UNVEILING THE UNUSUAL: AN INSIGHT INTO PEDIATRIC ADRENOCORTICAL TUMOURS

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Introduction

- **Adrenocortical tumors(ACTs)** are extremely rare in children, comprising **0.2%** of all pediatric neoplasms with annual incidence of **0.3-0.38 per million children**[1]
- Peak incidence seen in children under **5 years** with higher prevalence in **females**[1]
- Most common presentation is **virilization**(characterized by linear growth acceleration, advanced bone age, pubic hair, hirsutism, acne, deep voice, muscle hypertrophy, clitoromegaly or increase in penis size)followed by Cushing's syndrome[2]

Case report

CLINICAL PRESENTATION

9 month old female child presented with a progressive perineal swelling & hair on pre pubic region.

CLINICAL EXAMINATION

- **Clitoromegaly**-2.4cm(Prader stage1);hair on labia &pre pubic region;moon facies.
- P/A-5x5 cm mass in right lumbar region.
- Serial BP monitoring- Elevated BP

INVESTIGATIONS

- Elevated serum **DHEA-S(4628ug/dL)** & testosterone(13ng/ml); cortisol(no diurnal variation-8ug/dL);LH&FSH(<0.3mIU/ml)
- Serum **ACTH** levels suppressed(**3pg/ml)**
- **Dexamethasone suppression test**-No decrease in cortisol.
- **CECT abdomen**-5.2x5.7x5.2 cm well defined, heterogeneously enhancing lesion;multiple prominent vessels ;non enhancing necrotic areas,present in right suprarenal region.
- **Bone age assessment**(left hand Xray):wrist-1.5 years; phalanges-2years

Management

- Pheochromcytoma ruled out before surgery
- **Exploratory laprotomy**-Right adrenal gland excision;sent for histopathology.
- **HPE**-oncocytic variant of adrenocortical neoplasm of uncertain malignant potential (**Wieneke's index** score-3) and staged as **pT2**(pTNM AJCC 8th edition)
- **Post op**:Serum levels of androgens and cortisol were normal soon after surgery

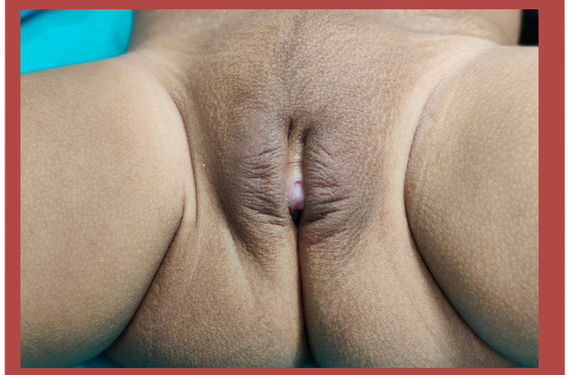

Fig1. Clitoromegaly & hair on labia & pre pubic region

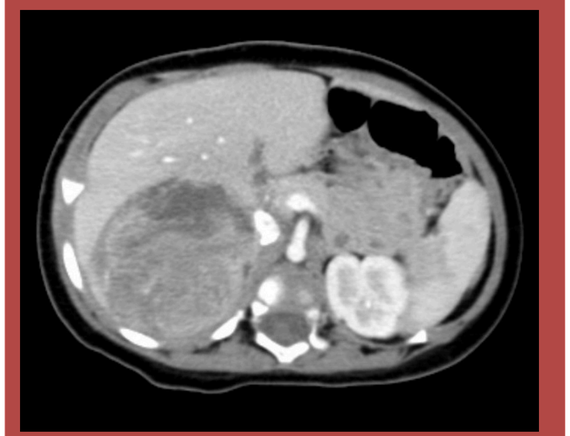

Fig2.CECT Abdomen(axial view)

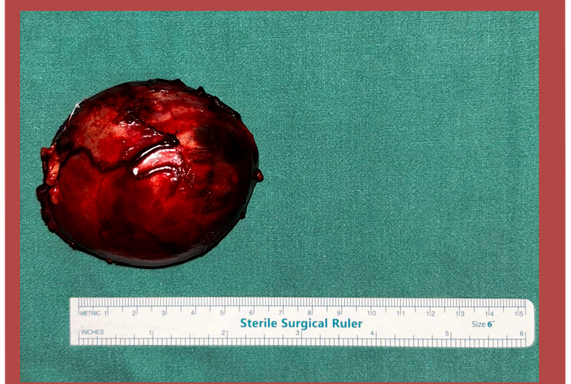

Fig3. Gross specimen of tumour post resection

Discussion

- Complete tumor resection(**R0**) remains the cornerstone of treatment and offers the best chance of cure [3]
- **Postoperative follow-up**:Imaging & endocrine workup (DHEA-S, testosterone, and cortisol) every 3 months in the 1st year,4 months in the 2nd year,6months in the 3rd year & once a year from 4th year to monitor for any recurrence.
- **Bone age assessment** should be done once a year before puberty & every 6 months during puberty until epiphyseal closure.
- **Mitotane** is the only drug approved for ACTs with malignant behaviour (adjuvant therapy)[4]
- **Radiotherapy** is not an option as many children with ACT carry germline TP53 pathogenic allelic variants predisposing them to cancer & radiation may increase the incidence of secondary tumors[5]
- **P53 gene** mutations are the most frequently inherited genetic abnormalities linked to a higher incidence of ACTs in familial cancer syndromes. This can be best inferred from the founder effect of TP53 gene mutation in the population of brazil having 10-15 times the annual worldwide incidence of ACTs[6]. Hence **genetic testing** is important to screen for other associated cancers.
- **CT Thorax** indicated in all patients(Lungs & liver:Most common site of metastasis)[7]
- **Delayed diagnosis** poses the risk for:premature epiphyseal fusion,causing **short stature**;permanent **voice** changes;progressive clitoromegaly;upstaging of tumor.
- **Differential diagnoses**:Congenital adrenal hyperplasia; exogenous androgen exposure;Mc Cune Albright syndrome;Glucocorticoid resistance.

Conclusion

All children under 5 years presenting with **virilizing or cushingoid** features should be evaluated for ACTs inspite of them being exceedingly rare.**Early diagnosis** with imaging and biochemical tests remains the cornerstone for a good prognosis before **irreversible pubertal changes** occur. A **multidisciplinary approach** including pediatric endocrinology, pediatric surgery, pediatric hematoncology & genetic testing department is required for planning the best course of treatment.

References

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