ADRENOCORTICAL TUMOURS IN CHILDREN: AIIMS EXPERIENCE

Gayatri Munghate,
Sandeep Agarwala, M. Srinivas,
Minu Bajpai, Veereshwar Bhatnagar,
Devendra K Gupta

Department of Pediatric Surgery,
All India Institute of Medical Sciences,
New Delhi, India
Adrenal Tumours: Introduction

- Adrenal cortical neoplasms are rare
  - Incidence < 0.2% of all pediatric tumours
  - 6% of all adrenal tumours are adrenocortical tumors
- Tumours are functional and aggressive
ADRENAL TUMOURS: AIMS

- To review the presentation, management and outcome of children with adrenocortical tumours
ADRENAL TUMOURS: MATERIALS AND METHODS

- Retrospective review
- 1992-2010
- All children diagnosed with adrenocortical tumours and enrolled in solid tumor clinic at AIIMS were included
- Records were reviewed
- Presentation, management, and outcome were studied
ADRENAL TUMOURS: RESULTS- 20 PATIENTS

- Age range
  3 - 132 months
  Median – 36 months
- Right : Left  9:11

AGE DISTRIBUTION

- <5 YEARS
  11 (55%)
- >5 YEARS
  9 (45%)

SEX DISTRIBUTION %

- BOYS (11)
- GIRLS (9)
ADRENAL TUMOURS: PRESENTATION

- Weight gain: 13 (65%)
  - 4 had pain in abdomen also
- Chest pain: 1 (5%)
- Fever: 1 (5%)
- Abdominal mass: 1 (5%)
Adrenal Tumours: Presentation

- Precocious puberty and hirsutism: 2 (10%)
- Virilization: 13 (65%)
Adrenal Tumours: Presentation

- Cushing’s syndrome: 18 (90%)
  - Virilisation: 13
- Hypertension: 13 (65%)
- Abdominal mass: 11 (55%)
- Liver metastasis: 2 (10%)
- IVC thrombus: 3 (15%)
**Adrenal Tumours: Management**

n=20

- **No surgery** (4 patients)
  - Deaths 3
  - 1-IVC thrombus

- **Surgery** (16 patients)
  - Survivors 13
  - Deaths 3
  - 2-IVC thrombus
    - 1-Recurrence
ADRENAL TUMOURS: RESULTS N= 20

- Two (10%) had liver metastasis at presentation
  - One of them also underwent hepatic resection
- Three (15%) had IVC thrombus
  - Two resection of adrenal tumor
    - One post-op chemotherapy- Carboplatin + Etoposide
  - One died pre-operatively
- Three patients (15%) developed recurrences
  - All local recurrences
    - One had pulmonary recurrence also
  - 1 expired
  - 2 discontinued therapy
Adrenal Tumours: Overall survival

- n = 20
- Deaths: 6
- All deaths occurred at 1-3 months of diagnosis
- Median survival 3 months

OS 67.53% (95CI 40.95-84.15)
Adrenal Tumours: Overall survival - Age group

- Patients divided in two groups based on age
  - Age < 5 years: 11
  - Age > 5 years: 9
- p = 0.25 (not significant)
ADRENAL TUMOURS: OVERALL SURVIVAL - IVC THROMBUS

Kaplan-Meier survival estimates, by thrombus

No IVC thrombus (n = 17):
OS = 79%

IVC thrombus (n = 3):
OS = 0%
P = 0.0003
ADRENAL TUMOURS: REVIEW OF LITERATURE

- Rare tumours in children – 0.2%
- Slight female preponderance -1.4-1.9:1
- 95% - endocrine dysfunction
- 50% have palpable mass
  - often not the presenting complaint
- Patients often seen at advanced stage

ADRENAL TUMOURS: REVIEW OF LITERATURE

- Nonfunctional tumours are rare
- Aggressive course
- Surgery –
  - Primary modality of treatment
  - Only chance of cure
- Lack of effective chemotherapy
  - Ongoing trials of new chemotherapy regimen

Adrenal Tumours: Review of Literature

- Factors associated with poor prognosis*
  - Older age at diagnosis
  - Stage 3 to 4 disease
  - Cortisol hypersecretion
- Survival rate >67% if complete resection done
- Median survival – 21 months**

*Veytsman I et al, J Clin Oncol 2009; 27:4619-4629
ADRENAL TUMOURS: CONCLUSIONS - CURRENT STUDY

- Majority (90%) presented with endocrine dysfunction.
- Palpable mass is seen in 55% of patients at presentation.
- 10% had metastasis at presentation.
- Overall outlook is dismal
  - Overall survival 67%
  - Median survival-3 months
- Presence of advanced disease at presentation associated with poor prognosis