A 51-Year-Old Woman With a Liver Mass

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A 51-year-old healthy woman received a thoracic and abdominal computed tomography scan during an evaluation for community-acquired pneumonia. The abdominal scan showed an incidental 10.0 × 6.5 × 3.0-cm cystic hepatic mass that appeared benign radiologically. Because laboratory studies were within normal limits at the time of the scan, the decision was made to re-evaluate the hepatic mass with additional imaging in 6 months. The patient was treated for her pneumonia and recovered without incident. Repeat computed tomography scan of the chest and abdomen 6 months later showed that the cystic liver mass had increased in size to 22.0 × 15.0 × 8.0 cm. Laboratory evaluation at this time revealed the following liver function test abnormalities (with reference ranges in parentheses): aspartate aminotransferase, 859 U/L (5–55 U/L); alanine aminotransferase, 513 U/L (3–50 U/L); total protein, 5.2 g/dL (6.0–8.2 g/dL); albumin, 2.1 g/dL (3.5–5.0 g/dL); total bilirubin, 1.0 mg/dL (0.2–1.3 mg/dL); and alkaline phosphatase, 117 U/L (30–125 U/L). Carcinoembryonic antigen, α-fetoprotein, CA 125, and CA 19-9 serum levels were within normal limits. The patient underwent a right hepatic lobectomy.

The resected specimen weighed 2400 g and measured 25.0 × 19.0 × 8.5 cm. The surface showed a variegated tumor composed of 2 attached and circumscribed locules. Together, the 2 locules of tumor comprised a mass measuring 23.0 × 16.0 × 7.0 cm (Figure 1). The tumor contained areas of hemorrhage, glistening myxoid areas, necrosis with cystlike formations, and solid, soft tan areas. Histologically, the mass was circumscribed but not encapsulated. The sheets of tumor cells were stellate and spindle-shaped with strikingly pleomorphic, hyperchromatic nuclei and plentiful homogeneous eosinophilic cytoplasm (Figure 2, A). Other areas showed smaller, more basophilic and spindle-shaped nuclei with less eosinophilic cytoplasm and a looser, less cohesive pattern (Figure 2, B). Bizarre cells, multinucleated giant cells, and mitotic figures, including atypical mitoses, were present. The myxoid background varied in density. Spherical globules of eosinophilic material were present both within the cytoplasm of some neoplastic cells as well as in the intercellular matrix.

What is your diagnosis?

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Pathologic Diagnosis: Undifferentiated Embryonal Sarcoma of the Liver

Abstract

We describe an incidentally diagnosed large hepatic tumor in a previously healthy 51-year-old woman. The tumor showed gross, microscopic, immunohistochemical, and ultrastructural features of undifferentiated embryonal sarcoma, which is a primitive mesenchymal tumor usually encountered in the pediatric population. Very few cases of undifferentiated embryonal sarcoma have been reported in patients older than 15 years. This neoplasm in adults appears to exhibit some differences from those neoplasms found to be in the pediatric group. Prognosis is poor for both children and adults with this tumor.

Evaluation of the tumor shows classic gross, microscopic, immunohistochemical, and ultrastructural features of undifferentiated embryonal sarcoma. The name of the tumor underscores its defining light microscopic characteristic: no specific cell lineage differentiation.1,2 Although the tumor is considered to be of primitive mesenchymal origin, few studies have evaluated tumor cells ultrastructurally.3 Studies that include ultrastructural analysis, however, reveal a variable list of primitive mesenchymal cell types, and the exact histogenesis of the neoplasm remains in question.3 The predominance of cases of embryonal sarcomas present in pediatric patients, and, as a result, most reports that evaluate occurrences in adults suffer from small numbers. As in this case, most embryonal sarcomas are located in the right lobe of the liver.2 The cut surface of these well-demarcated and occasionally encapsulated tumors is variegated, with gray, tan, and yellow cystic and gelatinous tissue, with or without areas of hemorrhage or necrosis.2

Most of the neoplastic cells are stellate or spindle-shaped. The architecture varies from a dense, cellular picture to a loose assembly of cells with a myxoid-appearing background. Pleomorphism, bizarre cells, multinucleated giant cells, and mitoses are seen.2 The tumor cells in this case show strong positivity for vimentin (Figure 3, A), desmin (Figure 3, B), α1-antitrypsin (Figure 3, D), and CD68. Tumor cells are weakly and focally positive for muscle-specific actin and keratin 8/18, and are negative for cytokeratin AE1/AE3, S100 protein, α-fetoprotein, and smooth muscle actin. Characteristic of this tumor are the spherical globules of eosinophilic material that are present both within the cytoplasm of the large tumor cells as well as within the intercellular matrix. These globules are positive for desmin (Figure 3, B), diastase-resistant periodic acid–Schiff (Figure 3, C), α1-antitrypsin (Figure 3, D), and muscle-specific actin. These globules are shown to be lysosomal granules when evaluated ultrastructurally, and may be caused by tumor cell apoptosis.2

Not only do the tumor cells in this case contain the characteristic densely packed lysosomal granules (Figure 4, small arrow), but occasional bundles of myofilaments are also seen (Figure 4, large arrow).4 Some reports suggest that the presence of myofilament bundles seen by electron microscopy indicate differentiation toward smooth muscle, and that the tendency to develop this feature is a key distinguishing factor between pediatric and adult cases of embryonal sarcoma.4 Several other features of adult cases of embryonal sarcoma (patients presenting at an age older than 15) have slight variations from the usually encountered pediatric cases. Pediatric cases of embryonal sarcoma tend to present with an equal sex distribution, but there is a female preponderance in adults. Adult cases tend to have a less prominent myxoid component seen histologically than most pediatric cases.4 Few cases of embryonal sarcoma have been reported in adult patients, however, and additional review of adult cases is warranted to carefully consider the differences between adult and pediatric patients and the treatment and prognosis implications these differences may carry.4,5 Although embryonal sarcoma is generally a disease of childhood, it does occur in the adult population, and it is crucial to keep this entity in the differential diagnosis when evaluating a liver mass, regardless of patient age.

References

