

Embryonal rhabdomyosarcoma of the common bile duct and porta hepatis in an infant: A rare presentation

Tumors of any type causing obstructive jaundice which arise in the biliary tree in early childhood are rare. Embryonal Rhabdomyosarcoma (RMS) of the biliary tree isn't frequent tumor, that commonly arises from the common bile duct. Herein we describe the patient with embryonal RMS originating in the biliary tree.

16 month old male child was brought to the Department of Pediatrics with complaints of abdominal pain and jaundice for post four months. On physical examination, the child had slight hepatomegaly and icterus. Ultrasonography and computed tomography imaging showed a multicystic tumor in the porta hepatis, with dilated intra-and extrahepatic biliary ducts. The decision was to perform a left hepatectomy and resection of extrahepatic bile ducts. The macroscopic examination of the specimen revealed a soft, pale tan mass measuring 70x50 mm in the porta hepatis and neighbouring structures. Histological examination showed typically botryoid variant of embryonal rhabdomyosarcoma, with "cambium layer" of tightly packed tumor cells beneath the epithelium (Figure 1 a, b). 25 atypical mitotic figures per HPF were registered. Tumor cells showed positive for desmin and myogenin (Figure 1 c, d). By these results, the case was diagnosed as embryonal RMS. Postoperatively, he followed up for almost two-year and received seven courses of combined chemotherapy (endoxan, farmorubicin and vincristine) and radiotherapy. However, a new mass recurred.

Rhabdomyosarcoma is a malignant tumor arising from cells committed to skeletal muscle differentiation. Though this tumor is commonly seen in children and infants, the common sites are head and neck, genitourinary tract and retroperitoneum (1). The biliary system accounts for 1% of all RMS occurring in childhood. Symptoms of the onset of the embryonal RMS are anorexia, abdominal pain, fever, and features of

obstructive jaundice (2). The differential diagnosis of a mass in biliary tree in a child includes hepatoblastoma, choledochal cyst and mesenchymal hamartoma (3). The prognosis of patients with rhabdomyosarcomas depends on the primary site, the histological subtype, tumour size and staging, the latter according to the Intergroup Rhabdomyosarcoma Study Group criteria, which is based on disease extent, local and regional resectability (4). Some authors suggest aggressive surgery is unwarranted for embryonal RMS and these conclusions were drawn based on tumours of the proximal biliary tree where complete clearance is not feasible (5). Patients with embryonal RMS generally respond very well to chemotherapy. However, the biliary tract being an unfavorable site for RMS, a prolonged follow-up is necessary to evaluate the outcome of treatment.

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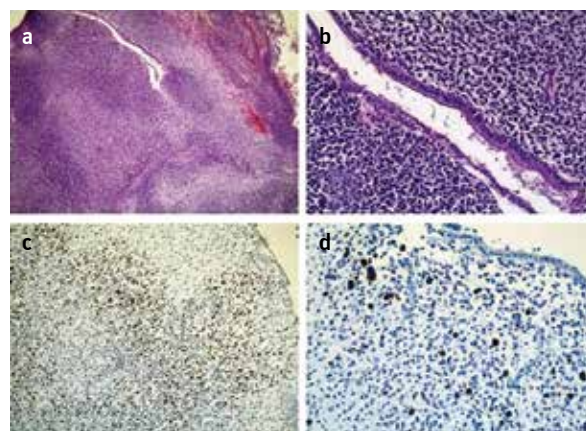


Figure 1. a-d. Primitive spindle-shaped tumor cells are roundish and have pleomorphic nuclei and large eosinophilic cytoplasm (H&E, x100) **(a)** Cambium Layer: The mucosa, with underlying hypercellular zone of poorly differentiated cells (H&E, x200) **(b)** The primitive tumor cells show diffusely nuclear positive staining for Myogenin (Myogenin x 200) **(c)** The tumor cells show positive focally staining for Desmin (Desmin x 200) **(d)**.

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