PEDIATRIC CUSHING'S DISEASE AND PARADUODENAL TUMOR

Ana Valea[1,2], Maria Breaban[2], Mara Carsote[3], Andra Morar[2], Dan Pop Dumitru[4], Carmen Emanuela Georgescu[1,2], Cristina Ghervan[1,2]

EP 70

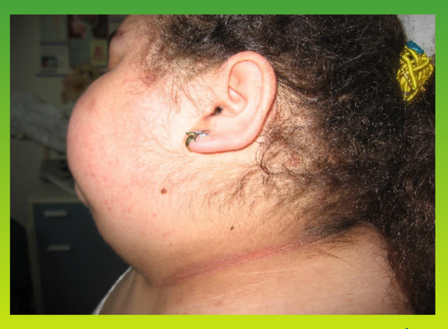
1. Iuliu Hatieganu University of Medicine and Pharmacy, Cluj-Napoca, Romania
2. Endocrinology Clinic, Cluj-Napoca, Romania
3. Department of Endocrinology Carol Davila University of Medicine and Pharmacy, Bucharest & C.I.Parhon National Institute of Endocrinology, Bucharest, Romania
4. Department of Radiotherapy, Prof. Dr. Ion Chiricuta Oncology Institute Cluj-Napoca, Romania

Introduction

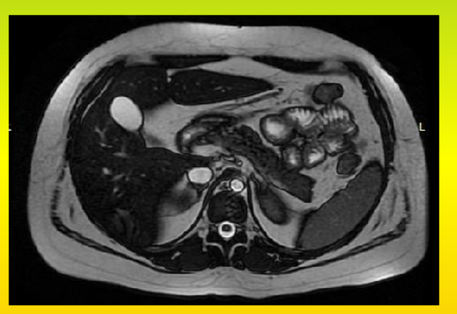
Cushing's disease (CD) in youth represents a challenge especially related to the therapy and long-term management.

Material & Methods

Suggestive endocrine panel and imagery is exposed.



Paraduodenal mass



Date	Parameter	Value	Normal Range
September 2009	ACTH	48.9pg/ml	7.2-63.3 pg/ml
	Morning plasma cortisol (8 a.m)	486.4nmol/l	172-497nmol/l
	Night plasma cortisol (8 p.m	220.1nmol/l	71.1-286nmol/l
	Plasma cortisol after 1 mg overnight DXA	347.6nmol/l	<50nmol/l
	Plasma cortisol after 2 days x 8 mg DXM	41.7nmol/l	
January 2010	ACTH	95.56pg/ml	7.2-63.3 pg/ml
	Morning plasma cortisol	404.7 nmol/l	172-497nmol/l
	Plasma cortisol after 1 mg DXA	81.2 nmol/l	<50nmol/l

Case presentation

A 19-year male was diagnosed at age of 12 with CD. Clinical assessment revealed: weight gain, headache, hyperpigmentation predominantly on the areas subjected to friction. Hormonal profile found: plasma ACTH of 48.9pg/ml (N:7.2-63.3 pg/ml), baseline plasma cortisol (8 a.m) of 486.4nmol/l (N:172-497nmol/l) and (8 p.m.) of 220.1nmol/l (N:71.1-286nmol/l), plasma cortisol after 1 mg overnight Dexamethasone (DXM) suppression test of 347.6nmol/l (N:<50nmol/l) and after 2 days x 8 mg DXM of 41.7nmol/l, suggestive for ACTH-dependent hypercortisolism. The initial pituitary MRI did not detect anomalies but a subsequent examination described a pituitary microadenoma. Adominal CT scan with and without enhancement and abdominal MRI revealed a periduodenal tumor of 12/8 mm, which raised the question of a neuroendocrine tumor which was not confirmed by pathological report based on biopsy (via superior digestive endoscopy). The patient underwent transsphenoidal surgery with selective removal of the pituitary microadenoma and persistent CS intermittently treated with steroidogenesis inhibitors and imagery follow-up of the periduodenal mass. Currently, high plasma ACTH levels of 95.56pg/ml with elevated basal plasma morning cortisol of 404.7 nmol/l and inadequate suppression after 1 mg DXM overnight of 81.2 nmol/l. Pituitary MRI was status quo and pasireotide was recommended.

Conclusion.

Despite the potential differential diagnosis with ectopic Cushing's syndrome related to the paraduodenal tumor, pediatric CS represents a rare yet severe event and the methods to achieve the disease control in this particular population are still suboptimal.