Rare causes of paediatric virilizing tumours

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Introduction

Excessive androgen secretion from gonads, adrenal gland and tumors arising from germ cells lead to gonadotropin independent precocious puberty in male and virilization in females. Rapid progression of symptoms with peripheral precocity need urgent evaluation to identify the underlying etiology. We report three cases of malignancy with excess androgen secretion.

Case 1

4 year old boy presented with iso-sexual precocious puberty, cushingoid feature, progressive abdominal distension
And generalized acne for six month duration.

Examination

- Wt 19.5 kg (90th 97th)Ht 107.5 cm (75th 90th)
- Adult body odor, Greasy skin, Acne
- Hirsutism, no gynaecomastia
- BP 114/69mmhg (99th)
- No hepatomegaly, left side palpable mass, no free fluid
- Axillary hair +, Pubic hair III, Phallus 8.5cm good width
- Testicular volume R/ 6ml, L/6-8ml

Investigations

USS abdomen, CT abdomen and pelvis
L/ supra renal mass with midline shift of adjacent structures. No distant metastasis.

Management.

Surgical resection of the tumour and followed up with regular USS abdomen and DGEAS

Bone age	9 years
ODST(<50)	562.3nmol/l (<50)
Testosterone	61.05nmol/l
DHEAS (0.9- 5.8)	>40.71µmol/l
LH (0.08-3.9)	0.07IU/l
FSH (0.1-1.3)	0.06IU/l

Diagnosis
Adrenocortical carcinoma

Case 2

13 year old girl presented with virilization for 6 months

- Hirsutism, deepening of voice and facial acne

Examination

- Hirsutism, boldness, acne, masculinization, deep voice
- Not pigmented
- Bp110/80mm/hg
- Abdomen soft
- Breast stage 1, axillary and pubic hair stage 111, clitoromegaly 2.5cm

Investigations

USS, MRI -Abdomen and pelvis
Large soft tissue lesion with multiple cystic areas in the R/ ovary

Management

underwent R/salpingo oophorectomy
Intra operatively found to have 1cm diameter lesion on L/ ovary as well wedge biopsy was taken.
Referred to oncologist for further follow up and planned for L/oophorectomy

α FP	109ng/ml
β HCG	< 0.7mIU/ml
DHEAS	3.3 µmol/L
T. Testosterone	23.82nmol/L
LH	4.14IU/L
FSH	1.63IU/L
S. Cortisol	169.7 nmol/L

Diagnosis
Bilateral Sertoli Leydig cell tumour of ovaries

Case 3

8 year old boy presented with deepening of voice penile and axillary hair growth and increase height velocity for 1 month duration

Examination

- Not dysmorphic , not pigmented, no café au lait spot , no boney deformities
- RS – Nl
- BP 107/73mm/hg
- Abdomen – Soft no palpable masses
- Axillary hair I, pubic hair stage II
- B/L testis 3-4ml
- Penile length 9.5cm

Investigation

Chest X ray –no widening of mediastinum
CT and MRI abdomen - Normal
MRI brain - Normal
CT chest reveled - Anterior mediastinal mass

Management

As frozen section biopsy was inconclusive underwent total resection of tumour and received cisplatin based chemotherapy

X ray bone age	**5-6 years**
LH	<0.07 IU/L
FSH	<0.05 IU/L
17 OHP	8.17 nmol/l
Testosterone (0.1- 1)	5nmol/l
βHCG	261.9mIU/l

Diagnosis
Seminoma without other evidence of germ cell components

Conclusion

Rapid progression of virilization need urgent evaluation for androgen secreting tumours with imaging and tumour markers. Need long term follow up with frequent imaging and tumor markers to identify early tumour recurrence and appropriate treatment.