Pediatric Adrenocortical Tumors (PACT)

A single tertiary center experience: Clinical, Biological and Pathologic Characteristics Analysis

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Authors have nothing to disclose.

Background

PACT are primary tumors of the adrenal cortex, with heterogeneous presentation and varied behaviors in terms of progression. Generally diagnosed by clinical manifestations linked to hormone excess. They are more frequent in females with maximal incidence between 1-4 years of age. Complete tumor resection is the only intervention that provides the best chance for cure. Efficacy of adjuvant treatment in advanced disease (stages III and IV) is undetermined.

Aim

To evaluate the demographic, clinical, biochemical, and pathologic characteristics in a cohort of PACT in a single tertiary institution of Argentina.

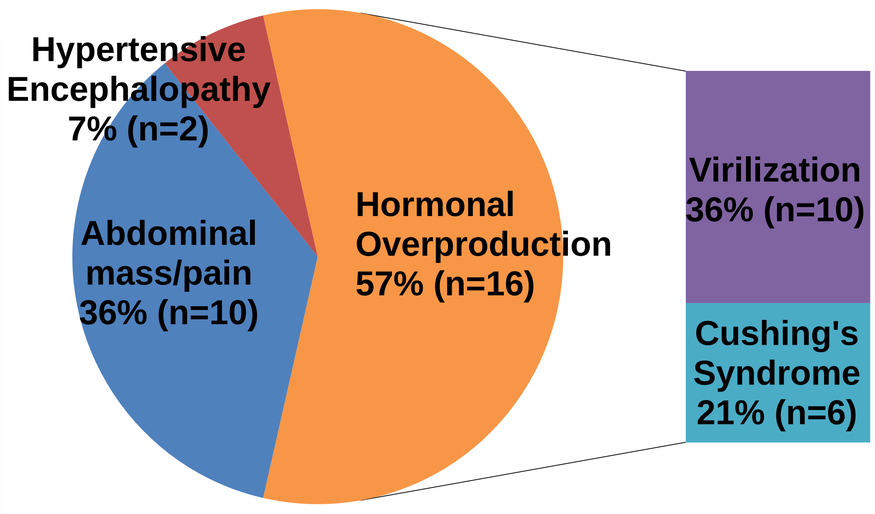
Subjects and Methods

Retrospective review of 28 medical records of PACT (chronological age (CA) <18 years (y) treated between 1987-2017. Data analized were: Demographical, clinical (Height-SDS, BMI-SDS, and bone age (BA), biochemical (serum DHEAS levels), and histological features (Wieneke index[1]). Staging (ST) according to COG system[2], and therapeutic interventions (surgery and adjuvant chemotherapy, Achemo).

Results

Demographical distribution of patients

Clinical complaint at diagnosis



Mean duration of symptoms was 10.9 months.

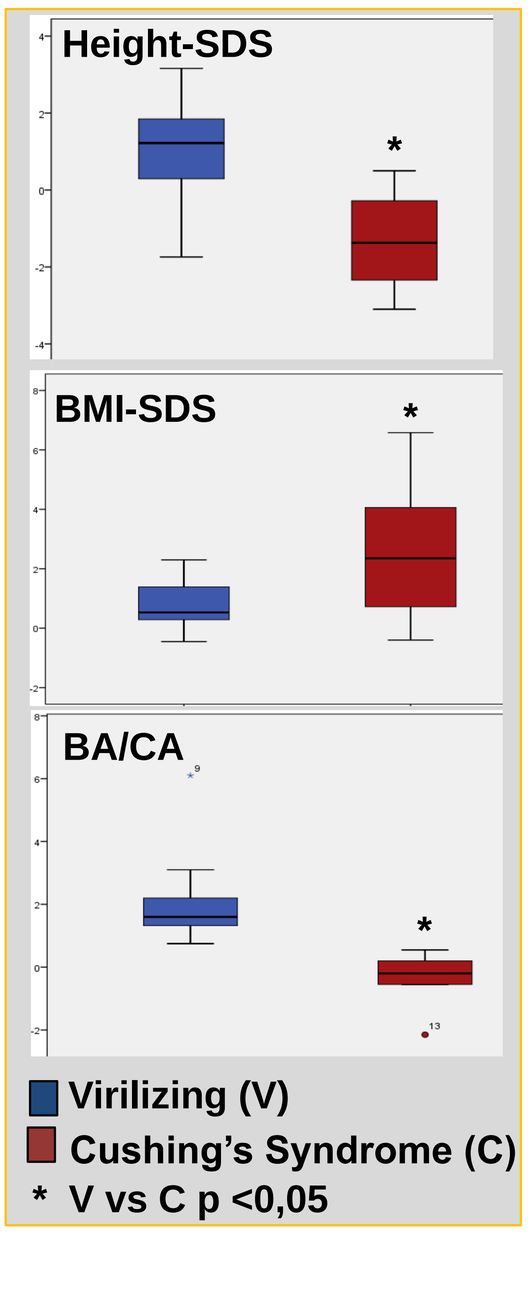
Distribution by age and sex

Feature	n	%
CA, years (median 2.8y)		
< 5	21	75%*
5 - 12	4	14.3%
13 - 18	3	10.7%
Sex		
Female	20	71.4%*
Male	8	28.5%
Female-male ratio, by age		
< 5	2,5:1	
5 – 12	1:1	
13 – 18	3:0	

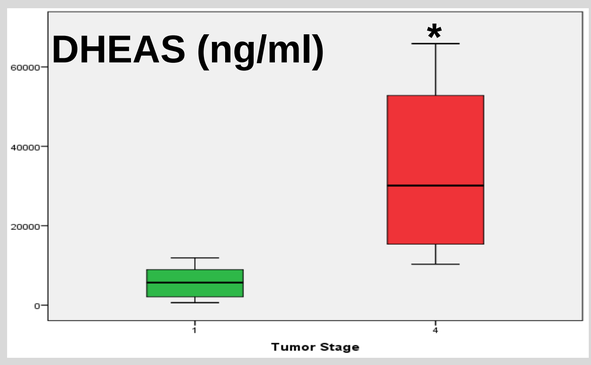
*X^2 p <0.001

Antropometric features according hormonal predominance

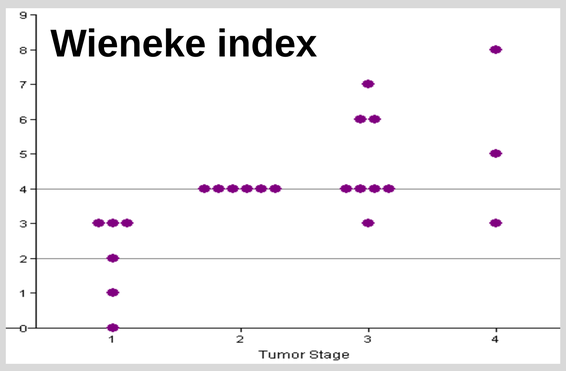
Clinical Features	n	%
Virilization	16	57.1%
Cushing's Syndrome	10	35.7%
Mixed	2	7.1%



Biochemical and Pathological features according to COG staging

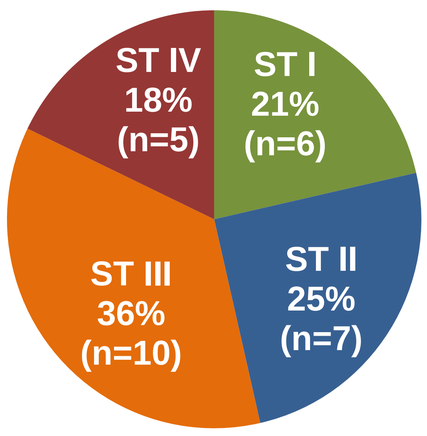


*DHEAS ST IV vs I p <0.05



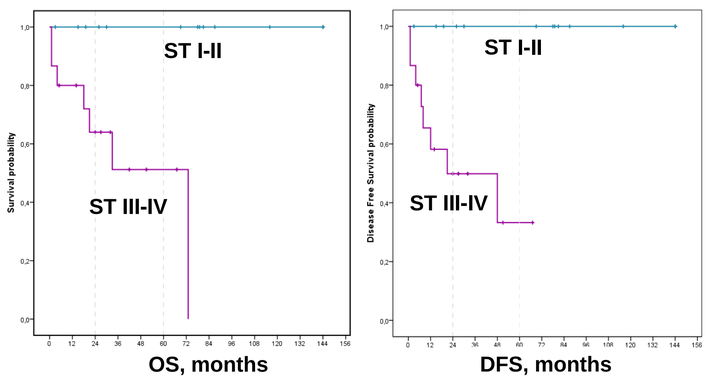
*X^2 p 0.04

COG Staging



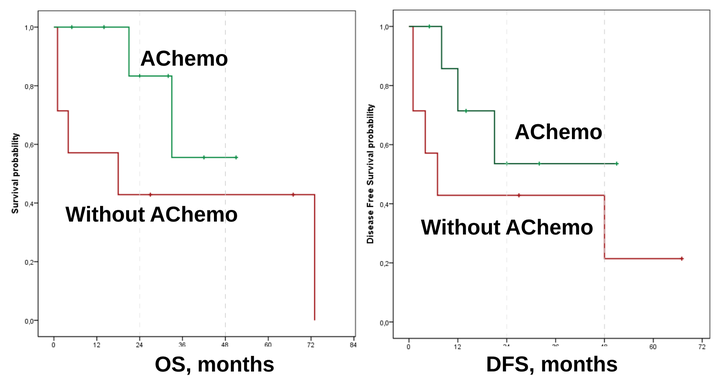
Total adrenalectomy: 26/28 patients.
Achemo: 8/15 patients. (cisplatin, etoposide, and doxorrubicin)
Median follow up: 3.64y. (range 0-12y)

Overall (OS) and Disease Free Survival (DFS) according to COG staging



5-year OS and DFS 100% for ST I-II vs 51% (95%CI 21-82) and 33% (95%CI 1.2-65) for ST III-IV respectively. LogRank p 0.002.

Overall and Disease Free Survival according to AChemo



A tendency of higher 2-year OS and DFS on AChemo (n=8; 83%, 95%CI 53-112 and 53%, 95% CI 14-92 respectively) vs without AChemo (n=7, 43%, 95%CI 6-80) was found. Log Rank p NS.

Conclusions

We reported the experience in our cohort of 28 PACT seen in a single center of Argentina over 30 years. Height-SDS and BMI-SDS mirror ACT hormonal secretion. Very high serum DHEAS levels might be use as a biological marker of tumor stage. Less advanced disease were associated with best patient outcomes. Long term follow-up is needed to draw valid conclusions of using AChemo.

References:

1 Wieneke JA, Thompson LD, Heffess CS. Adrenal cortical neoplasms in the pediatric population: a clinicopathologic and immunophenotypic analysis of 83 patients. Am J Surg Pathol. 2003 Jul;27(7):867-81

2 Ribeiro RC, Pinto EM, Zambetti GP, et al: The International Pediatric Adrenocortical Tumor Registry initiative: Contributions to clinical, biological, and treatment advances in pediatric adrenocorticaltumors. Mol Cell Endocrinol 2012, 351:37-43,.

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Adrenals and HPA Axis

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