Case Report

Hepatic epithelioid hemangioendothelioma in a young nigerian presenting with massive hemorrhage: A case report and review of the literature

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Abstract

Hepatic epithelioid hemangioendothelioma (HEHE) is a rare sarcoma of vascular origin. Massive hemoperitoneum (due to spontaneous rupture) and hematemesis (due to hemobilia) are rarer manifestations of HEHE. We present a 22-year-old male with 6 weeks' history of worsening noncolicky right hypochondrial and epigastric pain that later became generalized. Initial clinical diagnosis of perforated duodenal ulcer was made. Emergency exploratory laparotomy revealed multiple hepatic and intraperitoneal tumors with hemoperitoneum. Histology of the biopsied masses revealed HEHE. He subsequently developed massive hematemesis secondary to hemobilia (which was confirmed at endoscopy) from the HEHE. Contrast-enhanced computed tomography done on account of rapidly increasing hepatomegaly demonstrated multiple peripherally enhancing hepatic masses and peritoneal nodules with hemoperitoneum. He was placed on Avastin-based chemotherapy because of the vascular proliferating nature of the tumor. Despite relatively stable postoperative period, our patient had repeated episodes of massive hemobilia and finally died after one cycle of chemotherapy.

Keywords: Epithelioid hemangioendothelioma, hemobilia, hemoperitoneum, hepatic

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INTRODUCTION

Hepatic epithelioid hemangioendothelioma (HEHE) is a rare vascular neoplasm with nonspecific clinical manifestations and variable imaging findings.[1] Its spontaneous rupture is extremely rare, and presentation with hematemesis from hemobilia is even rarer.[1] To the best of our knowledge, we are reporting the first case of HEHE with hemoperitoneum and hemobilia in a young Nigerian adult, highlighting the salient radiological features.

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CASE REPORT

A 22-year-old male presented at our hospital with 6 weeks' history of worsening noncolicky right hypochondrial and epigastric pain which became generalized 4 days

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before presentation. He had progressive abdominal distension, anorexia, and early satiety but no weight loss or hematemesis.

He was acutely ill-looking, severely pale, weak, tachycardiac, and dehydrated. Abdominal examination revealed generalized rebound tenderness, guarding, and hypoactive bowel sounds. Abdominal paracentesis yielded hemoperitoneum. The initial clinical diagnosis was perforated duodenal ulcer.

Urgent preoperative ultrasound (USS) revealed lobulated enlarged liver with heterogeneous parenchyma and intraperitoneal anechoic fluid with septations [Figure 1]. Findings on emergency exploratory laparotomy included 1.5 L of hemoperitoneum, hepatomegaly with multiple nodular and cystic lesions over the diaphragmatic and inferior surfaces of the liver [Figure 2], with ruptured nodules on its inferior surface. Biopsy was taken for histology and the abdomen was closed up.

Histology revealed numerous slit-like channels lined by atypical malignant endothelial cells with hyperchromatic nuclei and thin rim cytoplasm and surrounded by hyaline stroma [Figure 3]. The histologic diagnosis was HEHE.

Postsurgery, he had several episodes of hematemesis which necessitated a cumulative transfusion with 32 pints of blood. Endoscopy showed blood refluxing from the 3rd part of the duodenum, without esophageal varices, ulcers, or gastric lesion, suggesting hemobilia.

Rapidly increasing postsurgery hepatomegaly warranted an abdominopelvic contrast-enhanced computed tomography (CECT) which showed multiple hypodense hepatic masses and peritoneal nodules with peripheral enhancement (PE) in arterial and portal venous (PV) phases [Figure 4]. The PE was not seen in the delayed phase and intra-abdominal collection was noted [Figure 5].

He was subsequently placed on bevacizumab (Avastin)-based chemotherapy (adriamycin and 5FU) but died after the first cycle.

DISCUSSION

Epithelioid hemangioendothelioma (EHE), first defined as a specific entity in 1982,[2] arises from vascular endothelium and is usually intermediate between hemangioma and angiosarcoma.[3] EHE has been described in many organs such as liver, spleen, bone, brain, meninges, breast, heart, head and neck soft tissues, stomach, and lymph nodes.[4] Our patient had histologically confirmed EHE of the liver.

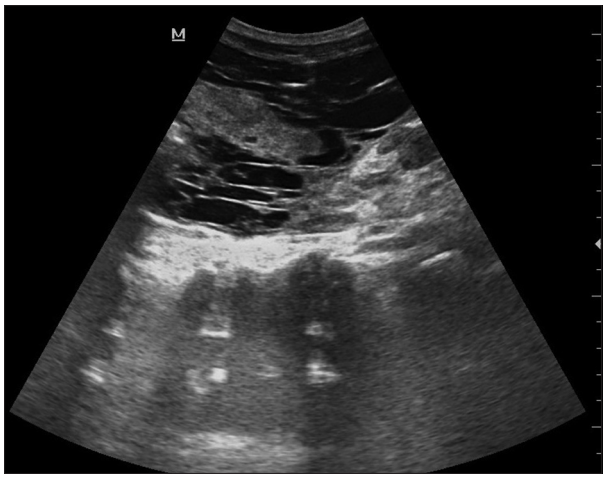

Figure 1: B-mode USS of the abdomen showing anechoic collection with septations in the abdominal cavity

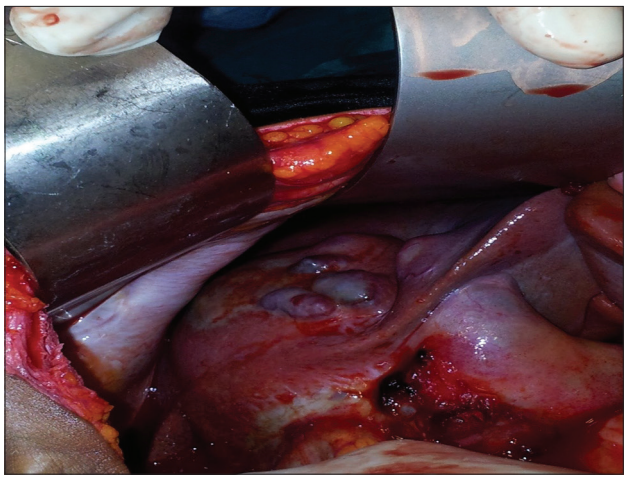

Figure 2: Multiple tumor nodules on the diaphragmatic surface of the liver seen at surgery

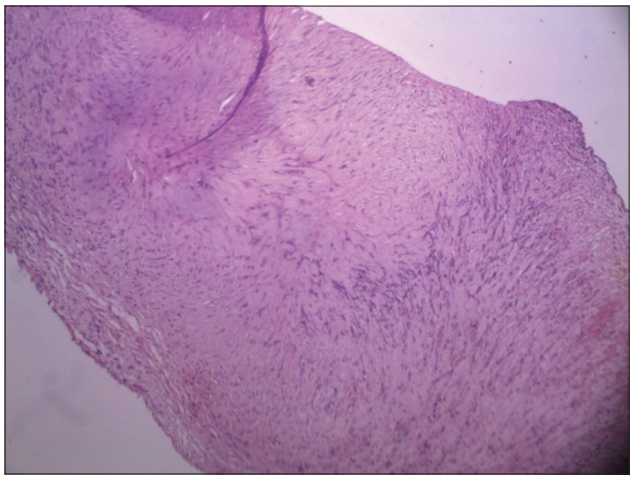

Figure 3: Histologic section showing numerous slit-like channels lined by atypical endothelial cells and surrounded by hyaline stroma, (H and E)

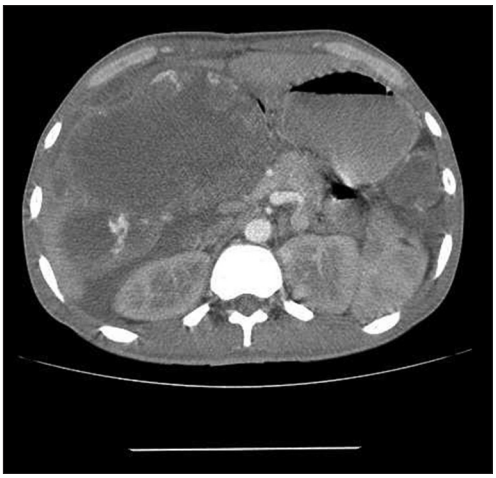

Figure 4: Axial contrast-enhanced computed tomography of the abdomen in arterial phase showing hypodense hepatic and intraperitoneal masses (anterior to the spleen) with peripheral enhancement and intra-abdominal hypodense collection (in hepatorenal pouch)

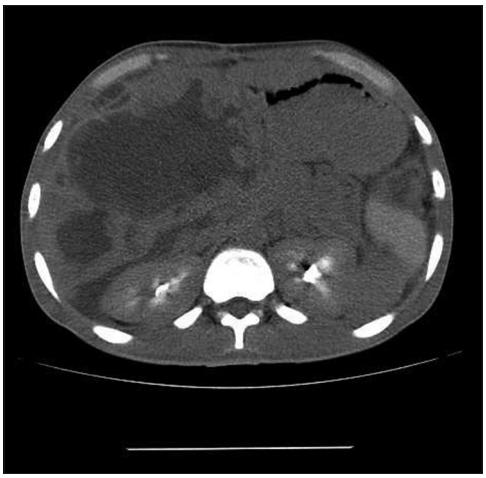

Figure 5: Axial contrast-enhanced computed tomography of the abdomen in delayed phase showing the hypodense hepatic masses and intraperitoneal tumor nodule (open arrow) which have lost their peripheral enhancement

It is rare in children <15 years and more common in middle-aged women with a male-to-female ratio of 2:3.[5] Our patient was a 22-year-old male.

Although there is no known predisposing factor to this tumor, it has been reported in vinyl chloride workers, asbestos workers, women on oral contraceptives, alcoholics, chronic liver disease (CLD), hepatitis B and C patients and following hepatic trauma,[6] none of which was present in this patient.

Clinical manifestations of HEHE include right-upper quadrant (RUQ) pain, abdominal distension, weight loss, portal hypertension, Budd–Chiari syndrome, and liver failure.[5] Our patient presented with RUQ pain and abdominal distension. Rarely, there is a spontaneous rupture producing hemoperitoneum.[1] Our patient had massive hemoperitoneum at presentation and hemobilia postsurgery.

Radiological features of HEHE can be demonstrated on CECT, contrast-enhanced magnetic resonance imaging (CE-MRI), USS, and scintigraphy.[7]

On CECT, multifocal hypodense tumors extending to the periphery of the liver that show PE on arterial and PV phases with loss of the PE on delay phase as seen in our patient are typical for HEHE.[7] In addition, extensive tumors may become confluent and diffuse while some tumors may become isodense to liver parenchyma on CE.[7] Capsular retraction, calcification, compensatory hypertrophy of the unaffected liver segments, and features of portal hypertension may be seen,[7] all of which were absent in this case.

Our patient did not do MRI due to its relatively high cost which would have depicted internal architecture of HEHE better than computed tomography.[7,8] On T1-weighted MRI, HEHE is hypointense with some lesions containing central areas of lower signal intensity than the remainder of the tumor.[7,8] On T2-weighted MRI, HEHE is heterogeneously hyperintense and may have a target appearance due to the presence of central hypointense zone (representing areas of hemorrhage, coagulation necrosis, and calcification) and peripheral hyperintense zone of cellular proliferation.[7,8] On CEMRI, PE occurs with a thin nonenhancing rim corresponding to a narrow avascular zone between normal liver parenchyma and the nodules.[8]

Differential diagnoses of HEHE include hemangioma, hepatocellular cancer (HCC), peripheral cholangiocarcinoma (PC), and metastasis. Hemangioma shows nodular PE on arterial phase with centripetal filling and eventual homogeneous enhancement (except in region of central scar and thrombosis)[9] on delay phase unlike HEHE with continuous PE (in the arterial and PV phases) which is lost on delay phase. HCC which is associated with CLD has variable enhancement which is rarely peripheral in arterial phase and becomes isodense on PV phase, unlike HEHE that retains its PE (in PV phase).[9] PC is associated with capsular retraction, similar to HEHE and intrahepatic bile duct dilatation, and shows heterogeneous enhancement with retention of contrast on delayed phase,[9] unlike HEHE. Metastases are usually multifocal like HEHE but usually show enhancement only in the arterial phase, unlike HEHE that maintains its PE into the PV phase.[9]

USS done in our patient showed intra-abdominal collection with septations suggesting inflammatory or neoplastic cause. The liver was enlarged and diffusely heterogeneous in our patient; however, discrete liver nodules be seen in some patients[7,8] which are mostly hypoechoic, but may be hyperechoic or isoechoic relative to background normal liver.

Scintigraphy was not done in our patient due to nonavailability in our environment. Decreased perfusion of involved areas with increased blood flow to uninvolved areas of the liver is seen on scintigraphy.[7]

Orthotopic liver transplantation is the best option in treatment as resection is not usually possible due to the multifocality of the tumor, with variable benefit from adjuvant chemotherapy or radiation when total resection is not possible.[10] Current chemotherapeutic approaches are directed at preventing endothelial proliferation. Our patient was commenced on bevacizumab-based chemotherapy (an inhibitor of vascular endothelial growth factor) which has been used in the literature.[10] He only had a single cycle of it before his demise.

There could be metastases to peritoneum, lymph nodes, omentum, muscle, skin, and mesentery,[6] and mortality rate in patients with metastatic disease is >60%.[10] The overall prognosis is considered more favorable than that of other hepatic malignancies[2,10] Our patient died about 2 months after presentation, from severe anemia secondary to several episodes of hematemesis from hemobilia.

CONCLUSION

This is a case of a 22-year-old male student with massive hemoperitoneum and hemobilia caused by ruptured HEHE. The patient died after one course of bevacizumab-based chemotherapy.

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Conflicts of interest
There are no conflicts of interest.

REFERENCES

1. Earnest F 4th, Johnson CD. Case 96: Hepatic epithelioid hemangioendothelioma. Radiology 2006;240:295-8.
2. Zhang W, Jambulingam PS, Silva MA, Taniere P, Bramhall SR, Mayer AD, *et al*. Orthotopic liver transplantation for epithelioid haemangioendothelioma. Eur J Surg Oncol 2007;33:898-901.
3. Mohtasham N, Kharrazi AA, Jamshidi S, Jafarzadeh H. Epithelioid hemangioendothelioma of the oral cavity: A case report. J Oral Sci 2008;50:219-23.
4. Nudo CG, Yoshida EM, Bain VG, Marleau D, Wong P, Marotta PJ, *et al*. Liver transplantation for hepatic epithelioid hemangioendothelioma: The Canadian multicentre experience. Can J Gastroenterol 2008;22:821-4.
5. Uchimura K, Nakamuta M, Osoegawa M, Takeaki S, Nishi H, Iwamoto H, *et al*. Hepatic epithelioid hemangioendothelioma. J Clin Gastroenterol 2001;32:431-4.
6. Chen HL, Bair MJ, Lin IT, Li CH, Chen YJ, Shih SC. An unusual case of hepatic tumour in an elderly patient. Int J Gerontol 2009;3:188-91.
7. Lyburn ID, Torreggiani WC, Harris AC, Zwirewich CV, Buckley AR, Davis JE, *et al*. Hepatic epithelioid hemangioendothelioma: Sonographic, CT, and MR imaging appearances. AJR Am J Roentgenol 2003;180:1359-64.
8. Kehagias DT, Moulopoulos LA, Antoniou A, Psychogios V, Vourtsi A, Vlahos LJ. Hepatic epithelioid hemangioendothelioma: MRI findings. Hepatogastroenterology 2000;47:1711-3.
9. Dahnert W. Radiology Review Manual. 7th ed. Philadelphia: Lippincott Williams and Wilkins; 2011. p. 735-40.
10. Barger J, Tanweer O, Liechty B, Snuderl M, Jafar JJ. Suprasellar epithelioid hemangioendothelioma: Case report and review of the literature. Surg Neurol Int 2016;7 Suppl 23:S596-602.