Case of the Month – March 2014

• 3 year old female with intermittent abdominal pain, nausea, vomiting, and palpable abdominal mass.

• Investigations reveal:
  – Elevated AST, ALT, LD, CRP, HCG, AFP >150 000
  – Afebrile

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Contrast Enhanced CT of the Abdomen/Pelvis

• What are the imaging findings?
• What is the differential diagnosis? Diagnosis?
Lungs
Findings

- Well-defined heterogeneous masses throughout both lobes of the liver on a background of hepatomegaly.
- Mass effect on adjacent liver parenchyma with compression and displacement of vessels.
- Left lower lobe soft tissue lesion.
Differential Diagnosis

• Liver abcess?
• Hepatoblastoma?
• Hepatocellular Carcinoma?
• Mesenchymal Hamartoma?
• Other?
Diagnosis

• Findings most in keeping with hepatoblastoma based on radiologic appearance, clinical presentation, and biochemical markers.

• Abscess unlikely given patient is afebrile and multi-focal nature of lesions (lung and liver).

• No known predisposing disorders for hepatocellular carcinoma.

• Mesenchymal hamartoma more common in children < 2 years with septated cystic/solid mass
Discussion

- Malignant embryonic hepatic tumor composed of epithelial cells or mixed epithelial/mesenchymal cells.
- Most common primary liver tumor in children (90% of tumors <5 years), elevated AFP common
- 95% occur before age of 5; M:F = 2:1
- Pathophysiology: etiology is unknown but extreme prematurity (< 1000g) increases risk by >15x. Maternal exposure to paints, metals, and smoking increase risk.
- Associated with other syndromes and genetic abnormalities like FAP and Beckwith-Weideman.
Discussion

- Characterized by heterogeneous enhancing well-defined mass/masses as presentation.
- Calcifications in 50% and more common in right lobe of liver. Average size ~10 cm.
- Metastasis is present in ~25% at diagnosis with the majority found within the lungs.
- Staged using PRETEXT system which evaluates the number of liver segments free from tumor.
- Treatment: Surgical resection +/- neoadjuvant chemotherapy which has high success rate (90% 3 year survival for standard risk, 50% for high risk). Transplantation is an option for unresectable disease.
References

• StatDx – Hepatoblastoma
