10 Rare Primary Malignancies of the Liver


CONTENTS

10.1 Introduction 153
10.2 Hepatoblastoma 154
10.2.1 Incidence and Clinical Presentation 154
10.2.2 Pathologic Findings 154
10.2.3 Imaging Findings 154
10.2.3.1 Ultrasound 154
10.2.3.2 Computed Tomography 154
10.2.3.3 MR Imaging 155
10.2.3.4 Angiography 155
10.3 Biliary Cystoadenoma and Cystoadenocarcinoma 155
10.3.1 Incidence and Clinical Presentation 155
10.3.2 Pathologic Findings 155
10.3.3 Imaging Findings 156
10.3.3.1 Ultrasound 156
10.3.3.2 Computed Tomography 156
10.3.3.3 MR Imaging 156
10.3.3.4 Angiography 156
10.4 Epithelioid Hemangioendothelioma 156
10.4.1 Incidence and Clinical Presentation 156
10.4.2 Pathologic Findings 156
10.4.3 Imaging Findings 156
10.4.3.1 Ultrasound 158
10.4.3.2 Computed Tomography 158
10.4.3.3 MR Imaging 158
10.5 Angiosarcoma 158
10.5.1 Incidence and Clinical Presentation 158
10.5.2 Pathologic Findings 159
10.5.3 Imaging Findings 159
10.5.3.1 Ultrasound 159
10.5.3.2 Computed Tomography 160
10.5.3.3 MR Imaging 160
10.5.3.4 Angiography 160
10.6 Other Sarcomas 161
10.6.1 Incidence and Clinical Presentation 161
10.6.2 Pathologic Findings 161
10.6.3 Imaging Findings 162
10.6.3.1 Ultrasound 162
10.6.3.2 Computed Tomography 162
10.6.3.3 MR Imaging 162
10.7 Lymphoma 162
10.7.1 Incidence and Clinical Presentation 162
10.7.2 Pathologic Findings 164
10.7.3 Imaging Findings 164
10.7.3.1 Ultrasound 164
10.7.3.2 Computed Tomography 164
10.7.3.3 MR Imaging 164

References 165

10.1 Introduction

Primary malignant neoplasms of the liver are classified by the cell of origin (Table 10.1). In this chapter, primary malignant liver tumors are discussed: hepatoblastoma, arising from hepatocytes; cystoadenoma and cystoadenocarcinoma, arising from biliary cells; epithelioid hemangioendothelioma, angiosarcoma and other mesenchymal sarcomas, arising from mesenchymal tissue; and finally primary lymphoma, arising from lymphomatous tissue.

Table 10.1. Malignant liver tumors

<table>
<thead>
<tr>
<th>Hepatocellular origin</th>
<th>Cholangiocellular origin</th>
<th>Mesenchymal origin</th>
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</thead>
<tbody>
<tr>
<td>Hepatocellular carcinoma</td>
<td>Cholangiocarcinoma</td>
<td>Angiosarcoma</td>
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<td>Fibrolamellar carcinoma</td>
<td>Cholangiocarcinoma</td>
<td>Epithelioid hemangioendothelioma</td>
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<td>Hepatoblastoma</td>
<td>Cystoadenocarcinoma</td>
<td>Leiomyosarcoma</td>
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<tr>
<td>Mesenchymal origin</td>
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<td>Fibrosarcoma</td>
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<td>Malignant fibrous histiocytoma</td>
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<td>Malignant fibrous histiocytoma</td>
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<tr>
<td>Primary lymphoma</td>
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<td>Primary lymphoma</td>
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10.2 Hepatoblastoma

10.2.1 Incidence and Clinical Presentation

Hepatoblastoma is the most frequent primary hepatic neoplasm in the pediatric age group (NELSON et al. 1996), constituting 43% of all tumors (DAVEY and COHEN 1996). The tumor is often detected before 3 years of age (BOECHAT et al. 1988; GÉOFFRAY et al. 1987), with a median survival of 1 year (FRIEDBURG et al. 1989). Hepatoblastoma is more frequent in males than in females, with a ratio of 3:2 (DAVEY and COHEN 1996). It is not associated with underlying cirrhosis but is more common in patients with hemihypertrophy, Beckwith-Weideman syndrome, Wilm's tumor, glycogen storage disease, diaphragmatic and umbilical hernias, and Meckel's diverticulum (DAVEY and COHEN 1996; NELSON et al. 1996; ROSAI 1996). Rare occurrence in siblings has been observed (HOROWITZ et al. 1987).

Presentation is most often due to enlargement of an abdominal mass with a few cases manifesting fever, pain, weight loss, and vomiting (NELSON et al. 1996); jaundice is rarely observed. The serum alpha-fetoprotein is elevated in 66% (HAAGA et al. 1994) to 90% (DAVEY and COHEN 1996) of cases.

10.2.2 Pathologic Findings

Hepatoblastoma is a malignant tumor of hepatocyte origin, which often contains mesenchymal components (ISHAK and GLUNZ 1967). On macroscopic inspection, hepatoblastoma is a solid, well-defined, sometimes lobulated mass, surrounded by a pseudocapsule (BOECHAT et al. 1988; ROSAI 1996). Although it is usually solitary, multiple lesions can be observed in less than 20% of cases (DAVEY and COHEN 1996; ROSAI 1996). Areas of necrosis and calcification are frequently present (BOECHAT et al. 1988).

Microscopically, it can be classified as an epithelial or mixed (epithelial-mesenchymal) neoplasm. Epithelial hepatoblastoma is composed of fetal or embryonal malignant hepatocytes or a combination of these (BOECHAT et al. 1988; MARTI-BONMATI et al. 1993). These cells are associated with extramedullary hematopoiesis pseudocapsule (BOECHAT et al. 1988; ROSAI 1996). Mixed hepatoblastoma has both an epithelial (hepatocyte) component and a mesenchymal component, consisting of primitive mesenchymal tissue and osteoid material and/or cartilage, which is responsible for the calcification seen on imaging studies.

The histologic classification has prognostic implications: the epithelial type, particularly if it has fetal hepatocytes, has a better prognosis. Embryonal epithelial cells are more primitive than fetal epithelial and mesenchymal cells, and tumors with this histologic type have a worse prognosis.

10.2.3 Imaging Findings

10.2.3.1 Ultrasound

Sonographically, hepatoblastoma appears as an echogenic mass that may have shadowing and echogenic foci corresponding to intratumoral calcification. Hyperechoic and/or cystic areas, corresponding to hemorrhage within the tumor, and/or necrotic areas may be present as well (KAUDE et al. 1981). Hepatoblastoma is associated with high Doppler frequency shifts that correlate with the neovascularity typical of this tumor (BATES et al. 1990).

10.2.3.2 Computed Tomography

On unenhanced scans, hepatoblastoma appears as a solid hypodense mass, which may occupy large portions of the liver. Fifty percent of hepatoblastomas show calcifications, which are particularly extensive.