A Pediatric Patient With Abdominal Pain

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Radiology Quiz

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A previously healthy 2-year-old girl presented to the pediatrician's office with chief complaint of abdominal discomfort.

History. The child's mother denied any history of fever, cough, nausea, vomiting, diarrhea, constipation, or weight loss. Her physical examination was unremarkable. There was no lymphadenopathy, bruises, or rash, and normal auscultation of heart and lungs could be heard. The patient's initial abdominal examination was performed by the general/family practitioner at an outpatient clinic. Her abdominal examination did not indicate organomegaly or tenderness. The patient's neurological and musculoskeletal examination was unremarkable as well.

Diagnostic Testing. Following her examinations, a radiologist performed antero-posterior (Figure 1) and lateral view (Figure 2) abdominal x-rays.

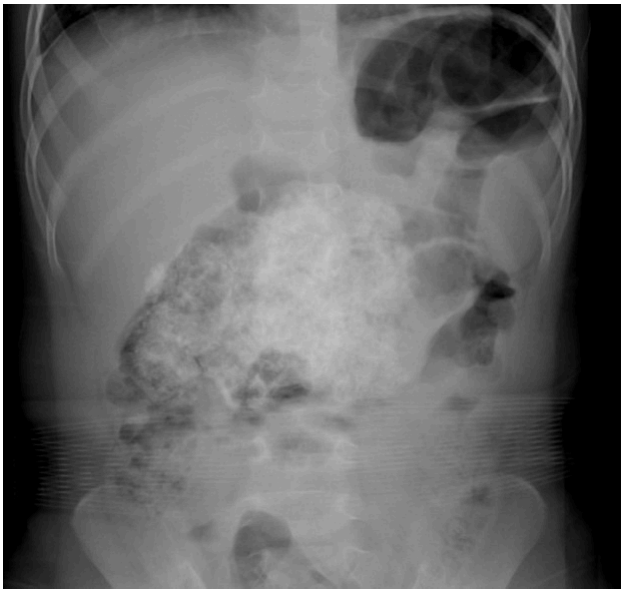

Figure 1. *Abdominal x-ray antero-posterior view.*

Answer: D. Large, calcified mass in the midline upper abdomen

Figure 1. *(Annotated) Abdominal x-ray antero-posterior view.*

Figure 2. *(Annotated) Abdominal x-ray lateral view*

Abdominal x-rays reveal a gently lobulated calcification in the midline upper abdomen, outside the lumen of the stomach and colon, which is consistent with a large calcified intra-abdominal mass. This finding was missed by the general radiologist, and the patient returned to the pediatrician's office about 6 weeks later with increased abdominal fullness, discomfort, reduced appetite, and anemia. An ultrasound examination was performed on the same day of the visit to the pediatrician's office, which revealed normocytic anemia and elevated liver enzymes.

Based on the x-rays, the differential diagnoses include a hepatic mass or a peritoneal mass. A renal or adrenal mass is unlikely because the mass is not retroperitoneal or off midline. The findings on this x-ray are not related to contrast because there is no history of any prior abdominal imaging. Fecal loading of the colon is generally not hyperdense as the x-rays indicate. Additionally, fecal loading is not confined to the midline upper abdomen.

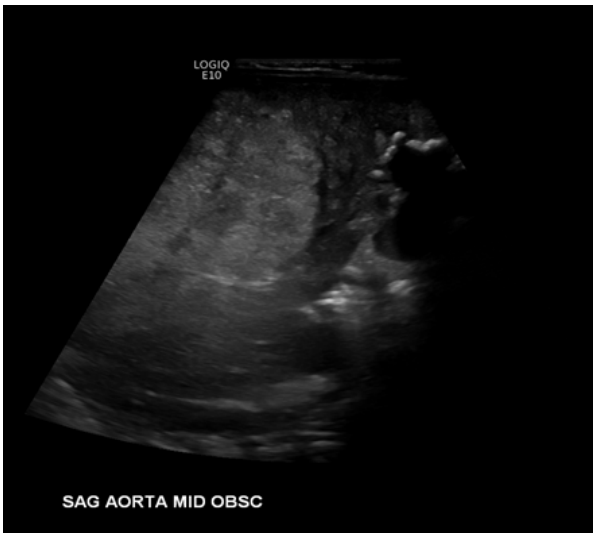

Figure 3. *Abdominal USG depicting the calcified abdominal mass.*

An abdominal ultrasonography (Figure 3) confirmed the calcified hepatic mass. Ultrasonography grayscale images show an infiltrative multifocal mass involving the majority of the liver with coarse chunky calcification in the mass in the left lobe of liver.

Figure 4. *Abdominal CT depicting the calcified abdominal mass in the left lobe of liver and non-calcified infiltrative multifocal masses in both lobes of liver.*

Treatment and Management. The patient was referred to a tertiary care center with the pediatric oncology service on the same day of the ultrasound examination. An abdominal CT scan confirmed the finding seen on the ultrasonography. There was no extension into the hepatic veins, portal vein, or inferior vena cava (IVC). There was retroperitoneal lymphadenopathy (not shown). An ultrasonography-guided biopsy confirmed a hepatoblastoma pre-treatment extent of tumor (PRETEXT) stage IV. The patient underwent 10 cycles of chemotherapy “Cisplatin and Doxorubicin” as per SIOPEL IV HR therapy followed by a living related donor partial hepatic transplant provided by the patient's mother. Patient is on sirolimus for immunosuppression.

Outcome and follow-up. The patient is now older than 18 months following completion of therapy. She has had few visits to the pediatric outpatient clinic for concerns of fever and cough in last 18 months without any significant concern for any other major illnesses. She continues to follow-up with her pediatric oncologist and pediatric gastroenterologist regularly.

Discussion. Hepatoblastoma is the most common primary malignant hepatic tumor in patients younger than 4 years of age. Most cases present in the first 18 months of life.[1] Patients usually present with painless abdominal mass.[1] Hepatoblastoma is a tumor of embryonic origin and can be seen occasionally on antenatal ultrasonography.[1] Most hepatoblastomas are sporadic, but may have association with Beckwith-Wiedemann syndrome, hemihypertrophy, familial adenomatous polyposis, prematurity with low birth weight, Gardner syndrome, glycogen storage disease, biliary atresia, and fetal alcohol syndrome.[1]

The usual presentation of hepatoblastoma is painless mass, but may present with nausea, vomiting, anorexia, pyrexia, anemia, abdominal pain, back pain, and jaundice. It may rarely present with peripheral precocious puberty due to beta human chorionic gonadotropin secretion. Serum alpha fetoprotein levels are elevated in up to 90% of cases.[1] Differentials include other hepatic masses like mesenchymal hamartoma, infantile hemangioma, neuroblastoma metastasis, rhabdomyosarcoma, and hepatocellular carcinoma.

PRETEXT staging is based on the number of involved hepatic segments, extrahepatic abdominal disease, involvement of portal vein, IVC/hepatic veins, and distant metastasis.

Abdominal x-rays may demonstrate a right upper quadrant mass and calcifications may be visible in 20-30% of cases on x-rays.[5] Ultrasonography demonstrates a hepatic mass with heterogeneous echotexture and dense calcifications in up to 50% of cases.[5] Heterogeneous hypodense mass is seen on computed tomography (CT), which is also helpful for staging.[3]

Most tumors are solitary but up to 15% of tumors can be multifocal like in our index patient.[4] Lung metastasis, lymph nodes, and extension into hepatic veins, IVC, or portal vein can be assessed on CT. Lungs are a common site of distant metastases.[1] CT may also demonstrate necrosis, calcification, and hemorrhage.[3] Liver MRI is helpful in delineating the margins and confirming other findings.[4]

Surgical resection is the treatment of choice, though presurgical chemotherapy may be given to reduce the tumor bulk. Liver transplantation is an approach that can potentially provide a cure in resectable tumors,[3] but IVC involvement indicates an unresectable disease.[1] Post-resection chemotherapy may be considered depending on the bulk of metastatic disease.[1] Long-term survival is about 60-70% depending on the staging for all ages.[1]

Conclusion. A previously healthy 2-year-old girl presented with abdominal discomfort. Her abdominal examination was unremarkable. Abdominal x-rays revealed a calcified mass in the midline upper abdomen. Follow-up ultrasonography revealed multifocal heterogenous hypoechoic masses in both lobes of liver with coarse chunky calcification in the left lobe of liver. CT of the abdomen confirmed the findings seen on the abdominal x-ray and ultrasonography. Additional retroperitoneal lymphadenopathy was also seen on CT scan. There was no extension into the hepatic or portal veins or IVC. Labs revealed normocytic anaemia and elevated liver enzymes. Based on the findings and liver biopsy, patient was diagnosed with hepatoblastoma PRETEXT stage IV. Patient received chemotherapy followed by living related donor partial hepatic transplant from her mother. Patient is on immunosuppressants, however otherwise doing well, meeting her developmental milestones and following her growth charts at 75th percentile for her weight and 50th percentile for her height.

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