Case Report

Extrahepatic transarterial radioembolization to treat fibrolamellar hepatocellular carcinoma: A case report

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Abstract

Fibrolamellar hepatocellular carcinoma (FL-HCC) is a rare primary liver tumor that typically presents at an advanced stage in early adolescents and adults with no underlying liver disease. Surgical resection is the first-line treatment, and patients who are not surgical candidates face limited treatment options with poor long-term outcomes. Herein we report the first documented, technically successful treatment of FL-HCC with extrahepatic spread using transarterial radioembolization (TARE) in a 16-year-old male patient with surgically unresectable disease. Subsequent imaging revealed tumor necrosis and a 20% reduction in size, and the patient survived 20 months post-treatment, a marked improvement relative to historical data in the literature. Further research should examine the potential role of yttrium-90 TARE in the treatment of FL-HCC patients with metastatic disease.

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Introduction

Fibrolamellar hepatocellular carcinoma (FL-HCC) is a primary liver tumor that usually presents at an advanced stage in early adolescents or adults at age 10–35 years with no underlying liver disease or cirrhosis [1]. FL-HCC often clinically manifests as abdominal fullness, pain, nausea, malaise, and weight loss; surgery is the only curative option at this time [2]. In a 2012 systematic review of 206 patients across 19 studies, 1 year and 5 year survival rates were 85% and 44%, respectively [3]. However, patients who are not surgical candidates experience limited treatment options with median overall survival of less than 12 months [4,5].

Although, liver-directed therapies such as transarterial chemoembolization (TACE) and transarterial radioembolization (TARE) are widely used in the treatment of HCC, their effectiveness in FL-HCC is poorly understood. In a 2018 case
A 16-year-old boy who initially presented with loss of appetite was diagnosed with FL-HCC in 2011 at an outside hospital and right trisegmentectomy was performed. He had no significant past medical history or family history otherwise. Informed consent was waived by our Institutional Review Board. Routine interval imaging in 2016 revealed a perihilar, subhepatic mass consistent with FL-HCC recurrence. Surgical resection was attempted but aborted due to the large size of the mass and a lack of suitable margins with involvement of the hepatic hilum, inferior vena cava, hepatic artery, right kidney, and hepaticojunostomy loop. The patient was treated with systemic chemotherapy and immunotherapy but experienced progression of his disease with enlarging size. Interventional radiology was consulted for consideration of locoregional therapy, with potential for subsequent surgical resection if the tumor was down-staged.

A computed tomography (CT) was performed and showed a tumor in the right upper abdomen measuring 10.0 × 7.5 × 14.0 cm (volume: 440 cm³) with tumoral invasion of the inferior vena cava, tumor thrombus near the confluence of the superior mesenteric vein and splenic vein, and regional nodal metastases (Fig. 1a). Whole body FDG-PET/CT at that time showed uptake by the large hepatic lesion, as well as fluorodeoxyglucose (FDG) uptake in prominent gastrohepatic, mesenteric and retroperitoneal lymph nodes. There were no other sites of disease. Mesenteric angiography to assess suitability for yttrium-90 TARE showed the majority of tumor supply was from the right inferior phrenic artery and the super-
rior mesenteric artery (Figs. 1b–c). TACE and bland embolization were considered but due to their limited efficacy in the authors’ experience, the decision was made to proceed with yttrium-90 TARE using resin microspheres. During TARE planning arteriography, nontumoral branches arising from the inferior phrenic (distal diaphragmatic and left branches) and superior mesenteric artery (jejunal branch to hepaticojejunostomy and inferior pancreaticoduodenal branch) were coiled to skeletonize tumoral blood supply (Fig. 2a). Embolization of parasitized extrahepatic vessels was consistent with other reports in the literature [8,9]. No tumoral supply was observed from the right renal and lumbar arteries. A Tc-99m MAA SPECT-CT confirmed absence of extratumoral uptake (Fig. 1d) and a lung shunt fraction of 8.2%.

TARE treatment was performed from a femoral approach using resin-based microspheres (Sirtex Medical, New South Wales, Australia). Radiation dosimetry using the Medical Internal Radiation Dose formula calculated 0.60 Gigabecquerel to each branch to achieve a target absorbed dose of 150 Gy. Arteriography from the treatment sites confirmed only tumoral enhancement (Fig. 2b). The superior mesenteric artery (SMA) tumor branch was treated using an antireflux catheter (TriSalus Medical Inc., Westminster, CO) to prevent nontarget embolization. The microcatheter was removed and disposed. A new 0.021in microcatheter was inserted to treat the right inferior phrenic artery branch. A Single-photon emission computed tomography (SPECT-CT) was performed after completion of radioembolization (Fig. 2c). No clinical symptoms or lab toxicities occurred after yttrium-90 radioembolization, including nausea, vomiting, or change in bowel habits. The patient was asymptomatic postprocedure and discharged after overnight observation.

Magnetic resonance imaging performed after radioembolization revealed necrosis and a 20% reduction in the size of the mass to 353 cm³ (Fig. 3). The patient developed progressive metastatic disease in the lungs and died 20 months after treatment.

Discussion

FL-HCC is a rare variant of HCC with limited treatment options beyond surgical resection. For unresectable FL-HCC, there are
no consensus guidelines; several management strategies have been described in the literature with mixed effectiveness and complication rates, including chemotherapy, ablation, and external beam radiotherapy [6,10,11].

TARE with yttrium-90 has demonstrated efficacy for FL-HCC in the liver with glass microspheres but has not been described in the treatment of extrahepatic FL-HCC. In this novel case report, we showed a 20% tumor volume reduction with no procedural complications. Thus, yttrium-90 TARE for extrahepatic FL-HCC metastases is feasible to perform with technical success. SMA branches may be radioembolized after careful planning arteriography, and tumor downstaging can be achieved. However, the long-term results of yttrium-90 TARE for extrahepatic FL-HCC metastases are not known, and all available treatment options should be considered for this rare tumor type.

FL-HCC is a primary liver tumor that typically presents in early adolescents and adults at an advanced stage. Patients who are not surgical candidates experience limited treatment options and have poor overall survival [3]. The effectiveness of liver-directed therapies such as TACE and TARE in the treatment of FL-HCC is not well understood. Here we present the first documented use of TARE to treat FL-HCC with extrahepatic spread. As a result of this case, further research should explore the potential role of yttrium-90 TARE as an option to reduce tumor volume in FL-HCC patients with metastatic disease.

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Fig. 3 – A 16-year-old male with fibrolamellar hepatocellular carcinoma with an axial contrast enhanced MRI of the abdomen obtained 3 months after radioembolization demonstrating reduction in tumor size (large arrows) to 9.0 x 7.3 x 13.0 cm³ (352 cm³) with areas of central necrosis (small arrows).