

An unusual cause of gastric extraluminal compression: fibrolamellar hepatocellular carcinoma

Case Report

Salvatore Dilettoso¹, Mario Uccello*², Alessandra Dilettoso¹,
Salvatore Gelardi¹, Benedetto Dilettoso¹

*1 ASP 3 Catania, Radiology Operative Unit, SS Salvatore Hospital,
Via Livorno 1, I-95047 Paternò, Italy*

*2 Department of Biomedical Sciences, University of Catania,
Via Androne 83, I-95124 Catania, Italy*

Received 17 April 2012; Accepted 27 July 2012

Abstract: Fibrolamellar hepatocellular carcinoma (FLH) is a rare variant of hepatocellular carcinoma that typically occurs in adolescents or young subjects in an otherwise normal liver. Correct recognition of FLH is extremely important, because a complete resection of the tumor can confer an excellent prognosis. FLHs can become large, exerting mass effects on adjacent organs before diagnosis. Unfortunately, there are very few imaging reports describing external compression of the stomach caused by a FLH. We report the case of a young female suffering from dyspepsia and abdominal pain caused by stomach compression resulting from a large FLH.

Keywords: *Fibrolamellar hepatocellular carcinoma • Liver neoplasms • Computerized tomography • Extraluminal compressive tumors • External compression of the stomach*

© Versita Sp. z o.o.

Abbreviations and acronyms

FLH = fibrolamellar hepatocellular carcinoma;
HCC = hepatocellular carcinoma;
CT = computerized tomography.

1. Introduction

Fibrolamellar hepatocellular carcinoma (FLH) is a rare variant of hepatocellular carcinoma (HCC) with unique clinical and histological features. Unlike conventional HCC, FLH occurs predominantly in young adults without chronic liver disease [1]. Correct recognition of FLH is fundamental, because complete resection of the tumor can effectively confer an excellent prognosis [1-3]. Nevertheless, FLH remains a deadly disease and frequently exhibits aggressive behavior [2]. Moreover, it can reach a large size before diagnosis, invading or compressing adjacent organs. As a result, clinical presentation is variable, and most patients can be asymptomatic or present with nonspecific gastrointestinal symptoms and signs.

Results of laboratory tests often demonstrate normal values or a slight increase in transaminase and alpha-fetoprotein levels [1,4]. A CT scan and other imaging modalities may be very helpful to physicians in both initial assessment and surveillance for recurrence, whereas pathology remains the gold standard in confirming a diagnosis of FLH [5].

2. Case report

A 28-year-old female was admitted with dyspepsia and vague abdominal pain that had persisted for one month; it was located over the left upper quadrant of the abdomen. She had no previous history of medical illness or excessive alcohol intake, nor were body weight loss, fever, nausea, vomiting, tea-colored urine, clay-colored stools, or jaundice indicated. Moreover, she denied a history of blood transfusion, foreign travel, and liver disease. Ultrasound examination revealed a heterogeneous solid mass measuring 7 cm in diameter over the left hepatic lobe. Biochemical tests showed that

* E-mail: mario_uccello@hotmail.it

the serum level of alanine aminotransferase, aspartate aminotransferase, alkaline phosphatase, total bilirubin, lactate dehydrogenase, proteins and albumin, alpha-fetoprotein, carcinoembryonic antigen, and carbohydrate antigen 19-9 were within normal values. The results for hepatitis B surface antigen and anti-hepatitis C virus antibody were negative. A CT scan confirmed the presence of a well-defined, hypodense liver mass with a calcification within a central scar (Figure 1) and areas of necrosis in an otherwise normal liver (Figure 2). The tumor was located in the segment II of the liver, had close anatomic relations of contiguity with the anterior gastric wall, and displaced and compressed the body and fundus of the stomach (Figure 3). A heterogeneous enhancement of the lesion was seen after contrast injection (Figure 4). The characteristic imaging findings and biopsy results led to a final diagnosis of FLH. The tumor was then completely resected with adequate margins, leading to full recovery from the initial clinical conditions. The patient remained asymptomatic and in stable condition, and postoperative CT scan showed no evidence of recurrence after a 6-month follow-up period.

3. Discussion

FLH is a distinctive subtype of HCC. Most conventional HCCs are detected in men older than 50 years who present with underlying liver cirrhosis or chronic liver disease [6,7], whereas FLH is typically not associated with liver disease [1]. By contrast, FLH classically manifests as a large hepatic mass in adolescents or young adults, with no gender predilection. Patients usually present with nonspecific gastrointestinal symptoms. Abdominal pain and malaise are the most common complaints in FLH; hepatomegaly or an abdominal mass is sometimes noted, and jaundice is occasionally found. Many patients also suffer from multiple signs and symptoms such as fever, diarrhea, and weight loss [1,8]. FLH often exhibits aggressive behavior, invading the main portal vein, the proper hepatic artery, the central intrahepatic bile ducts, or other adjacent organs, and/or presenting with extrahepatic lymph node or distant metastases [2,3,9]. Ultrasonography with color Doppler examination may represent a quick, noninvasive aid for the detection of a primary liver tumor and can distinguish it from



Figure 1. Nonenhanced CT scan demonstrates the presence of a hypodense mass with calcification within the central scar over the left hepatic lobe.



Figure 2. Portal venous CT image depicting hypodense areas of necrosis.



Figure 3. Hepatic arterial phase CT scan. The mass of about 7 cm in diameter exerts compression on the stomach.



Figure 4. Hepatic arterial phase CT scan shows a large mass with intense heterogeneous enhancement. An afferent vessel that supplies the tumor is clearly visible.

a liver metastasis [10]. Radiologically, FLH appears as a heterogeneous, sharply demarcated or lobulated mass, and is frequently localized in the left hepatic lobe. On CT scan, the tumor demonstrates heterogeneous enhancement on hepatic arterial phase images and often presents with calcification within the central scar [2,3,9]. A central scar is detectable in approximately 70% of FLHs and is composed of fibrous tissue emerging at the center of the lesion. A central scar can be seen in focal nodular hyperplasia and cavernous hemangioma as well, but central calcification is uncommon and its presence argues against the diagnosis of these neoplasms. Thus, when present, a central scar with calcification is a useful imaging feature that may suggest a diagnosis of FLH in patients with solitary liver masses [9]. In the case under discussion, we also noted massive areas of necrosis within the tumor that represent an infrequent finding in FLH and are typical of conventional HCC [2,3,9]. CT and MR images are helpful for initial assessment, staging, and monitoring for recurrence of FLH, whereas the diagnosis of FLH requires histological demonstration of the characteristic polygonal cells with abundant eosinophilic cytoplasm clustered in small groups separated by thick fibrous lamellae [5]. Therefore, liver biopsy is still considered the gold standard instrument for a correct differential diagnosis from other nodular liver

neoplasms. Such lesions include conventional HCC, hepatic adenoma, focal nodular hyperplasia, cavernous hemangioma, hepatoblastoma, and liver metastases [1-3,5,9]. Our patient complained of dyspepsia resulting from gastric compression. The most common sources of extragastric compressions are adjacent normal organs, and both benign pathologic and malignant lesions. Although several organs can cause extrinsic compression of the stomach, under normal anatomical conditions, the spleen and splenic vessel are the most common causes of compression after gallbladder. When the stomach is compressed by a malignant tumor, liver metastatic tumors and conventional HCC represent the most common causes of external compression of the anterior wall of the gastric body [11]. There are very few reports describing dyspepsia resulting from gastric compression in FLH [3,8]. However, clinicians should take into consideration the existence of this unusual presentation, which can mimic a functional disorder of the stomach, to facilitate prompt identification of the tumor.

Acknowledgements

Authors thank Alessia Trovato for relevant contribution to language revision.

References

- [1] Craig J.R., Peters R.L., Edmondson H.A., Omata M., Fibrolamellar carcinoma of the liver: a tumor of adolescents and young adults with distinctive clinicopathologic features, *Cancer*, 1980, 46, 372-379
- [2] Ichikawa T., Federle M.P., Grazioli L., Marsh W., Fibrolamellar hepatocellular carcinoma: pre- and posttherapy evaluation with CT and MR imaging, *Radiology*, 2000, 217, 145-151
- [3] Ichikawa T., Federle M.P., Grazioli L., Madariaga J., Nalesnik M., Marsh W., Fibrolamellar hepatocellular carcinoma: imaging and pathologic findings in 31 recent cases, *Radiology*, 1999, 213, 352-361
- [4] Berman M.A., Burnham J.A., Sheahan D.G., Fibrolamellar carcinoma of the liver: an immunohistochemical study of nineteen cases and a review of the literature, *Hum. Pathol.*, 1988, 19, 784-794
- [5] Liu S., Chan K.W., Wang B., Qiao L., Fibrolamellar hepatocellular carcinoma, *Am. J. Gastroenterol.*, 2009, 104, 2617-2624
- [6] Bosch F.X., Ribes J., Cléries R., Díaz M., Epidemiology of hepatocellular carcinoma, *Clin. Liver Dis.*, 2005, 9, 191-211
- [7] Uccello M., Malaguarnera G., Pelligra E.M., Biondi A., Basile F., Motta M., Lipoprotein(a) as a potential marker of residual liver function in hepatocellular carcinoma, *Indian J. Med. Paediatr. Oncol.*, 2011, 32, 71-75
- [8] Arista-Nasr J., Gutierrez-Villalobos L., Nuncio J., Maldonado H., Bornstein-Quevedo L., Fibrolamellar hepatocellular carcinoma in Mexican patients, *Pathol. Oncol. Res.*, 2002, 8, 133-137
- [9] Blachar A., Federle M.P., Ferris J.V., Lacomis J.M., Waltz J.S., Armfield D.R., et al., Radiologists' performance in the diagnosis of liver tumors with central scars by using specific CT criteria, *Radiology*, 2002, 223, 532-539
- [10] Bogdanov M., Perisic M., Jurisic V., Stojacic-Djenic S., Kerkez M.M., Ultrasonographic examination of the liver circulation in patients with liver metastasis from colorectal cancer, *J BUON*, 2010, 15, 378-381
- [11] Chen T.K., Wu C.H., Lee C.L., Lai Y.C., Yang S.S., Tu T.C., Endoscopic ultrasonography to study the causes of extragastric compression mimicking gastric submucosal tumor, *J. Formos. Med. Assoc.*, 2001, 100, 758-761