


Variceal bleeding caused by oesophageal varices: A manifestation of hepatocellular carcinoma in a 17-year-old girl

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Abstract

The authors present the rare case of a 17-year-old girl referred to the medical assessment unit following a large upper gastrointestinal haemorrhage. On further evaluation, she was found to have a primary hepatocellular carcinoma with extensive metastases. In our patient, the tumour was deemed incurable by resection, liver transplantation or percutaneous treatment. She underwent palliative chemotherapy and passed away 11 months following presentation.

Keywords

Hepatocellular carcinoma, variceal bleeding, paediatric

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Introduction

Primary hepatocellular carcinoma (HCC) is a tumour of the liver, which most often occurs in the setting of chronic liver disease and cirrhosis. This following report merits review as it describes a rare case of a 17-year-old with no prior risk factors for HCC, presenting to hospital with acute variceal bleeding, which on further evaluation revealed a primary HCC with extensive metastases.

Case

The patient's guardian has given consent for this study to be published in a scientific journal. This case does not require ethics approval.

A 17-year-old Caucasian female, who was previously fit and well, presented to the medical assessment unit at a district general hospital following an episode of haematemesis. The vomitus contained approximately 200 mL of bright red blood with large clots. There was no abdominal pain. She had been in her previous state of good health up until 1 month prior to admission when she reported a 3-week history of anorexia and passing melanotic stools. Her last menstrual period was 3 months prior to presentation. She denied weight loss. Her past medical history was remarkable for osteomyelitis as a child. She had no recent foreign travel, and both parents were British Caucasian with no risk factors for chronic viral hepatitis.

On physical examination, she was haemodynamically stable and alert. Her abdominal examination revealed hepatosplenomegaly with ascites and mild peripheral oedema. She required five units of packed red cells and was started on a continuous infusion of omeprazole, as out of hours endoscopy was not available. She also received terlipressin every 4 h.

She had relatively high concentrations of aspartate aminotransferase (124 U/L), alkaline phosphatase (671 U/L), gamma glutamic pyruvic transaminase (253 U/L), alpha fetoprotein (AFP, 27 ng/mL) and C-reactive protein (104.5 mg/L). Her haemoglobin on presentation was 6.5 g/dL, and her leukocyte count was 14.6 ($\times 10^9/L$). A chronic liver disease screen was done, which included hepatitis serology, autoimmune hepatitis screen, antinuclear antibody (ANA), anti-mitochondrial antibody, smooth muscle antibody, CEA and CA 19-9, which were all negative. She had an urgent upper gastrointestinal endoscopy where she had a further episode of melaena and haematemesis dropping her haemoglobin level to 8.0 g/dL. Upper G.I. endoscopy done

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within 24 h revealed a large clot occupying the fundus of the stomach with fresh bleeding beneath. Her chest radiography showed widespread metastatic lesions and a large right hilum. Her abdominal ultrasound scan revealed a large mass in the right lobe of the liver with an enlarged spleen. Computed tomography (CT) scanning of her chest, abdomen and pelvis confirmed a primary hepatic malignancy staged T4 N1 M1. A liver biopsy confirmed the presence of HCC of the fibrolamellar variant (FL-HCC), which was diffusely positive for CK 17.

She was commenced on three cycles of palliative chemotherapy with sorafenib and epirubicin, cisplatin and capecitabine. However, she passed away in 11 months following initial admission.

Discussion/conclusion

HCC is the third leading cause of cancer-related deaths globally, with over 80% of cases being related to underlying chronic infection with hepatitis B and C virus.¹ In the United Kingdom, age-standardised rates for HCC are 6 per 100,000 in males and 3 per 100,000 in females. It is, however, the second and third fastest growing cancer in males and females, increasing by 38% and 28%, respectively, in the last decade.² This is attributed to cirrhosis of the liver secondary to alcoholic liver disease, viral hepatitis and non-alcoholic steatohepatitis.² HCC is particularly rare in the paediatric population within developed countries accounting for one child per year developing HCC in the United Kingdom.³

This case is also important and it describes a unique case of HCC in the paediatric population within the United Kingdom. In our patient, the fibrolamellar variant of liver cancer (FL-HCC) was observed. FL-HCC was first described by Edmondson⁴ in 1956. It differs from typical HCC in that it usually arises in a noncirrhotic liver, has a predilection for younger patients and the levels of AFP are normal or only marginally elevated.⁵ It remains a rare type of liver cancer with typical HCC still accounting for 60%–80% of all liver cancers.⁵ It was noted that 80% of FL-HCC presents before the age of 35, while only 11% occurs after 40 years of age.⁵ While some studies have suggested that FL-HCC has a better prognosis than typical HCC, other studies have not shown any statistically significant difference in outcomes.^{6–9} The 5-year survival rate for FL-HCC has been estimated at 37%–76%. The prognosis is dependent on tumour resectability and stage of disease with inoperable tumours with extensive metastases carrying a worse prognosis.⁵ This case is unique as it describes a FL-HCC occurring in the presence of cirrhosis with variceal bleeding. It also highlights the fact that these tumours present at an advanced stage where there is limited scope for surgical resection and thus the overall prognosis would be poor.

In addition, only 1% of cases of HCCs present with variceal bleeding.¹⁰ There were no documented cases in the literature of a paediatric patient with no prior risk factors for HCC presenting with variceal bleeding. Sachdeva et al.¹¹

presented the care of a 12-year-old girl who presented with variceal bleeding but positive hepatitis B serology. Okusaka et al.¹² and Kutz and Miah¹³ presented cases of upper gastrointestinal haemorrhage caused by direct invasion of the tumour into the duodenum; however, these patients had hepatitis C infection and cirrhosis secondary to alcoholic liver disease, respectively.

In this patient, a chest X-ray showing extensive metastases increased the suspicion of a lymphoma or a HCC. The latter was thought to be less likely given her age, race and negative hepatitis B and hepatitis C serology. However, ultrasonography findings and the presence of decompensated liver disease increased the suspicion of a primary liver tumour. Differential diagnosis at the early stage of illness was difficult especially in this patient with no prior risk factors for liver cancer. FL-HCC usually presents at an advanced stage with mild to moderate abdominal pain, weight loss and early satiety or with a mass in the upper abdomen. Rarely can it present with obstructive jaundice, diarrhoea or bone pain. In our patient, the tumour was deemed incurable by resection, liver transplantation or percutaneous treatment.

Declaration of Conflicting Interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship and/or publication of this article.

Ethics Approval

Our institution does not require ethical approval for reporting individual cases or case series.

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Informed Consent

Written and verbal informed consent was obtained from a legally authorised representative for anonymised patient information to be published in this article.

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