Slow progressive puberty and secreting adrenocortical tumour in a teenager

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INTRODUCTION

Secreting adrenocortical tumours (SACT) are rare in childhood and present with variable signs depending on the type of hormone excess. We describe a teenager with ACT presenting with slow progressing puberty.

CASE DESCRIPTION

A pre-menarchal 13.5 years old girl with a high BMI (28kg/m2), presented with slow progressing puberty. She had started her puberty at least three years previously with breast changes followed by pubic and axillary hair over two years before presentation. Her Tanner staging for puberty was B4,P4 without virilisation, acne and Cushinoid features. There was a strong family history of PCOS and Type2 Diabetes. Investigations were done to rule out PCOS.

RESULTS

- **Blood investigations:**
  Testosterone 3.6 nmol/L (0.2 – 2.9)
  DHEAS 27 µmol/L (1.8 – 10.0)
  17 OH progesterone 3nmol/L (1-10)
  Androstenedione 8.9 nmol/L (2-12)
  Oestradiol 98 pmol/L
  LH 1.9 U/L , FSH 12 U/L.
- Abdominal ultrasound: Left sided adrenal mass
- MRI scan of the abdomen: Well defined left sided adrenal mass
- 24 hour urine steroid profile: Pattern suggestive of a secreting ACT with raised dehydroepiandrosterone and pregnanetriol.

MANAGEMENT

At Surgery:

- A well defined left sided adrenal mass was removed with intact capsule.
- Tumour size was 125x95x75mm, weight 585g.
- Histology showed uncertain malignant potential with the presence of 3 potentially malignant absolute histological criteria (size, weight and presence of necrosis).
- The patient was started on hydrocortisone replacement.

FOLLOW UP

- Hydrocortisone replacement was weaned over three months.
- She developed light menstrual bleeding one month after surgery but no further menses afterwards.
- Following life style modifications her BMI improved (27.5kg/m2)
- PCOS was confirmed biochemically and by ovarian US morphology at the age of 15.5.
- At 2 year follow-up, there were no biochemical/radiological evidence of tumour relapse.
- She has recently developed glucose intolerance and started on Metformin. Current BMI is 27.4kg/m2.
- She is postmenarchal but her periods are irregular

DISCUSSION

- Adrenocortical carcinoma is a rare disease but one of the most aggressive endocrine tumors with a very poor prognosis.
- Distinction between adenoma and carcinoma is difficult and has historically relied upon tumour size and imaging, or histopathological evidence of local invasion, necrosis, number and presence of atypical mitoses or metastases.
- Smaller tumours (<10.5 cm or <400g) are more easily completely resected and hence have a better prognosis while larger tumours are less likely to be resected and have a poor prognosis.
- An early diagnosis of ACT is crucial but often delayed because of atypical presentation.
- In pre-menarchal / adolescent girls a diagnosis of PCOS can be challenging and on-going monitoring is needed to confirm it. In the differential diagnosis, even in the absence of virilisation, secreting ACT should be considered.

CONCLUSIONS

- This case illustrates a secreting ACT with lack of virilisation in a pre-menarchal teenager with slow progressing puberty and strong family history for PCOS.
- Awareness of this condition should be raised and be considered in the differential diagnosis of PCOS