

Case report

Curative resection by superior hepatectomy for advanced hepatoblastoma facilitated by the presence of a large inferior right hepatic vein

BJ Dicken¹, DL Bigam² and AMJ Shapiro²

Department of ¹Surgery and Section of ²Hepatobiliary and Transplant Surgery, University of Alberta, Edmonton, Alberta, Canada

Background

Despite the success of neoadjuvant chemotherapy some patients with hepatoblastoma remain unresectable due to the proximity of important vascular structures. We report an unconventional surgical resection via a superior hepatectomy in a 16-month-old infant with hepatoblastoma.

Case outline

Staging CT scan revealed extensive replacement of the superior portion of the liver with complete occlusion of the three hepatic veins, and with extension into the inferior vena cava and right atrium. Following chemotherapy the tumour was confined to the superior portion of the liver with obstruction of the right, middle and left hepatic veins, but with

a large patent inferior hepatic vein draining the inferior liver segments. Superior hepatectomy was performed without complication.

Discussion

Complete surgical resection offers the only chance of cure for patients with hepatoblastoma. This case illustrates that careful preoperative planning facilitated aggressive surgical clearance with superior hepatectomy for curative resection of an otherwise non-resectable tumour.

Keywords

hepatoblastoma, surgical resection, liver tumour, superior hepatectomy

Introduction

In up to 40% of patients with hepatoblastoma, curative resection is not possible either because tumour invades key vascular structures or because extrahepatic disease is present [1]. Cure is dependent on complete excision of the primary tumour mass through standard or extended resection, or occasionally by orthotopic liver transplantation [2, 3]. This report details an atypical superior hepatectomy in a 16-month-old infant with hepatoblastoma.

Case report

A 16-month-old male infant presented with an asymptomatic abdominal mass in April 1999. Computed tomography (CT) scan demonstrated a large liver mass occupying the superior portion of the liver with extension and occlusion of all three major hepatic veins including the inferior vena cava and right atrium. A percutaneous

liver biopsy confirmed an epithelial-type hepatoblastoma and the patient received combination chemotherapy. Follow-up CT scan revealed complete regression of the extrahepatic disease. A large calcified mass occupying the superior aspect of the right and left lobes of the liver persisted. An exploratory laparotomy for possible right trisegmentectomy was undertaken; however, the tumour was found to extend into segment II and was considered to be unresectable. The child was referred to the hepatobiliary/transplant service for consideration of a liver transplant. Magnetic resonance imaging (MRI) studies confirmed that the tumour was confined within the superior portion of the liver, occupying segments II, IVA, VII and VIII, with complete occlusion of the left, middle and right hepatic veins (Figure 1). A contrast study of the inferior vena cava confirmed complete absence of flow in the right, middle and left hepatic veins, but demonstrated a large patent inferior hepatic vein draining a large portion of uninvolved hepatic parenchyma (Figure 2). Segments I, III, IVB, V and VI were

Correspondence to: Dr AM James Shapiro, 2000 College Plaza, 8215-112 Street, Edmonton, Alberta, Canada T6G 2C8 (e-mail: amjs@islet.ca)

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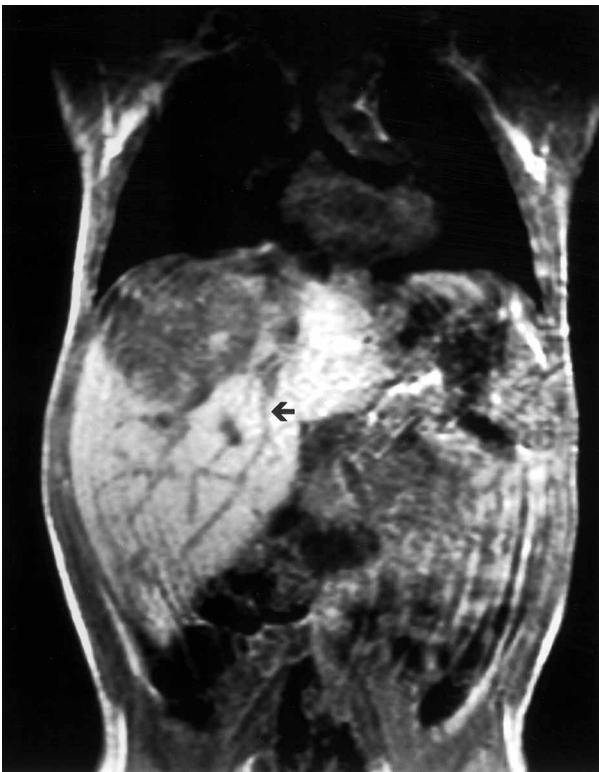


Figure 1. Preoperative coronal post-gadolinium T1-weighted MRI scan of a 16-month-old infant with hepatoblastoma, demonstrating absence of flow within all three hepatic veins, tumour encroachment on the inferior vena cava, and a large tumour mass occupying the superior segments of the liver. The arrow marks the presence of a prominent inferior accessory hepatic vein.

apparently free of tumour. Although this tumour was not amenable to conventional liver resection, superior hepatectomy with removal of all three hepatic veins and preservation of the inferior hepatic vein was considered a better alternative to transplantation. Given the risks of postoperative liver failure, a living related transplant work-up was conducted as a precaution.

Laparotomy was performed in October 1999 and revealed a large calcified tumour within the superior aspect of the liver occupying segments II, IVA, VII and VIII. The caudate lobe along with segments III, IVB, V and VI appeared to be free of tumour involvement. Proximal and distal vascular control was obtained, exposing the right, middle and left hepatic veins and carefully delineating the large inferior hepatic vein (Figure 3). Intra-operative ultrasound was conducted to assess tumour-free segments and their corresponding portal pedicles. The absence of tumour within the retrohepatic and superior hepatic vena cava with adequate venous drainage of the inferior liver segment

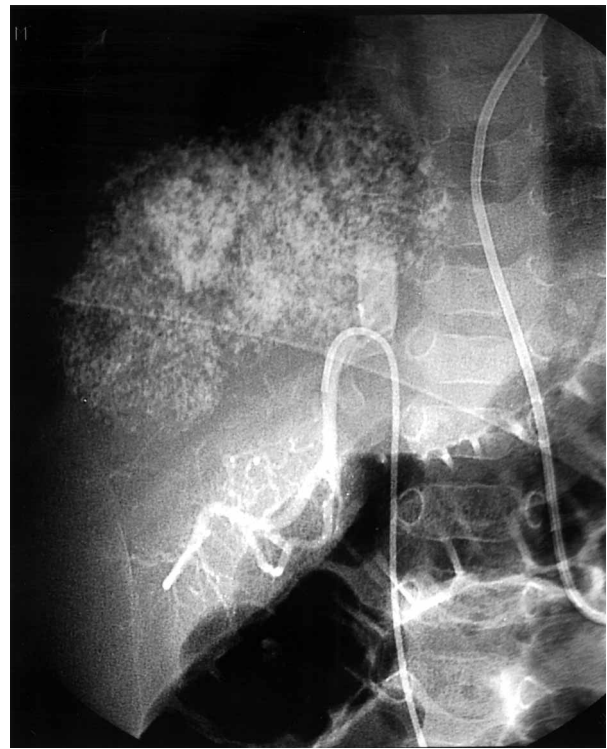


Figure 2. Contrast venogram of the retrohepatic vena cava, demonstrating a large inferior accessory hepatic vein and tumour occupying the superior portion of the liver, with obstruction of the inferior vena cava.

suggested that a superior hepatectomy could be performed in the absence of a caval replacement.

Total vascular occlusion was applied for 68 min, and a superior hepatectomy was performed. There was careful preservation of the portal pedicle branches to segments III, IVB, V and VI, with division of the pedicles to segments II, IVA, VII and VIII. Retrohepatic dissection was completed by dividing the right, middle and left hepatic veins, allowing removal of the specimen. Frozen section examination confirmed the absence of tumour within the hepatic veins. The patient made a full recovery and was discharged home on postoperative day 13.

Pathological examination revealed an epithelial hepatoblastoma measuring $11 \times 8 \times 4$ cm that was resected with microscopically negative margins. The pathological description demonstrated occlusion of the hepatic veins by fibrous and calcified tissue, but without residual tumour involvement.

The patient has remained well and is disease-free with no radiological or clinical evidence of recurrence at more than 4 years (January 2004) after operation.

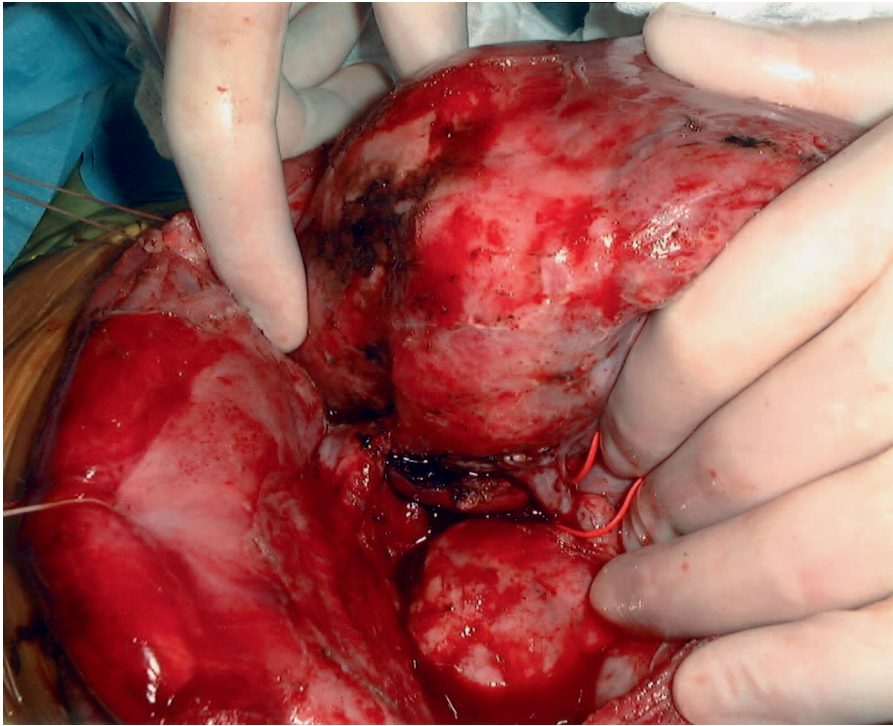


Figure 3. Intra-operative photograph demonstrating extensive tumour involvement of the superior portion of the liver with compression of the retrohepatic vena cava. Vascular loop marks preservation of a large inferior accessory hepatic vein.

Discussion

Delay in the diagnosis of hepatoblastoma is frequently associated with advanced disease at presentation, necessitating neoadjuvant chemotherapy and aggressive surgery. Some patients remain unresectable after chemotherapy owing to tumour involving key vascular structures. Orthotopic liver transplant is a potential alternative to extended resections, but requires lifelong immunosuppression and may accelerate proliferation of residual malignant cells [4–6].

Unconventional surgical techniques can be considered as an alternative to transplantation. La Quaglia *et al.* [7] advocated central hepatectomy for malignant tumours involving segments IV, V and VIII. Superina *et al.* [3] demonstrated the feasibility of an extended left hepatectomy in patients with hepatoblastoma involving all three hepatic veins, illustrating the importance of the retrohepatic veins in providing adequate liver drainage.

We have stressed the importance of careful preoperative planning for consideration of aggressive atypical hepatectomy in a non-resectable tumour. In our case, the recognition of the involvement of all three hepatic veins

with preservation of a large inferior vein allowed us to proceed with the planned superior hepatectomy.

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