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A case of cortisol producing tumour in 14 years old girl

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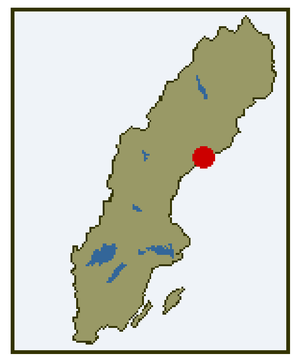


Background

Adrenocortical Carcinomas (ACC) represents 0.16% of all pediatric neoplasms in children age 0-14.9 years diagnosed between 1984-2010 according to Swedish Childhood Cancer registry[1]. International data report ACC to be most frequent in girls below 4 years of age[2].

Case

-3 years | -2 years | -1 years | -6 months | At referral

A very physically active 14 years old girl was referred due to growth velocity 0 cm over the last year. Her history revealed substantial weight gain, pubertal arrest premenarche and increased body hair.

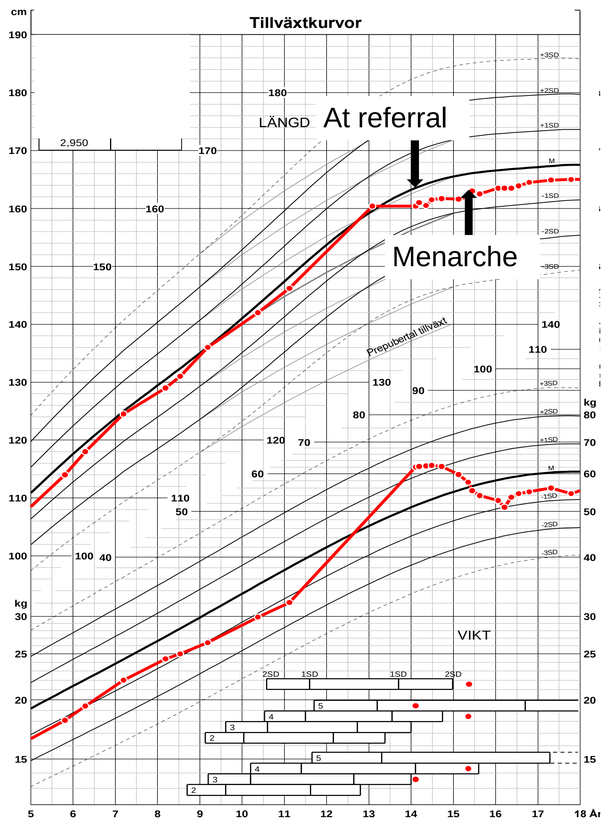
Clinical signs: Cushing Syndrome with increased abdominal fat, hirsutism, acanthosis nigricans and acne in face and shoulders. Height 160.4 cm, weight 62 kg, BMI 24.1 and pubertal stage Tanner B3, PH5.

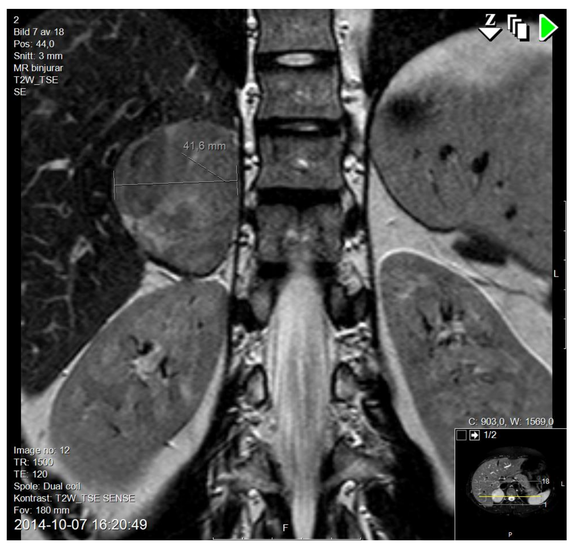
Investigation*: Serum-lab:* Cortisol 521 nmol/L, Testosterone 3.7 nmol/L (<1.8), DHEAS 15.4 µmol/L (1.8-10.0), 17-OHP 2.3 nmol/L (< 5-18).

At midnight ACTH <0.3 pmol/L (1.5-14), Cortisol 542 nmol/L (<50), without diurnal variation.

Dexamethasone suppression test 1 mg and 9 hrs later: morning S-Cortisol 632 nmol/L and Saliva Cortisol 9.85 nmol/L (<3).
24hrs U-Cortisol 1406 nmol/L/24h (<170), U-Steroid profile showed high androgen and cortisol content.

Genetic analyses for Beckwith-Wiedemann, Li-Fraumeni, MEN1 and Carney Complex that predispose for cancer development were negative.

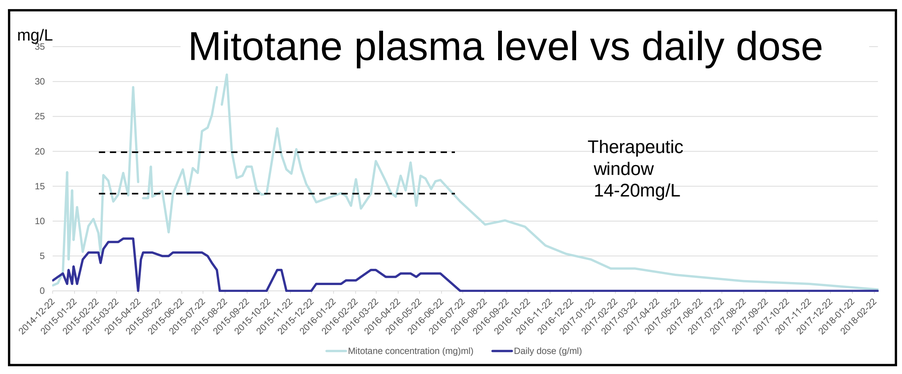




MRT of the adrenals shows a non-fatty 5x5x4 cm tumour in the right adrenal

Treatment

Surgery: The tumour was resected together with the right kidney and a part of the liver. The adrenocortical carcinoma was invading the capsule of the liver. No sign of metastasis or rest-tumour was shown by Metomidate-PET.
Chemotherapy: 8 blocks of multiple drug chemotherapy for 6 months, combined with 18 months of mitotane treatment for inhibiting steroid synthesis and for cytotoxic effect on the adrenal cortex. Hydrocortisone replacement started directly postoperative. The therapeutic window for mitotane was difficult to maintain due to side effects, especially nausea. Mitotane is stored in the fat tissue and released during weight loss, resulting in mitotane remaining in suppressive serum concentration (>0.5 mg/L) for 20 months after end of treatment.
Observation time is presently 4.5 years with no sign of relapse. Hydrocortisone substitution continues.



Pre treatment

2.5 years post surgery

Summary: A case of non-syndromic adrenocortical carcinoma in a 14 years old girl presenting with growth failure and Cushing syndrome.
Monitoring of growth velocity was the crucial factor for the patient to contact health care – patient monitoring of growth may improve health surveillance.
Mitotane treatment was challenging with the substance still detectable in suppressive serum concentration during 20 months after end of treatment.

All photos are published with patient's concent

References: 1. Swedish Childhood Cancer Incidence and Survival in Sweden 1984-2010, Report 2013 from the Childhood Cancer Registry
2. Michalkiewic E. et al. Clinical and outcome characteristics of children with adrenocortical tumors: a report from the International Pediatric Adrenocortical Tumor Registry. J Oncol. 2004 Mar 1;22(5):838-45.

Disclosures: The authors have no disclosures

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