World Journal of Gastroenterology

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DOI: 10.3748/wjg.v21.i3.1028

World J Gastroenterol 2015 January 21; 21(3): 1028-1031
ISSN 1007-9327 (print) ISSN 2219-2840 (online)

CASE REPORT

Focal nodular hyperplasia coexistent with hepatoblastoma in a 36-d-old infant

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Supported by National Key Clinical Specialty Construction Programs of China (2014-2016); and Medical Guide Project of Shanghai Municipal Science and Technology Commission, No. 134119a4100 (to Qiao ZW).

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Received: April 26, 2014
Peer-review started: April 30, 2014
First decision: June 10, 2014
Revised: July 3, 2014
Accepted: September 5, 2014
Article in press: September 5, 2014
Published online: January 21, 2015

Abstract

Focal nodular hyperplasia (FNH) is a benign hepatic tumor characterized by hepatocyte hyperplasia and a central stellate scar. The association of FNH with other hepatic lesions, such as adenomas, hemangiomas and hepatocellular carcinoma, has been previously reported, but FNH associated with another hepatic tumor is rare in infants. Here we report a case of FNH coexistent with hepatoblastoma in a 36-d-old girl. Computed tomography (CT) imaging showed an ill-delineated, inhomogeneous enhanced mass with a central star-like scar in the right lobe of the liver. The tumor showed early mild enhancement at the arterial phase (from 40HU without contrast to 52HU at the arterial phase), intense enhancement at the portal phase (87.7HU) and 98.1HU in the 3-min delay scan. A central scar in the tumor presented as low density on non-contrast CT and slightly enhanced at delayed contrast-enhanced scanning. This infant underwent surgical resection of the tumor. Histopathology demonstrated typical FNH coexistent with a focal hepatoblastoma, which showed epithelioid tumor cells separated by proliferated fibrous tissue.

Key words: Focal nodular hyperplasia; Hepatoblastoma; Infant; Computed tomography

Core tip: Focal nodular hyperplasia (FNH) is infrequent in infants, and hepatoblastoma is the most common primary malignant liver tumor in infants. The case reported here was a 36-d-old girl suffering from FNH coexistent with hepatoblastoma. Computed tomography imaging showed an ill-delineated, inhomogeneous enhanced mass with a central star-like scar in the right lobe of the liver. The patient underwent surgical resection of the tumor, and histopathology demonstrated typical FNH coexistent with a focal hepatoblastoma.

Gong Y, Chen L, Qiao ZW, Ma YY. Focal nodular hyperplasia coexistent with hepatoblastoma in a 36-d-old infant. *World J Gastroenterol* 2015; 21(3): 1028-1031 Available from: URL: http://www.wjgnet.com/1007-9327/full/v21/i3/1028.htm DOI: http://dx.doi.org/10.3748/wjg.v21.i3.1028

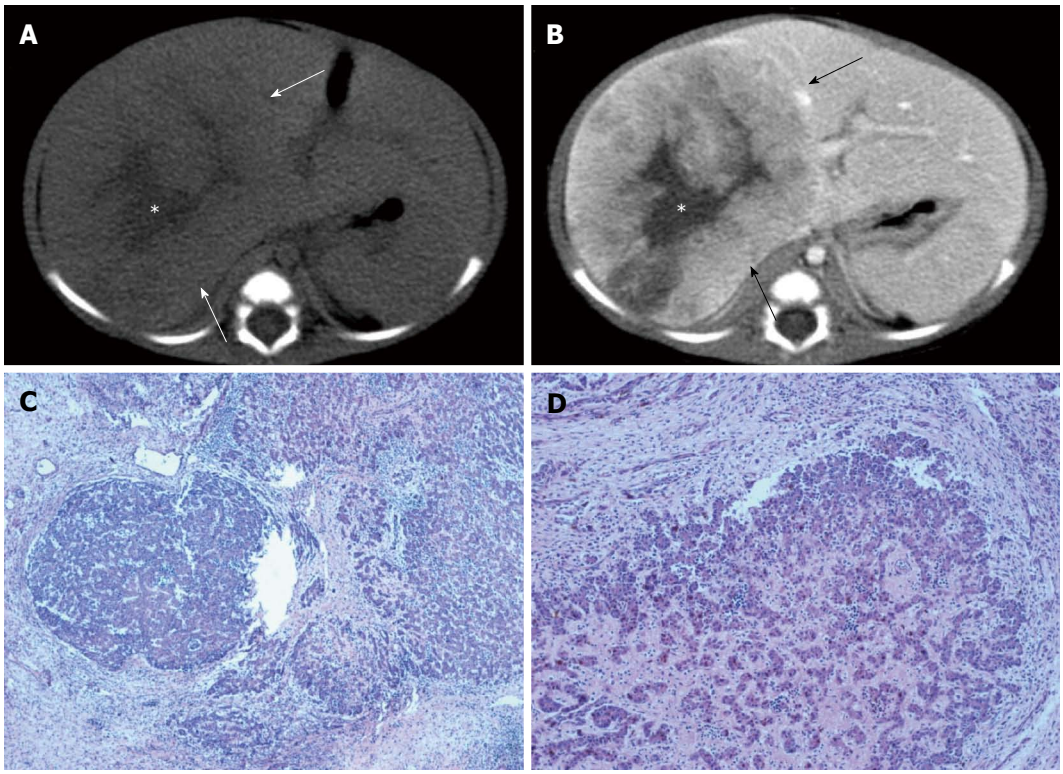

Figure 1 Computed tomography findings. A: Non-contrast computed tomography (CT) of the liver showed a slightly hypo-dense mass in the right lobe of the liver (arrow) with a hypo-dense central star-like scar (asterisk); B: Contrast-enhanced CT of the liver in the portal venous phase showed inhomogeneous and intense enhancement of the mass (arrow), with the central star-like scar (asterisk); C: Histopathology showed typical focal nodular hyperplasia, with hepatocytic nodules separated by bands of fibrous tissue (HE stain, original magnification × 50); D: Foci of hepatoblastoma showed epithelioid tumor cells separated by proliferated fibrous tissue (HE stain, original magnification × 100).

INTRODUCTION

Focal nodular hyperplasia (FNH) is a benign liver tumor, often asymptomatic and discovered incidentally[1,2]. Although FNH can be found at any age, it is rare in children, and comprises only 2% of all pediatric liver tumors[3]. There have been various reports on the association between FNH and other hepatic lesions, such as hepatocellular adenomas[4], hemangiomas[5], hepatocellular carcinoma[6] and metastases[7]. We herein report a case of FNH associated with hepatoblastoma in a 36-d-old female infant. To the best of our knowledge, only one case of FNH complicated by hepatoblastoma has been reported in a 4-year-old boy after treatment of stage IV neuroblastoma[8].

CASE REPORT

A 36-d-old girl presented to the hospital with a history of jaundice. Physical examination revealed a palpable solid mass fixed on the right quarter of the abdomen approximately 7 cm × 8 cm × 8 cm in diameter. The baby was full-term and delivered normally without asphyxia. Her mother had a history of progesterone administration before and in the early stages of pregnancy.

Sonography revealed a huge uneven hypo-echoic mass in the right lobe of the liver. Computed tomography (CT) imaging showed an ill-delineated, inhomogeneous enhanced mass with a central star-like scar in right lobe of the liver (Figure 1A, B). The tumor showed early mild enhancement at the arterial phase (from 40HU without contrast to 52HU at the arterial phase), intense enhancement at the portal phase (87.7HU) and 98.1HU in the 3-min delay scan, with slight enhancement of the central scar.

Laboratory evaluations included serum total bilirubin (TBIL) 93.1 μmol/L (normal 5.1-17.1 μmol/L), direct bilirubin (DBIL) 42.6 μmol/L (normal 0-6 μmol/L) and an alpha-fetoprotein (AFP) level of 74390 ng/mL (normal 0-77 ng/mL). Serum markers for both hepatitis B and hepatitis C virus were negative.

The infant underwent surgical resection of the tumor. The histopathology demonstrated FNH coexistent with hepatoblastoma (Figure 1C and D).

DISCUSSION

Up to 75% of pediatric liver tumors are malignant[9], and FNH constitutes 5% of benign liver tumors in children[10]. Although FNH is found at any age, even in newborn[11,12], it is rare in children, and two age peaks have been described, around 6 years and 18 years[10].

The pathogenesis of FNH is not well understood[2,13-15]. The most widely accepted theory is that FNH is the result of congenital or acquired vascular abnormalities[14,15]. Numerous studies have reported an increased incidence of

FNH in long-term survivors of childhood malignancies or following hematopoietic stem cell transplantation[8,14-17]. It was thought that FNH was a complication of chemotherapy or radiation therapy in these patients[15].

The features of FNH on MRI and CT include homogeneity, arterial phase enhancement suggestive of hypervascularity, a lack of lesion capsule, and the presence of a central scar[15,18]. Ultrasound, CT, and especially MRI have proven effective at distinguishing FNH from other benign and malignant lesions in adults. In children, due to its infrequency and more variable imaging features, the accuracy of radiologic diagnosis of FNH is challenging. Atypical or variable radiologic features such as absence or non-enhancement of the central scar may be more common in children than adults[9]. Atypical features make it difficult to differentiate FNH from other benign or malignant lesions, including hepatic adenoma, hemangioma or infantile hemangioendothelioma, hepatoblastoma, and hepatocellular carcinoma.

Although there is no evidence in the literature of malignant degeneration of FNH, there are several reports of FNH occurring simultaneously with malignant neoplasms[6-9]. Gutweiler *et al*[8] described a case of hepatoblastoma concomitant with FNH after treatment of neuroblastoma in a 4-year-old boy. Similarly, Lautz *et al*[9] described a patient with hepatocellular carcinoma and concomitant FNH with a history of neuroblastoma. At present, most children with FNH should continue to undergo surgical resection due to symptoms, increasing size, or inability to confidently rule out malignancies[9].

COMMENTS

Case characteristics

A 36-d-old girl presented to the hospital with a history of jaundice.

Clinical diagnosis

Physical examination revealed a palpable solid mass fixed on the right quarter of the abdomen.

Differential diagnosis

Hepatoblastoma, hemangioma or infantile hemangioendothelioma.

Laboratory diagnosis

Serum level of total bilirubin was 93.1 umol/L, direct bilirubin was 42.6 μmol/L, and alpha-fetoprotein level was 74390 ng/mL.

Imaging diagnosis

Sonography and computed tomography (CT) imaging revealed a mass in the right lobe of the liver. The tumor showed early mild enhancement at the arterial phase, intense enhancement at the portal phase and slight enhancement of the central scar in the 3-min delay scan.

Pathological diagnosis

Histopathology demonstrated typical focal nodular hyperplasia (FNH) coexistent with a focal hepatoblastoma.

Treatment

The infant underwent surgical resection of the tumor.

Related reports

There are several reports of FNH occurring in children after treatment of malignant neoplasms. However, no cases of FNH and hepatoblastoma have been reported in infants.

Experiences and lessons

Hepatoblastoma is more common in infants than FNH. The tumor in this infant showed a typical central scar on CT imaging, and histopathology confirmed FNH coexistent with a focal hepatoblastoma.

Peer review

FNH coexistent with a focal hepatoblastoma is rare in infants. The dynamic CT images in this infant are helpful to diagnose FNH in the liver, but it is challenging to confirm a coexistent focal hepatoblastoma.

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P- Reviewer: Chen Y, Otte JB, Tang KF **S- Editor**: Ma YJ
L- Editor: A **E- Editor**: Zhang DN

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ISSN 1007-9327