


Adrenal Cortical Neoplasms



Adrenal Cortical Neoplasms



- Adenoma
- Carcinoma

Adrenal Cortical Carcinoma



- distinguished clinically by:
 - size (> 6 cm)
 - metastatic behavior
- both carcinomas and adenomas may be hormonally active (Cushing's syndrome)

Adrenal Cortical Carcinoma: Pathologic Features

- Size (>100 gm)
 - Median weight in published series:
 - 510 gm, 714 gm, 849 gm, 1190 gm, 1210 gm
- Mitotic activity
- Necrosis
- Grossly variegated appearance, cystic change
- Capsular or vascular invasion
- Involvement of adjacent organs
- Nuclear pleomorphism less reliable (seen in both benign and malignant tumors)

Adrenal Cortical Carcinoma: Incidence

- 1 per 1 million people
- Female:Male 1.5:1
- Most common in 5th decade
- Smaller peak in 1st decade
- Bilaterality rare,
 - but adrenal mets in up to 15%

Adrenal Cortical Carcinoma: Clinical Presentation

- abdominal or flank pain
- discomfort
- fullness
- 30% with palpable mass
- weight loss

Adrenal Cortical Carcinoma: Clinical Presentation

- intermittent low grade fever
 - related to tumor necrosis
- hypoglycemia
- hypercalcemia
- up to 67% with metastasis at diagnosis

“FUNCTIONING” ADRENAL CORTICAL CARCINOMA

- abnormal steroid production
- 26 to 76% in various series
- “pure” syndrome, such as Cushing’s
- “mixed” endocrine syndrome or any biochemical evidence of steroid excess
- various enzymatic dysfunctions
- loss of sensitivity to ACTH
- feminization uncommon (gynecomastia)
- hyperaldosteronism very uncommon

Adrenal Cortical Carcinoma: Radiologic Appearance

- Usually large
- Low intensity areas correspond to necrosis
- Calcification
- Irregular contrast enhancement
- MRI: heterogeneous T1-weighted images
 - signal intensity $>$ fat on T2-weighted images

Adrenal Cortical Neoplasms: pregnancy-related issues

- association with pregnancy is rare
 - usually Cushing's syndrome patients are infertile
- Cushing's syndrome in pregnancy associated with:
 - fetal wastage, prematurity, adrenal insufficiency, virilization, maternal hypertension, diabetes, pre-eclampsia, pulmonary embolism
- if Cushing's syndrome develops during pregnancy:
 - majority (16/22) associated with adenoma(8), or carcinoma (8) (Guilhaume et al, France, 1992)
- Cushing's disease observed rarely (4 pts, 2 with ectopic ACTH)

Adrenal Cortical Carcinoma: Prognosis



- Mortality Rates
 - 2 yr 50%
 - 5 yr 84%
 - 10 yr 90%

Adrenal Cortical Carcinoma: Sites of Regional and Distant Metastasis (50 Autopsies, NIH)

Liver	92%	Peritoneum	16%
Lung	78%	Kidney	12%
Retroperitoneum	48%	Diaphragm	12%
IVC (obstruction related edema, ascites)	28%	Heart	12%
Intestinal serosa	22%	Spleen	8%
Lymph nodes		Pancreas	8%
Abdominal	32%	Thyroid	6%
Thoracic	26%	Brain	4%
Bone	18%	Skin	2%

Adrenal Cortical Carcinoma: Prognostic markers

- Mitotic index
 - $< 20/50$ hpf = low grade (58 mo median surv.)
 - $> 20/50$ hpf = high grade (14 mo median surv.)
- Proliferation markers (Ki67, PCNA, S-phase)
- DNA ploidy, more common in carcinomas than adenomas
- Multiple aneuploidy
- Epidermal growth factor receptor
- c-myc

Childhood Adrenal Tumors

- 92 % Neuroblastoma
- 6% Adrenal cortical carcinoma
- 2% Pheochromocytoma
- Estimated incidence for ACA in children (< 20 yr)
 - 19 new cases/yr
 - 3 cases/1 million children/yr
 - 0.3% of childhood neoplasms
 - 6% of all ACA
- Most have virilization, Cushing's (usually mixed)
 - "herculean" habitus, deepening voice
 - clitoromegaly, facial and pubic hair