A 5-MONTH-OLD male infant was admitted to the hospital with a 3-day history of fever associated with anorexia, fussiness, and decreased sleep. Information from a review of systems and history of present illness revealed a several-week history of constipation and gastroesophageal reflux. On physical examination the infant appeared healthy and had a nontender, firm, abdominal mass palpated in the right upper quadrant. The mass extended 9 cm below the costal margin. By auscultation, bowel sounds were present, and a diffuse low-pitched bruit was heard over the mass. Findings from the remainder of the physical examination were unremarkable.

Results of serum chemistry, liver function tests, prothrombin time, and partial thromboplastin time were normal. The complete blood cell count was remarkable for a hemoglobin level of 91 g/L and a platelet count of $203 \times 10^9$/L. Urine catecholamine levels were normal. His $\alpha_1$-fetoprotein level was greater than 60 µg/L (reference range, 0-15 µg/L). An abdominal radiograph showed an upper abdominal mass crossing the midline not seen on radiography performed 2 months earlier during evaluation of the patient’s gastroesophageal reflux. An abdominal ultrasonogram (Figure 1) and a computed tomographic scan (Figures 2, 3, and 4) of the abdomen were performed.

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Denouement and Discussion

Hemangioendothelioma of the Liver

Figure 1. Ultrasoundographic scan of the liver demonstrates a markedly enlarged, inhomogeneous liver with multiple hypoechoic nodules of varying sizes scattered throughout the parenchyma.

Figure 2. Computed tomographic scan of the liver shows an enlarged liver with multiple smoothly margined, round, low-attenuation masses of variable size distributed through all segments of the liver.

Figure 3. Immediate post–contrast injection images demonstrate enhancement through multiple low-attenuation regions, some enhancing uniformly and others enhancing peripherally.

Figure 4. Delayed post–contrast injection images at 3 minutes after injection show contrast filling the central portions of the previously peripherally enhancing nodules.

Hemangioendothelioma of the liver is the most frequent vascular tumor diagnosed during the newborn period. It is a benign, solitary or multicentric highly vascular tumor that arises from supporting tissues of the liver and skin.1 A hemangioendothelioma may develop areas of fibrosis, calcification, hemorrhage, or cystic degeneration, and it typically enlarges during the first 1 to 6 months of life. The natural history involves regression and complete or near-complete involution by age 12 to 18 months.1,2

These vascular tumors are symptomatic 85% of the time.3 Symptoms usually are present by age 2 months.3 The most serious complication is high-output congestive heart failure, which requires vigorous therapy with digitalis, diuretics, and oxygen, with planned, definitive treatment of the hemangioendothelioma.1-3 The younger the patient at presentation, the more severe the cardiac failure.2 Other complications are consumptive coagulopathy and thrombocytopenia (Kasabach-Merritt syndrome), respiratory and feeding problems owing to compression of neighboring structures, hepatocellular carcinoma, and hemorrhage.1,2,4

The ultrasonographic appearance of a hemangioendothelioma is a highly echogenic, a complex, or a hypoechoic mass.1 Dynamic, contrast-enhanced computed tomography is diagnostic for hemangioendothelioma of the liver.1 Hemangioendotheliomas appear as multiple round or oval, well-circumscribed, homogeneous, hypodense masses that initially enhance at the periphery with progressive centripetal filling in of enhancement after intravenous injection.1 Washout of the contrast medium occurs within 5 minutes of administration.1 Radionuclide or magnetic resonance imaging can be helpful. Radionuclide scintigraphy with technetium Tc 99m–labeled erythrocytes shows increased activity during the blood-pool phase and on delayed imaging.1 On magnetic resonance imaging, hemangioendotheliomas are well-defined masses with lower signal intensity than normal liver tissue on T1-weighted images and higher signal intensity on T2-weighted images.1 After the administration of gadolinium diethylenetriamine pentaacetic acid, progressive centripetal enhancement is observed.1

Laboratory data may indicate consumptive coagulopathy,1,2,4 αfetoprotein levels are usually moderately increased.3 Hemangioendotheliomas are known to regress spontaneously; however, treatment of complications is often necessary. Medical therapy includes corticosteroids and interferon alfa-2A.1-3 Corticosteroids have shown a 30% to 60% response rate.2 Interferon alfa-2A inhibits capillary endothelial cell migration, endothelial cell proliferation in vitro, and angiogenesis in animals.3 Surgical therapy is used if the hemangioendothelioma is solitary or has minimal collateral vessels.1-3 The hemangioendothelioma may be excised by partial heptectomy or its afferent vessels can be embolized or ligated.1,3

Our patient was initially treated for 2 months with daily subcutaneous injections of 2 million U/m2 of IFN-α-2A, but he showed no notable response. He was then treated with high-dose systemic steroids for 1½ months with initial involution of the hemangioendothelioma. Seven months after diagnosis, he continues treatment with interferon alfa-2A. Almost 50% of pediatric liver tumors are benign.4 Differentiation of a benign hepatic mass from a malignant hepatic mass, such as hepatoblastoma, is essential.

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