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IMAGES IN PEDIATRIC CARDIOLOGY

Fibrolamellar Hepatocellular Carcinoma With Cardiac Spread Causing Severe Inferior Vena Cava Obstruction in a 9-Year-Old Child

Jarrod D. Knudson · Jason F. Goldberg · Nancy A. Ayres

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Fibrolamellar carcinoma (FLC), a subset of hepatocellular carcinoma (HCC), is a very uncommon form of pediatric liver cancer, with fewer than 200 cases reported in the literature [3]. It is found primarily in older children and young adults, clustering between the ages of 10 and 35 years [4].

Fibrolamellar carcinoma is not associated with a history of cirrhosis or underlying liver disease, distinguishing it from HCC and hepatoblastoma (a much more common tumor in children) [4]. The clinical syndrome of FLC is characterized by malaise, abdominal pain, hepatomegaly/abdominal mass, and elevation of transaminases and alpha-fetoprotein [4]. The prognosis is variable because the tumor is quite chemotherapy resistant [3]. Lymph node and peritoneal metastases are common, and nearly half of FLC patients experience development of distant metastases to sites such as the mediastinum, abdominal organs, skeletal muscle, and pericardium [3]. However, to our knowledge, no cases of inferior vena cava (IVC) or right atrial spread in children have been reported to date. We report a case of FLC involving a 9-year-old child, with direct extension into the IVC and right atrium, resulting in severe IVC obstruction.

J. D. Knudson (✉) · N. A. Ayres
Division of Pediatric Cardiology, Department of Pediatrics, Baylor College of Medicine, Texas Children's Hospital, 6621 Fannin St., MC-19345-C, Houston, TX 77030, USA
e-mail: knudson@bcm.edu; jarrodknudson@gmail.com

J. F. Goldberg
Department of Pediatrics, Baylor College of Medicine, Texas Children's Hospital, Houston, TX 77030, USA

Case Report

A 9-year-old boy with no significant medical history was transferred from an outside hospital for evaluation of a hepatic tumor with cardiac spread. He had presented to his primary care physician with fatigue, and examination had shown a palpable abdominal mass. The boy underwent computed tomography of the abdomen and chest, which exhibited a liver mass that extended into the IVC and right atrium with bilateral pleural effusions. He was transferred to our institution for evaluation and treatment.

The boy's initial vital signs included a pulse of 138 beats/min, a respiratory rate of 39 breaths/min, and a blood pressure of 97/71 mmHg. He was intubated, and thoracostomy tubes were placed to evacuate pleural effusions. A hepatic biopsy performed at the outside institution was reviewed by our pathology division, and a tissue diagnosis of FLC was confirmed. Cardiology was consulted for sinus tachycardia, which was thought to be secondary to intravascular hypovolemia in the context of diuresis for pleural effusions and ascites.

An echocardiogram was performed, showing a large, severely obstructive IVC mass (Fig. 1). The mass extended through the IVC and occupied the majority of the right atrial cavity (Fig. 2). However, there was laminar blood flow around the mass and through the tricuspid valve (Fig. 2). Preoperative magnetic resonance imaging (MRI) was contemplated but not performed due to the risk associated with an additional general anesthetic.

Both general and cardiothoracic surgery took the patient to the operating room for attempted resection of the tumor. However, exploratory laparotomy showed direct extension of the tumor into the IVC. Given the extensive caval invasion, the IVC could not be dissected free, and the patient underwent cholycystectomy and Roux-en-Y

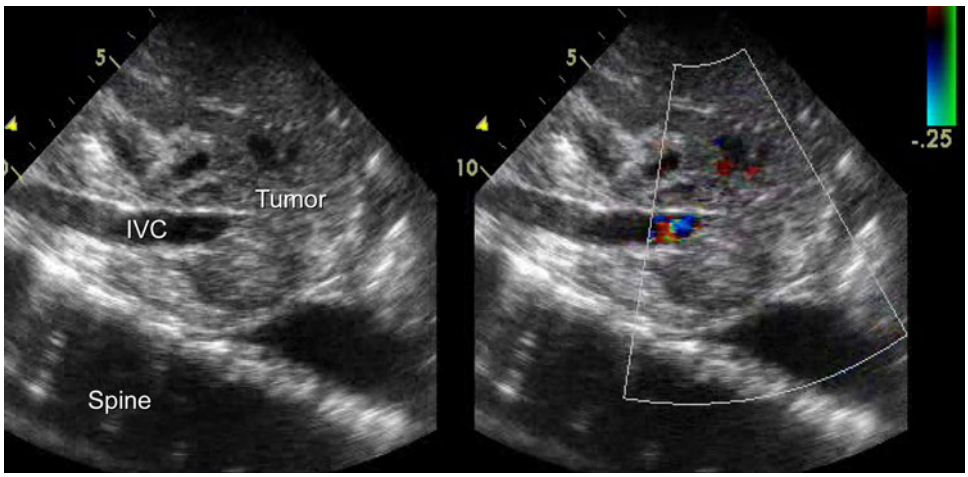

Fig. 1 Sagittal subxiphoid view showing the hepatic tumor penetrating and filling the inferior vena cava (IVC). Additionally, there is complete IVC obstruction, as evidenced by color-flow Doppler imaging (*right*)

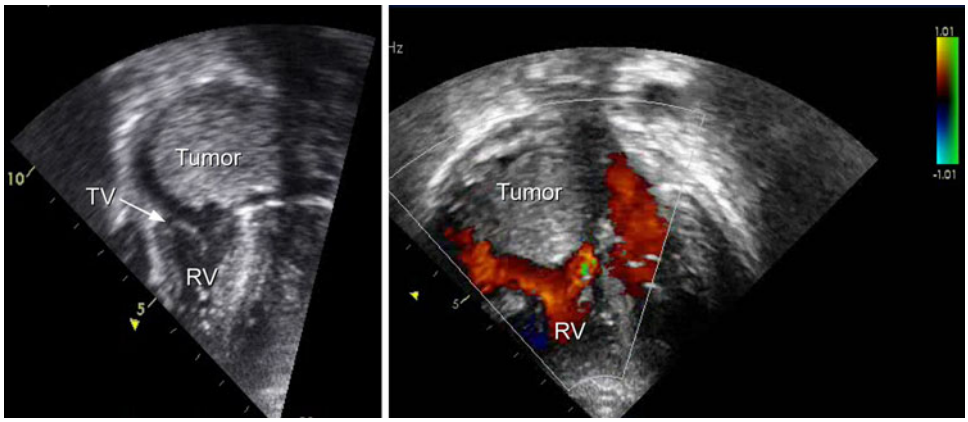

Fig. 2 Tumor occupying the majority of the right atrial cavity (*left*). Despite the extent of atrial involvement, there is laminar right ventricular inflow across the tricuspid valve, as evidenced by color-flow Doppler imaging (*right*). *TV* tricuspid valve, *RV* right ventricle

hepaticojejunostomy. No intracardiac exploration was performed. At this writing, the patient is receiving palliative chemotherapy and has undergone a Denver shunt for refractory ascites.

Discussion

Primary cardiac tumors are extremely rare in adults, who are 20 times more likely to have metastatic cardiac lesions. In a large autopsy series, one in five adults dying of cancer was found to have cardiac metastases [2]. In contrast, primary cardiac tumors comprise the majority of cardiac tumors in children, with cardiac metastatic disease being the exception. According to a recent international multicenter experience characterizing cardiac tumors in children by MRI, the majority of cardiac tumors in children are benign primary tumors (e.g., fibromas and rhabdomyomas), with malignancies representing approximately 15 % of the cases studied [1]. Although rare, cardiac extension/metastasis from distant primary tumor sites does occur in children and should be considered in the differential diagnosis when a child with a cardiac mass is evaluated.

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