The management of adrenal cell carcinoma in a single tertiary centre: A 25 year experience

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Background

Adrenal cortical carcinoma (ACC) in children is rare and aggressive, with the mainstay of treatment being surgical resection. Although, there have been recent improvements in outcomes with chemotherapy, the prognosis is still quite poor and often patients present with advanced disease. Focus on endocrine management has not been previously described.

Objective

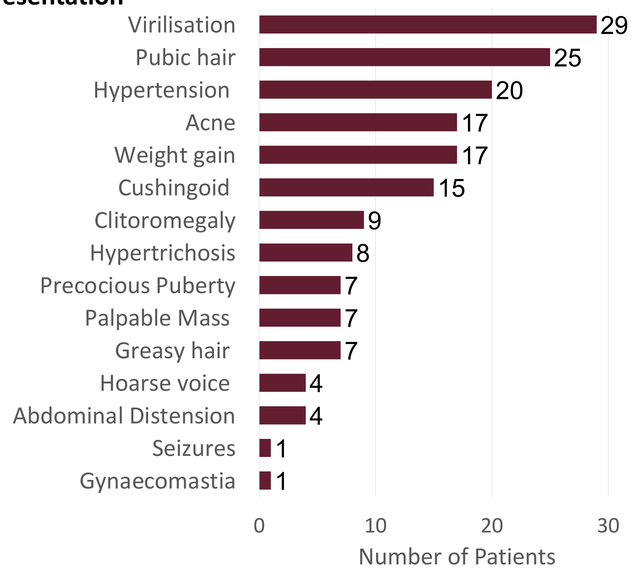
- Further characterise
  - Presenting features and biochemical markers to support earlier diagnosis
  - Strategies to manage refractory hypertension
- Describe longer term endocrine management in this cohort

Methods

Retrospective review of patient records at a single centre between 1996 and 2021 of patients with an ACC.

Results

Presentation



Results

Management	Detail	
Refractory Hypertension	Metyrapone (n=3)	Ketoconazole (n=3)
Surgical Resection	All Patients Evolved overtime to become laparoscopic	
Adjuvant Chemotherapy	32% of patients received	
	Mitotane (n=8)	
Hydrocortisone	Most treated with IV infusion All were discharged on hydrocortisone replacement (10mg/m^2/day; duration 0.1-8y, median 1y) Those treated with mitotane required higher doses (15-17mg/m^2/day)	
Fludrocortisone	3 patients (all associated with mitotane)	
Thyroxine	2 patients (all associated with mitotane)	

The mortality rate was 32% in this cohort, although comorbidities were contributing factors in at least two patients.

Results

34 patients

Age at diagnosis: 2 weeks – 10 years

Median 2.35 years

17 Female

17 Male

Malignancy Risk (Histology and Size)

High (n=17)
Borderline (n=2)
Low (n=15)

Genetic Mutation identified in 38%

TP53 mutation (n=10)
n=8 high risk

Mutation associated with *Beckwith Wiedemann syndrome* (n=3)
n=1 borderline n=2 high risk

Investigation		Result
Androstenedione ↑		88%*
DHEA-S ↑		61%*
Testosterone ↑		93%*
Cortisol	Cortisol ↑	95%*
	Loss of diurnal variation documented	33%*
Urine Steroid profile	Pre-surgery	Elevated
	Post-surgery	Normal

* Percentage of cases

Key messages

- The most frequent presentations are virilisation, hypertension, cushingoid appearance, and rarely gynaecomastia
- This highlights the essential role of genetic diagnosis in influencing disease progression and treatment
- Hydrocortisone
  - Our practice has evolved to include post-operative intravenous hydrocortisone infusion in all patients to mitigate the effect of the rapid fall in cortisol
  - The duration of requirement in this cohort was highly variable
- Future Research Direction
  - Newer agents such as pasireotide or mifepristone to decrease cortisol concentrations in those with refractory hypertension, as they may have fewer side-effects and reduce morbidity

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