Clinical Pathological Correlation
Case #1

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Case Presentation – History

• **ID**
  - 24 yo M
  - Referred from GP to Hematologist for polycythemia

• **PMHx**
  - Depression – started Tx last year

• **PSHx**
  - Negative

HPI

- Recent trip to India in February 2004
  - Was well throughout trip, no sick contacts
- Decrease 
- No HA
- No visual changes
- Rare dizzy spells
- Intermittent paresthesias in fingers

SHx

- No injectable steroids
- Weekend binge drinker
- Smokes < 1 ppw
- NO illicit drugs

History Cont’d

- **FHx**
  - Dyslipidemia
- **Meds**
  - Bupropion x 1 yr
- **ALL**
  - NKDA

Physical Exam

- Healthy slim young man (24 yo)
- No lymphadenopathy
- Abdomen soft, non tender, no masses
- No HSM
- CVS & RESP exams N
- O2 saturation 98% RA
Investigations

- Hb 207 [135-175]
- WBC 11.5 [4.0-11.0]
  - Differential N
  - No eosinophilia
- Plt 203 [125-350]
- INR 1.24 [0.9-1.20]
- Lytes N
- Cr N

Impression

- Polycythemia NYD

Plan

- EPO level
  - Rule out occult EPO secreting tumors
- Abd US
- CT Head
- Phlebotomy for 400 cc of blood for symptomatic improv't w target Hb of 160

Polycythemia

- HCT > 54% M
- HCT > 51% F
- Absolute polycythemia
  - Incr in RBC mass
- Relative polycythemia
  - Decr in plasma vol

Symptoms – Polycythemia

- HA
- Blurry vision
- Dizziness
- Strokes
- Cardiac ischemia
- Peripheral thromboses

DDx of Polycythemia

- Primary
  - Polycythemia rubra vera
  - Primary congenital and familial polycythemia
- Secondary
  - EPO secreting tumor
  - Chronic hypoxia
  - Carboxyhemoglobinemia
  - Cushing's syndr
  - Corticosteroids, exogenous androgens
  - Altitude
  - R to L shunt
CT Head

- Normal

Radiology

Abdominal US
**Imaging Summary**

- **Liver**
  - Segment 6
  - 3 x 4 x 5 cm multiloculated cyst with thickened septa
  - ? old (post-infectious) vs hydatid cyst

- **RK**
  - 2.5 x 3 x 3 cm echo poor mass interpolar cortex
  - Mild enhancement on CT
  - Solid hypovascular mass w Doppler

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**CT Abdomen - Summary**

- **Liver**
  - 4 x 2.5 cm multiloculated cystic lesion segment 6
  - Thick septations - no solid component
  - Unlikely to be abscess re. healthy presentat’n
  - Findings suspicious for Echinococcus (travel Hx)

- **RK**
  - Solid lesion RK measuring 3.5 x 2 cm
  - Incr enhancement
  - R renal vein N
  - No LN’s seen
  - Lesion extends into renal sinus
DDx

- Solid renal mass
  - Malignant
  - Benign
  - Infectious
  - Inflammatory
- Liver Cyst
  - Hydatidiform

Echinococcus

- Tissue infect'n d/t *Echinococcus granulosa*
- Uncommon in Canada
- Present in
  - Central Asia
  - Middle East
  - Mediterranean
  - S. America
  - US - AZ, CA, NM, UT

Echinococcus - Risk Factors

- Exposure to
  - Intermediate hosts
    - Cattle, sheep, pigs, deer
  - Found in feces of the definitive host
    - Dogs, wolves, coyotes
  - Rare infection in US, rarer in Canada

Special Tests

- Right kidney FNA
  - Negative for
    - PMN’s, organisms, Gram stain, AFB, Fungus

- Liver segment 6 FNA
  - Negative for
    - PMN’s, organisms, Gram stain, O&P, AFB, Fungus
    - Few lymphocytes & macrophages present
**Special Tests**

- Renal Core Bx
  - Negative for RCC
  - Diagnostic for metanephric adenoma
    - Fat, possibility of other tumors cannot be excluded based on small sample size
- DDX
  - Adult Wilms’ tumor
  - Metanephric adenofibroma
  - Low grade papillary RCC


**Urologic Management**

- Open Right Radical Nephrectomy
  - Subcostal approach off tip of 12th rib
  - Extension of mass into renal sinus prevented partial Nx
  - Adrenal sparing

**Gross Pathology**

- Metanephric adenoma
  - 4 x 3 x 2 cm
  - Tumor confined to kidney
  - Perinephric fat, pelvi calyceal urothelial mucosa not involved w tumor
  - No vascular or lymphatic invasion
  - Reactive fibrosis w/in tumor c/w Bx site

**Final Pathology**

**Metanephric Adenoma**

- 1995
- 2005
Metanephric Adenoma

- Newly described tumor
- Benign biological behaviour
- Described in 1995: Jones EJ et al, Davis CJ et al.
- Rare: 80 well documented cases in literature (1999)
- Occurs in children and adults: M:F 1:4

Symptoms (SSx)
- Flank pain
- Gross hematuria
- Palpable mass

Labs
- Polycythemia
- hyperCa

Microscopic Findings
- Bland architecture – rare mitoses
- Resembles epith component of Wilms’
- Extremely cellular
- Basophilic epithelial cells
- Lacks fibrous interface w/ adjacent renal parenchyma
- High nucleus:cytoplasm ratio

Gross Pathology
- Size 0.3-15.5 cm
- Tumor may regress in form of scarring
- Calcification in 20%
- Gray, yellow, or tan
- Sharply circumscribed; satellite lesions
- Hemorrhage or necrosis may be evident
- Non encapsulated
- No multi-centricity

Microscopic Pathology
- Metanephric Adenoma–like Tumors of the Kidney
- Report of 3 Malignancies With emphasis on Discriminating Features
- Jones EJ et al, Davis CJ et al, Pins MJ et al.
Microscopic Findings

- Nucleoli inconspicuous
- No infiltrative growth
- No vascular invasion
- Minimal cytoplasm
- Mitotic figures – rare or absent

Metanephric Adenoma

Needle Bx

- Tightly packed small tubules with little intervening stroma

Metanephric Adenoma

- N parenchyma
- Tumor is sharply demarcated from surrounding N renal parenchyma

Metanephric Adenoma

- Small uniform round acinar tubular structures separated by scant stroma
- Lining epithelial cells have scant cytoplasm & hyperchromatic nuclei

Genetics

- No chromosome trisomy 7 & 17
- Unrelated to papillary RCC & Wilms’

Metastatic Potential

- 1 case report of a metastatic metanephric adenoma in a 7 yo F
  - Mets to paraAO, hilar, AO bifurcation LN’s

Pesti T, Jones DC. Human Pathology 2001;32(1):101-4

Synchronous Malignancy

- Case report of 8 yo F with metanephric adenoma and foci of pRCC and metastasis


Metanephric Adenoma

- Tight packaging of small acini
- Near solid appearance
- No cytologic atypia
- Mitotic figures rare

Metanephric Adenoma

- 50% of cases have papillary structures
- Minute cysts into wc blunt papillary structures protrude
- Resemble immature glomeruli
- Typ no bld vessels in these papillary structures

Metanephric Adenoma

- Psammoma bodies are commonly seen in MA
- 1st defined by EC Jones

Summary

- Almost always benign clinical course with paraneoplastic syndrome
- Primarily a pathological Dx
  - lack of clinical, radiographic, or cytologic means of making definitive Dx