5 yr complete clinical remission after single adrenalectomy for severe occult ACTH-dependent Cushing's syndrome

Daniel Grigorie[1,2], Andra Caragheorgheopol[1], Carmen Iordachescu[1], Bogdan Stanescu[1,2], Dan Hortopan[1], Alina Sucaliuc[1,2]
[1]National Institute of Endocrinology; [2] "Carol Davila" University of Medicine and Pharmacy, Bucharest, Romania

Introduction.Complete long-term clinical remission in occult ectopic ACTH syndrome after a single adrenalectomy is unexpected.

Case report. 5 yr ago, a 54-yr-old man was admitted because of resistant HTA, multiple severe vertebral fractures, muscle weakness and Cushingoid features of at least one year.

Adrenal tests were diagnostic for ACTH-dependent Cushing'syndrome: ACTH= 263 pg/ml, high plasma (62 µg/dl) and urinary free cortisol (UFC=1256 µg/24h) with no suppression after both low-dose (cortisol= 21.3 µg/dl; UFC= 255.36 µg/24h) and high-dose dexamethasone (cortisol= 32.95 µg/dl; UFC= 1193.2 µg/24h).

A pituitary MRI showed no identifiable tumor. CTs of the thorax and abdomen and a ^{18}-FDG-PET CT scan failed to identify a possible tumor.

A right adrenal laparoscopic adrenalectomy was performed. One mo postsurgery ACTH decreased to 79.8 pg/ml and cortisol to 16.7 µg/dl, nonsuppressible with low dose DXM (7.37 µg/dl); low testosterone and 25OHD normalised, and bone turnover markers increased significantly (osteocalcin 10 x).

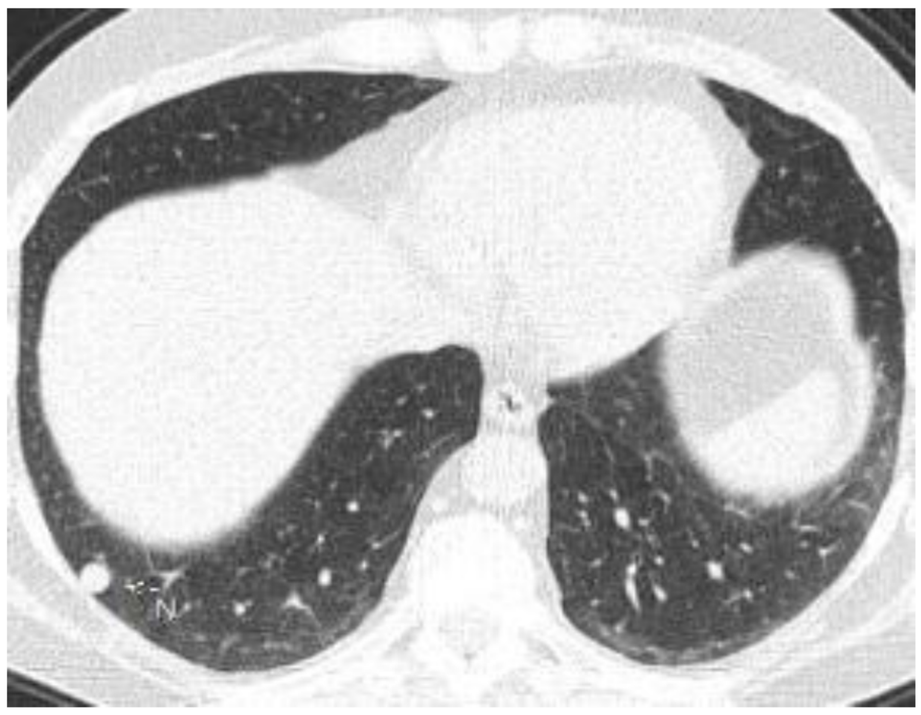
As clinical improvement was significant the patient declined the second adrenalectomy.Ten mo postsurgery all Cushingoid features disappeared and the patient was in very good clinical condition with mild HTA and normal blood chemistries as he is now.

Adrenal tests were performed annualy: they fluctuated, with serum cortisol/UFC normal or high normal but nonsuppressible (or paradoxically increased); ACTH also was around 100 pg/ml. BMD increased progressively for 5 yr (2.8%/yr at the FN).

	Baseline	1 mo postop	2 mo postop	1 yr postop	5 yrs postop
Plasma cortisol (µg/dl)	62	16.7	5.7	10.3	8
ACTH (pg/ml)	263	79.8	90.7	97.5	102.5
UFC (µg/day)	1256	213	127.4	127	94
Low dose DXM supression (µg/ml)	21.3	7.37	2.94	19.7	6.04
High dose DXM supression (µg/dl)	32.9	NA	NA	NA	44

Two years ago a CT scan revealed a small pulmonary nodule (10 mm) but a SMS (^{111}-indium-pentetreotide) scan was negative; the nodule was stable on recent CT.

We wait for the patient consent to operate the pulmonary nodule.



Conclusion:The case show an unexpected rapid and complete clinical remission of a severe occult ectopic Cushing' syndrome. The abnormal cortisol feed-back persisted for the duration of follow-up.

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