Abdominal Mass in an Infant

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Case Presentation

A previously healthy 8-month-old girl was brought to the pediatrician with increasing abdominal girth. On physical exam, the patient was tachypneic, and her abdomen was distended. Constipation was suspected and the patient was referred for a conventional radiograph of the abdomen, which demonstrated a large abdominal mass (Fig. 1A). An abdominal ultrasound (not shown) and contrast-enhanced CT of the abdomen and pelvis (Fig. 1B and C) was performed to further characterize the mass. Laboratory evaluation revealed an elevated serum alpha-fetoprotein of 394 ng/ml (reference range 0-99 ng/ml). Serum CEA and b-HCG were normal. The serum LDH was low at 185 U/L (normal range 208-274 U/L). The patient underwent laparotomy with subsequent definitive surgical management at a tertiary pediatric treatment facility.

Figure. AP supine view of the abdomen (A) demonstrates diffuse soft tissue density filling the right flank and midline of the abdomen. Air-filled bowel loops are displaced into the left upper quadrant. Axial (B) coronal (C) contrast-enhanced CT images show a solid and cystic mass distorting the inferior margin of the right hepatic lobe.
Key Imaging Finding

Large solid and cystic right upper quadrant mass in an infant

Differential Diagnoses

Renal origin – 55%
- Hydronephrosis
- Polycystic kidney disease
- Mesoblastic nephroma
- Nephroblastomatosis – Wilms tumor spectrum
- Renal vein thrombosis
- Ectopic kidney

Gastrointestinal origin – 15%
- Duplication cyst
- Mesenteric cyst
- Omental cyst
- Meconium pseudocyst

Pelvic origin extending into abdomen – 15%
- Ovarian cyst
- Hematocolpos
- Sacrococcygeal teratoma

Adrenal origin -10%
- Adrenal hemorrhage
- Neuroblastoma
- Teratoma

Hepatobiliary origin – 5%
- Benign
  - Hepatic Cyst
  - Choledochal cyst
  - Infantile hemangioendothelioma
  - Mesenchymal hamartoma
  - Infantile hemangioma
- Malignant
  - Hepatoblastoma
  - Undifferentiated embryonal sarcoma
  - Undifferentiated rhabdomyosarcoma of the biliary tree

Discussion

The differential diagnosis of infantile abdominal masses is broad and based on age, location, and structure of the lesion. In infants, there are 3 primary locations for masses: renal (55%), gastrointestinal (GI) tract (15%), and extension of a pelvic mass (15%). Ultrasound is often the first imaging modality used in the evaluation of an abdominal mass since it is widely available, does not expose the infant to radiation, and can usually be performed without sedation. The origin of the tumor can frequently be determined by ultrasound. The lesion can be classified as cystic, solid, or mixed. Vascularity of the lesion and evidence of vascular invasion should be evaluated prior to biopsy or resection.

Renal masses comprise the majority (55%) of abdominal masses in infants. Hydronephrosis is most common with other etiologies including polycystic kidney disease, mesoblastic nephroma, nephroblastomatosis – Wilms tumor spectrum, renal vein thrombosis, and ectopic kidney. GI tract masses account for about 15% of abnormalities. Common causes include duplication cysts and mesenteric or omental cysts, as well as meconium pseudocysts. Pelvic masses extending into the abdomen make up another 15% and include ovarian cyst, hematocolpos, and sacrococcygeal teratoma. Non-renal flank masses make up 10% of cases and include adrenal hemorrhage (most common in neonates), neuroblastoma, and teratoma.

Hepatobiliary masses are least common, comprising only 5% of abdominal masses in infants. The most common benign hepatic tumors of infancy are infantile hemangioendothelioma, mesenchymal hamartoma, and infantile hemangioma. Hepatoblastoma is the most common primary malignant hepatic tumor in this age group. Other malignant liver tumors in infants include undifferentiated embryonal sarcoma and embryonal rhabdomyosarcoma of the biliary tree.

This mass was surgically proven to be a mesenchymal hamartoma, which is the second most common benign liver tumor in children. It was successfully resected without complication or need for additional therapy. This entity usually presents as a multicystic mass in the liver in a child younger than 2 years of age. The stroma is usually cystic or mixed but can contain angiomatous components and be
multifocal. There are reports that patients may have increased occurrence of a balanced translocation at the long arms of chromosomes 11 and 19.

CT images are notable for large feeding vessels. With contrast administration, there is enhancement of the septae and solid (stromal) elements. Pathologically, mesenchymal hamartoma is composed of myxomatous mesenchyme and malformed bile ducts which may bulge from the liver or may even appear pedunculated. Rapid expansion can occur with accumulation of fluid in the cystic spaces. Complications include ascites, jaundice, and rarely congestive heart failure.

If the tumor is detected prenatally, there is increased risk of preterm delivery. In utero percutaneous decompression of the larger cysts improves prognosis. This tumor is often very large at initial detection, making surgical excision complicated. Although rare, cases of malignant transformation into undifferentiated embryonal sarcoma are reported. Although the tumor grows rapidly in young children, over time it may shrink. It may be followed with ultrasound in asymptomatic patients.

Diagnosis

Hepatic mesenchymal hamartoma

Summary

When an abdominal mass is discovered in an infant, conventional radiography is often performed first followed by ultrasonography. The most common etiologies of an abdominal mass in an infant include renal, GI, and pelvic masses, followed by adrenal and hepatic lesions. By determining the organ of origin, internal characteristics, and vascularity of the lesion, the extensive differential diagnosis can be narrowed. Additional imaging with CT or MRI can be of benefit if surgical excision is contemplated.

References