed in both 2014 and 2018 because of invasive tumor growth and double vision. She was then put on temozolomide. Patient #2 (w, 58y) presented with apoplectic gonadotropinoma in 2013. She also underwent 3 courses of TSS as well as stereotactic radiosurgery because of repeated tumor growth leading to visual field disturbances and double vision. Despite these measures, the tumor could not be controlled and she, as well, was put on temozolomide in 2018. In both cases costs were reimbursed by the patient's health care insurance and in both the first cycle was conducted with 150 mg/ body surface area (BSA) with escalation to 200 mg/BSA in the second. After only 2 cycles, double vision resolved in both patients and the tumor had shrunk by approximately 20% on MRI in patient #1 and even more in patient #2. In both patients, temozolomide dose was reduced again to 150 mg/ BSA due to side effects. Nevertheless, in both patients tumor volume further continued to decrease under therapy.
Conclusion: This promising clinical course after exposure to temozolomide with early, significant tumor shrinkage in two heavily pretreated patients with aggressive pituitary adenomas indicates that this therapy can be considered also in older patients and may yield astonishing results. Although temozolomide is increasingly becoming a therapeutic option for those patients whose pituitary tumors are refractory to standard therapies, further research and observance over time of temozolomide therapy in aggressive pituitary adenomas and carcinomas is indicated.

Pediatric Endocrinology

PEDIATRIC ENDOCRINE CASE REPORTS II

46 XX DSD Due to POR Deficiency

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Background
PORD (P450 oxidoreductase deficiency) is a rare form of CAH with marked phenotypic variations due to differences in the degree of steroid hormone excess/deficiency. PORD results in17αhydroxylase/17,20lyase- CYP17, 21αhydroxylase- CYP21, and aromatase- CYP19A1 inhibition. In the absence of characteristic skeletal features of Antley Bixler phenotype, differentiating PORD from other types of CAH is challenging.
Case details
04 day child, second of the non-identical twins, product of 2nd degree consanguinity, third in birth order was brought with abnormal genitalia. The other male twin had no genital ambiguity but had pigmented scrotum. There was no adrenal crisis in index case or maternal virilization. First child is normal female. Child was hemodynamically stable weighed 02 kgs and measured 51 cms, had no hyperpigmentation, skeletal deformities or dysmorphic features. Phallus was 10mm, clitoral index 40mm^2 with single urogenital opening and posteriorly fused labia (anogenital ratio 0.6).Gonads were not palpable. Karyotype was 46XX with normal Mullerian structures and non-visualized gonads on ultrasonography. Biochemical workup showed random plasma glucose level of 99mg/dl and normal electrolytes. Baseline serum 8am Cortisol was 1.15 mcg/dl (normal 5-18 mcg/dl) and 17OHP was 20 ng/ml (normal < 02 ng/ml). Serum Androstenedione level was 0.39 nmol/L (normal 0.5-3.4 nmol/L).