**INTRODUCTION**

Endogenous Cushing’s Syndrome is a rare disorder in children and Cushing’s disease (CD) caused by an ACTH-secreting pituitary adenoma is responsible for 75–80% of cases. Transsphenoidal surgery is now considered as a first-line therapy in CD but a proportion of pediatric patients who undergo pituitary surgery for CD don’t achieve cure or remission. Definitive treatment, such as surgery and/or radiotherapy, rather than long-term medical therapies, is currently recommended for the management of pediatric CD. Nelson’s syndrome is a potentially severe complication of bilateral adrenalectomy, more frequent in children than in adults. We report a case of CD in pediatric age submitted to bilateral adrenalectomy with subsequent Nelson’s syndrome.

**CASE REPORT**

**History**
- Onset 2 year rapid weight gain with associated growth failure
- Marked plethora and stretch marks
- Mood change/lethargy for 2-3 months
- Truncal obesity, “moon” face with facial plethora, marked purple striae in inner thighs and buttocks, hyperpigmentation of scrotum and penis

**Investigation**
- Basal cortisol: 85.8 mcg/dl (8.0-17.0)
- Basal ACTH: 59.1 pg/ml (0-46)
- 24h urinary free cortisol: 1098 mcg/24h (75-270)
- Cortisol after low dose dexamethasone test: 71 mcg/dl
- Cortisol after high dose dexamethasone test: 50 mcg/dl
- Positive CRH testing with a greater than 40% incremental rise in serum cortisol
- Brain MRI: no clear adenoma, enhancement in the right lateral portion of the pituitary
- Abdomen CT and MRI: hepatic steatosis, no other abnormalities described

**Management**
- Jan/2004: Ketonozal 200mg 3 times daily
- Feb/2004: Liver dysfunction
- April/2004: Ketonozal ceased due to liver dysfunction
- April/2004: Transferred to a specialized center
- May/2004: Ketoconazole ceased due to development of hypertension (BP: 155-95 mmHg)
- Propranolol + co-amolizide

**Follow-up**
- 2008: Significant skin hyperpigmentation
- Elevated plasma ACTH (>1250pg/mL)
- Brain MRI: 2mm pituitary microadenoma
- Conservative approach decided in order to preserve pituitary function

**Nelson’s Syndrome**
- 2010: Brain MRI: 7 mm microadenoma
- 2013: Brain MRI: 8-9 mm microadenoma
- 2016: Brain MRI: Stable, 9 mm pituitary microadenoma
- 2017: Brain MRI: Stable, 9 mm pituitary microadenoma
- 2018: Scheduled for pituitary surgery

Normal growth and pubertal development. No signs and symptoms of CD. No compressive symptoms. No pituitary deficits.

**CONCLUSION**

Treatment of pediatric CD may be challenging once the aim is to cure hypercortisolism and to preserve pituitary function, in order not to compromise normal development. Once transsphenoidal hypophysectomy is not always possible or available, bilateral adrenalectomy remains a therapeutic option in life-threatening situations. Pituitary surgery should be the first-line treatment option for Nelson’s syndrome. Despite surgical intervention, adjuvant radiotherapy may be required in some patients as progression of Nelson’s syndrome tumors may occur.