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Case Report

Imaging of a glomus tumor of the liver in a child

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ABSTRACT

Glomus tumors occur preferentially in the subcutaneous tissue of the fingers and toes, but are extremely rare in visceral organs. Although, there have been several reports of glomus tumors in the liver in adult patients, there have yet been no publications reporting glomus tumors of the liver in children. Here, we report a case of an 11-year-old girl who was admitted with a 2-week history of progressive dyspnea on exertion and vomiting. Upon physical examination, she was found to have hypertension and a palpated smooth, firm mass at the epigastrium. Abdominal MRI revealed a well-defined exophytic hypervascular mass with intratumoral hemorrhage at segment 3/4b of the liver. Ultrasound-guided biopsy revealed it to be a glomus tumor. An ultrasound conducted at a 1-month follow-up after preoperative embolization revealed that the mass had decreased in size. A subsequent exploratory laparotomy with left hepatectomy was performed and the histologic results confirmed the diagnosis.

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Introduction

Glomus tumors are a type of mesenchymal hamartoma originating from the neuromyoarterial glomus body that surrounds the arteriovenous anastomoses. The glomus body is responsible for hemodynamic control including that of blood pressure, circulation, and body temperature. Although in the majority of glomus tumors are benign, in some cases, they are potentially malignant. Glomus tumors occur in the organs

that are rich in glomus bodies such as the digits, palms, and soles of the feet. Approximately 75% of glomus tumors occur in the hands, mostly in the subungual region of the fingertips [1]. Glomus tumors are extremely rare in the visceral organs but have been reported in the mediastinum [2], respiratory tract [3,4], gastrointestinal tract [5,6], and urogenital tract [7,8]. There have been several publications reporting glomus tumors in the hepatobiliary systems of adult patients [9–15], but to our knowledge, there have been no reported cases of glomus tumors of the liver in children.

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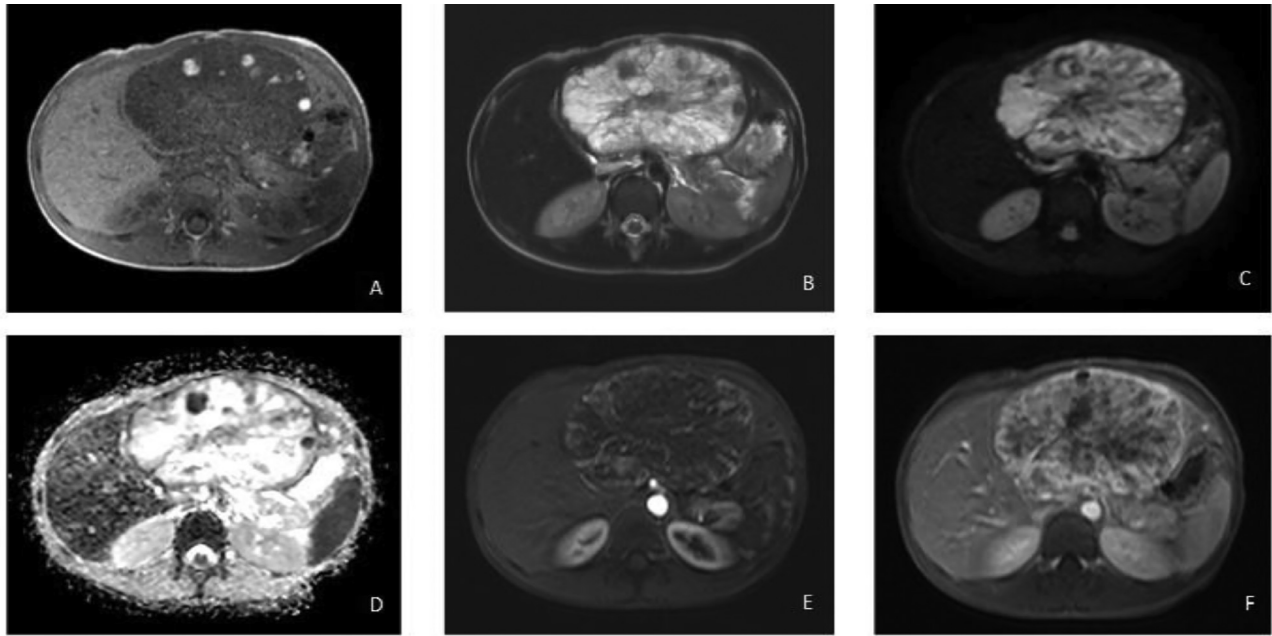


Fig. 1 – Abdominal MRI revealed a 12-cm well-defined exophytic mass at hepatic segment 3/4b that showed hypointense signal on T1W with foci of T1W hyperintensities representing intratumoral hemorrhage (A). It showed heterogeneously hyperintense signal on T2W (B), restricted diffusion (C, D), and peripheral enhancement on arterial phase (E), and progressive enhancement and septal enhancement on delayed images (F).

Case presentation

A previously healthy 11-year-old girl was admitted with a 2-week history of progressive dyspnea on exertion and vomiting. Her family history was unremarkable. Physical examination revealed hypertension and a smooth, firm mass at the epigastrium. A systolic apical murmur was noted upon heart examination. A liver function test showed only elevated cholesterol (396 mg/dl). Other laboratory tests (complete blood count, blood chemistry test, renal and liver function test, coagulation test, hepatitis profiles, and alpha-fetoprotein test) were within the normal limits. An echocardiogram found mitral and tricuspid regurgitation and left ventricular systolic dysfunction. An abdominal CT scan from previous hospital found a left lobe liver mass. Abdominal MRI (Fig. 1) revealed a 12-cm well-defined exophytic mass at hepatic segment 3/4b which showed hypointense signal on T1W with foci of T1W hyperintensities representing intratumoral hemorrhage. It showed heterogeneously hyperintense signal on T2W, restricted diffusion, and peripheral enhancement on arterial phase and progressive enhancement and septal enhancement on delayed images. The patient's liver parenchymal background was normal, and there were no other suspicious lesions. An ultrasound-guided biopsy was performed percutaneously under local anesthesia, and histopathological study revealed it to be a glomus tumor. The patient underwent preoperative embolization, and a celiac angiogram revealed a hypervascular tumor, which fed from A3 branch of left hepatic artery (Fig. 2A). Embolization was performed using a mixture of polyvinyl alcohol (PVA) 355-500 micron, and

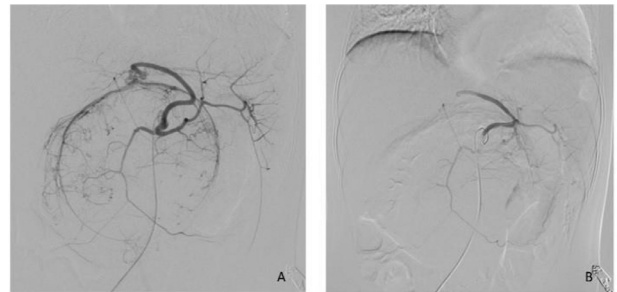


Fig. 2 – (A) The celiac angiogram revealed a large hypervascularized tumor at the inferior portion of the left hepatic lobe fed by A3 branch of left hepatic artery. (B) Left hepatic artery embolization using administered mixture of PVA 355-500 and contrast material.

contrast material (Fig. 2B). An ultrasound conducted at a 1-month follow-up revealed that the tumor had decreased in size to 8 cm (Fig. 3). An exploratory laparotomy with left hepatectomy was subsequently performed.

The pathology results showed dilated vascular channels surrounded by uniform neoplastic cells with round nuclei, fine chromatin, inconspicuous nucleoli, and pale eosinophilic cytoplasm, as well as a well-defined cytoplasmic border (Fig. 4A, 4B). No atypia or mitotic figures were present.

Immunohistochemical (IHC) staining of the tumor was positive for h-Caldesmon (Fig. 4C), smooth muscle actin (SMA) (Fig. 4D), and CD34 (Fig. 4E). Other IHC tests, including those for

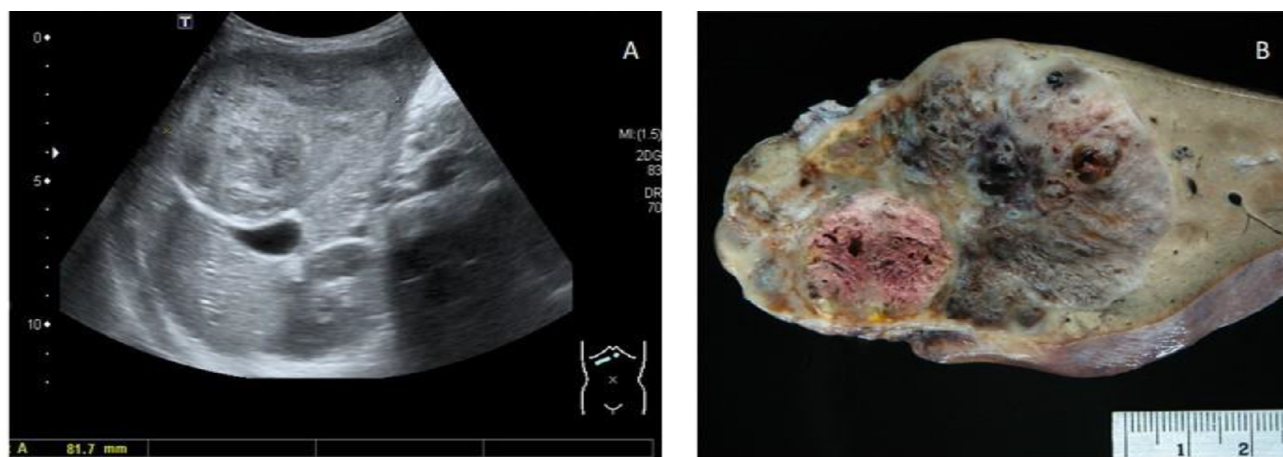


Fig. 3 – An ultrasound conducted at a 1-month follow-up after embolization and gross pathology. (A) The ultrasound revealed that the tumor had decreased in size to 8 cm. (B) A well circumscribed, inhomogeneously tan-brown mass with areas of necrosis and hemorrhage.

AE1/AE3, Heppar1, CD31, desmin, and myogenin, were negative. Based on these findings, the mass was diagnosed as glomus tumor and of uncertain malignant potential.

Discussion

Hepatic neoplasms account for 2% of all childhood tumors and about 6% of pediatric abdominal neoplasms [16]. Approximately two-thirds of liver tumors in children are malignant, particularly after 6 months of age [17]. Primary malignant tumors are the third most common type of intra-abdominal malignancy after Wilms tumors and neuroblastoma. The most common malignant hepatic tumors in children are hepatoblastomas, hepatocellular carcinomas (HCC), undifferentiated embryonal sarcomas, angiosarcomas, and embryonal rhabdomyosarcomas, respectively [16]. In addition to vascular tumors and mesenchymal hamartomas, the entities of benign liver tumors in adults can be found in children such as focal nodular hyperplasia (FNH), hepatic adenoma, and nodular regenerative hyperplasia [16,18].

Because of the patient's age, normal serum alpha fetoprotein (AFP), and MRI tumor characterization, we were not able to specifically diagnosis the tumor as any of the types of liver tumors that commonly develop in children. Angiosarcoma is a rare malignant tumor that occurs primarily in elderly patients, but has been known to occur in young girls who have previously been diagnosed with infantile hemangioendothelioma [19]. This condition can manifest in 1 of 4 patterns: Multiple nodules, a large dominant mass, a mixture of dominant mass and multiple nodules, and diffuse micronodular infiltration of the liver [20]. The MRI features of angiosarcoma are intratumoral hemorrhage, heterogeneous enhancement, and progressive enhancement on delayed phase without central filling, which were consistent with our findings here.

Although there was peripheral arterial enhancement, epithelioid hemangioendothelioma and hemangioma were ruled out due to the lack of progressive centripetal appearance. FNH was not considered due to incomplete contrast enhancement, no central hyperintense T2W, and delayed enhancement of the central scar. Hepatic adenoma was entirely excluded from the possible diagnoses because of the patient's age, incomplete contrast enhancement, and the mass not becoming isointense to the liver on portovenous and delayed phases. Hepatocellular carcinoma was also less likely as the patient's serum AFP and liver parenchymal background were normal and there was no washout of contrast material on portovenous and delayed phases.

As the liver mass was large and lacked specific imaging features, further ultrasound-guided biopsy was performed in order to allow for definite pathologic tissue diagnosis.

There have been several cases reported of glomus tumors of the liver in adults. Kenn et al reported a glomus tumor of the liver that appeared as a well-defined mass with moderately hyperintensity on T2W with an early spoke-wheel-like enhancement [9]. Amoueian et al reported on multiple fused cystic liver masses appearing on a CT scan [12]. Jaiswal et al reported a case of a hypervascular liver mass with a prominent feeding artery and draining portal vein [11]. A report by Hirose et al described a case of a solid-cystic liver mass filled with blood, the solid portion of which showed progressive enhancement on a CT scan [15].

Preoperative embolization has the advantage of being able to decrease vascularity in hypervascular tumors, which minimizes blood loss during the operative procedure.

At a 1-month follow-up, the tumor in our case had decreased in size from 12 cm to 8 cm according to abdominal ultrasound (Fig. 3A). The residual liver mass was completely resected (Fig. 3B). Histopathology and immunostaining for SMA strongly confirmed the diagnosis of a glomus tumor of uncertain malignant potential. The patient had not experienced recurrence at a 2-year follow-up.

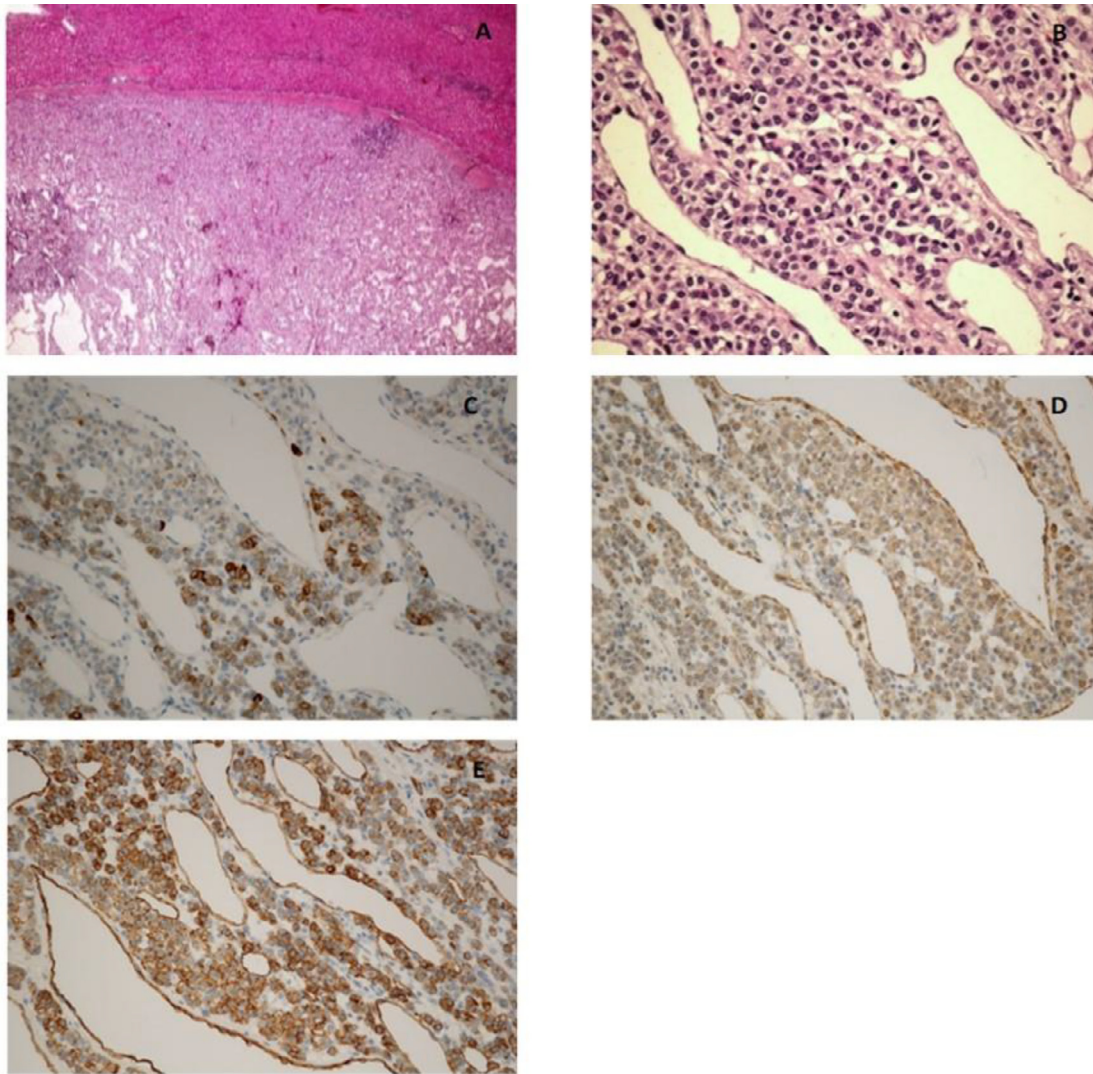


Fig. 4 – Histology of the tumor (A) A well-circumscribed mass composed of cellular neoplastic nodules with slit-like and dilated thin wall blood vessels (H&E 40 \times). (B) The neoplastic cells were uniform with round nuclei, fine chromatin, inconspicuous nucleoli, and pale eosinophilic cytoplasm, as well as well-defined cytoplasmic borders (H&E 400 \times). No atypia or mitotic figures were present. IHC study for h-caldesmon (C) showed reactivity. The tumor was also reactive to SMA (D) and CD34 (E).

Conclusions

Primary glomus tumors of the liver rarely occur in children. However, they should be considered in the differential diagnosis of hypervascular liver masses. Although most of these tumors are benign, malignant transformation in benign glomus tumors has been reported [21]. Therefore, tumor removal with preoperative embolization should be considered.

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The authors have no conflict of interest directly relevant to the content of this article.

Our institutional review board approved the study, and we obtained written informed consent from the patient.

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