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Aggressive recurrence of primary hepatic epithelioid hemangioendothelioma after liver transplantation

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CASE PRESENTATION

A 40-year-old woman with history of hypertension presented on April 2013 with three-month history of right upper abdominal pain, nausea, vomiting, jaundice, 6 kg weight loss and progressive abdominal distension. Physical examination revealed jaundice, hepatomegaly and hard liver with moderate ascites.

Laboratory investigations revealed normal prothrombin time/partial thromboplastin time and abnormal liver function: total bilirubin (26.5 mg/dL); direct bilirubin (22.2 mg/dL); aspartate aminotransferase (369 IU/L); alanine aminotransferase (49 IU/L); alkaline phosphatase (552 IU/L). Viral markers for hepatitis A, B and C were nonreactive. Serum alpha fetoprotein level was 2.6 µg/L.

Computed tomography (CT) of the chest, abdomen and pelvic showed diffuse neoplastic infiltrative involvement of the entire liver with sparing of small patchy areas of the liver with a mass effect. There was no evidence of metastasis on positron emission tomography scan and bone scan was negative.

Ultrasound (US)-guided liver biopsy was consistent with hepatic epithelioid hemangioendothelioma (HEHE). All tumour cells were positive for endothelial markers including CD31, CD34 & factor VIII-related antigen. The patient underwent living related donor liver transplantation on June 2013.

The explanted liver weighed 3222 g and measured 33 cm × 24 cm × 12 cm. The capsular surface showed foci of capsular retraction (Figure 1A). Cross-sections of the resected liver showed multiple white geographical areas alternating with the liver tissue, involving the entire surface area. The white areas were rimmed by hyperemic edge (Figure 1B).

Microscopic examination revealed diffuse infiltration by malignant epithelioid cells with signet ring morphology and massive areas of necrosis, the cells showed similar positive reaction to endothelial markers (Figures 2A to 2D). The tumour showed aggressive morphology with abundant vascular and neural infiltration. The gall bladder was extensively infiltrated by the same tumour cells, which reached the mucosal surface.

Postoperatively, her liver enzyme levels were normalizing and repeat liver Doppler US after transplantation revealed patent vessels and small hepatic hematoma. She was then discharged on prednisone, tacrolimus and mycophenolate mofetil (Cellcept, Genentech, USA)

One month later, she presented with severe abdominal pain and ascites and elevated liver enzyme levels. CT of the abdomen revealed innumerable diffuse hypodense lesions in the liver, enlarged periaortic, periportal and aortocaval lymph nodes noted in the areas and

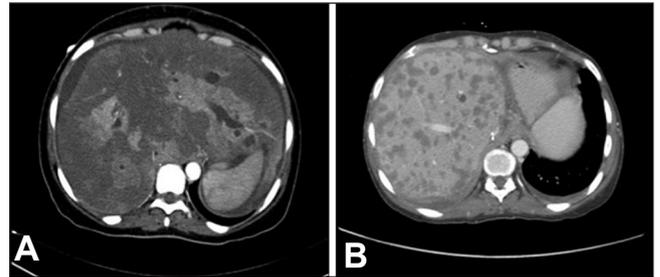


Figure 1 A Computed tomography (CT) abdomen revealing diffuse neoplastic infiltrative involvement of the entire liver, with sparing of small patchy areas of the liver. B CT of the abdomen showing post-liver transplant recurrence with innumerable diffuse hypodense lesions in the liver

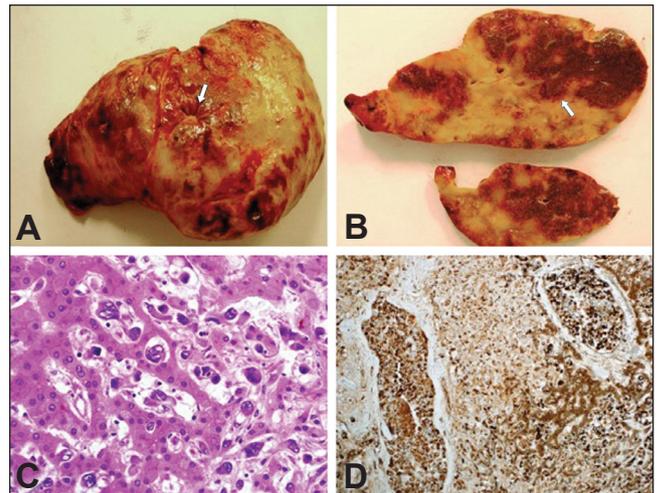


Figure 2 A Hepatectomy specimen with focal capsular retraction (arrow). B Cross-section of the liver revealing white sclerotic areas with hyperemic rim (arrow). C Malignant epithelioid hemangioendothelioma cells with sinusoidal infiltration (hematoxylin and eosin stain, original magnification ×40). D Tumour cells positive for CD31 and CD34 (immunohistochemical staining, original magnification ×40)

moderate ascites. Liver biopsy revealed recurrence of the disease. The patient received supportive treatment; unfortunately, she passed away one month later.

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DISCUSSION

The first series of 32 HEHEs was described in 1984 (1). Mehrabi et al reviewed 434 reported cases between 1984 and 2005, 44% of patients underwent liver transplantation (LT) with 96% and 54.5% one-year and five-year survival respectively (2).

Rodriguez et al (3) reviewed 110 patients diagnosed with HEHE who underwent 126 LTs between 1987 and 2005 from the United Network for Organ Sharing database. The one, three and five-year patient survival rates were 80%, 68%, and 64%, respectively; 32% of those who died were due to recurrent HEHE recurrent (3). Lerut et al (4) analyzed 59 patients reported in the European Liver Transplant Registry, the one-, five- and 10 year patient survival rates were 93%, 83%, and 72%, respectively. Nine (15.3%) patients died due to recurrent disease (4).

This patient was 40 years of age at the time of LT, similar to the average age reported by the European registry (4). There are six reported cases, including ours, receiving living donor LT for HEHE.

The explanted liver size was the largest reported in the literature (33 cm × 24 cm × 12 cm) compared with (17 cm × 14 cm × 13 cm), with an explanted liver weight of 3222 g compared with 1250 g. The large tumour burden may have contributed to the very early aggressive recurrence. The average time of recurrence is 49 months (range six to 98 months); but can occur up to 12 years post-LT (5). To our knowledge, this is the shortest recurrence after LT (within three months of LT).

CONCLUSION

Recurrence post-LT in HEHE must be explored and assessed further. Survival >10 years must also be documented and reported in the literature because recurrence is an unpredictable event post-LT.

DISCLOSURES: The authors have no financial disclosures or conflicts of interest to declare.

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