CASE REPORT – OPEN ACCESS

International Journal of Surgery Case Reports 29 (2016) 76–79

Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.casereports.com

Huge focal nodular hyperplasia presenting in a 6-year-old child: A case presentation

Lin Zhuang[a,1], Chuangye Ni[b,1], Wenbing Din[b], Feng Zhang[b], Yi Zhuang[c], Yawei Sun[a], Dong Xi[a,*]

[a] Department of General Surgery, Wujin Affiliated Hospital of Jiangsu University, Changzhou, 213161, China
[b] Department of Liver Surgery, First Affiliated Hospital of Nanjing Medical University, Nanjing, 210029, China
[c] Department of Emergency, Wujin Affiliated Hospital of Jiangsu University, Changzhou, 213161, China

ARTICLE INFO

Article history:
Received 21 September 2016
Received in revised form 23 October 2016
Accepted 23 October 2016
Available online 27 October 2016

Keywords:
Focal nodular hyperplasia
Child
Benign liver tumors
Computed tomography

ABSTRACT

INTRODUCTION: Focal nodular hyperplasia (FNH) is a benign lesion of the liver which is usually found in healthy adults, however, FNH is rare in children, and comprises only 2% of all pediatric liver tumors. Herein, we report the case of a 6-year-old child (male) with a huge FNH which size is more than 10 cm. This could be the biggest FNH among all children's FNH cases ever reported.
CASE PRESENTATION: A 6-year-old boy was found a hepatic space-occupying lesion two years ago. As the time went by, the lesion became bigger gradually. The last CT examination showed the size of the tumor to be 10.5 × 9.9 cm in the right hepatic lobe. This child underwent surgical resection of the tumor which was confirmed as FNH (11 × 8 × 7 cm) by pathology.
CONCLUSION: FNH is a benign lesion of the liver, and it is characterized by hepatocyte hyperplasia and a central stellate scar. It is uncommon for FNH to be diagnosed in children. Such huge FNH (about 11 cm) is extremely rare. Surgical operation may be the effective method to cure huge FNH.

1. Introduction

Primary tumors of the liver, including malignant and benign tumors, constitute 1%–2% of all pediatric tumors [1]. Focal nodular hyperplasia (FNH) is a benign lesion of the liver which is usually found in healthy adults [2]; however, FNH is rare in children, and comprises only 2% of all pediatric liver tumors [3]. FNH is a well-circumscribed and lobulated tumor, which is often asymptomatic and incidentally discovered [4]. On gross examination, the typical architecture of FNH includes bile ducts and a central stellate scar, which also contains blood vessels that supply the hyperplastic process. Microscopically, the proliferating cells are almost identical to the surrounding hepatocytes [5]. Chen et al. [6] showed children with an indefinite diagnosis should undergo surgical treatment. Herein, we managed a 6-year-old boy with a huge FNH. The diagnosis could not be established based on imaging studies and needle biopsy, thus the boy underwent a surgical procedure.

2. Case report

A healthy 6-year-old boy was referred to us for evaluation of a hepatic space-occupying lesion, which was first detected approximately 2 years ago. Imaging studies, including abdominal ultrasound and computed tomography (CT), were performed 2 years ago. An abdominal ultrasound scan revealed a 6.1 × 5.2 cm lesion with inhomogeneous and a well-demarcated low echo in the right lobe of the liver. On CT image of the upper abdomen, a plain scan showed a 6.5 × 5.4 cm circular shadow with a slightly low density in the right hepatic lobe, which displayed a central star-like scar in the low-density area. On the contrast CT scan, the shadow was more prominent in the arterial phase and decreased slightly in the venous phase. In the delayed phase, the density was close to the liver parenchyma and the lower-density area disappeared in the right hepatic lobe. The tentative diagnoses were FNH or hepatoblastoma based on imaging. The patient was referred to our hospital for further therapy. He had no symptoms of nausea or vomiting. The physical examination showed mild hepatomegaly. Laboratory testing in our hospital revealed the following: alanine aminotransferase (ALT) and aspartate aminotransferase (AST) were 23.2 U/L (normal, 15–40 U/L) and 37.0 U/L (normal, 9–50 U/L), respectively; viral serologic tests for hepatitis B and C were negative; and the alpha-fetoprotein (AFP) level was 0.7 ng/ml.

During hospitalization, the re-examination showed that the lesion had enlarged slightly. The patient had CT in our hospital,

Abbreviations: FNH, focal nodular hyperplasia; ALT, alanine aminotransferase; AST, aspartate aminotransferase; CT, computed tomography.

* Corresponding author.
E-mail address: xi-dong76@hotmail.com (D. Xi).
[1] These authors contributed equally to this work.

http://dx.doi.org/10.1016/j.ijscr.2016.10.053

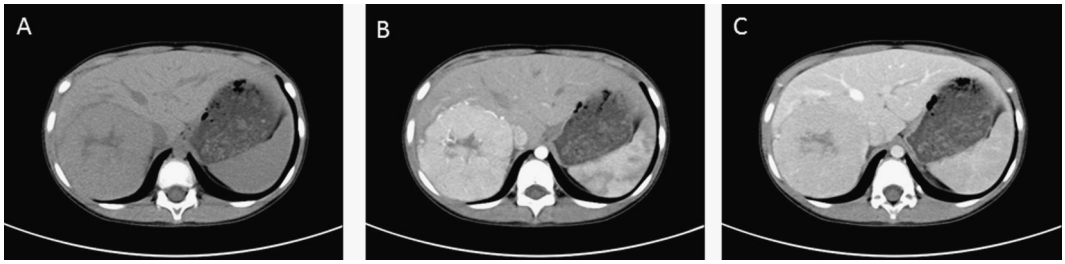

Fig. 1. (A) Plain scan showed the lesion was low density with a lower density scar showed in the center. (B) In the arterial phase, the lesion was significantly enhanced without central scar tissues and showed a clear capsule. (C) The lesion was decreased in the venous phase.

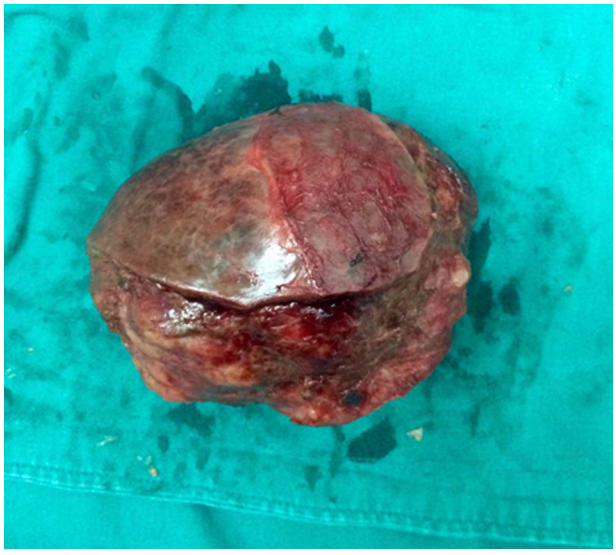

Fig. 2. Gross pathology of the resected tumor shows an 11-cm tumor with a round-like appearance, compatible with the imaging features.

which showed the size of the shadow to be 10.5 × 9.9 cm in the right hepatic lobe and the spleen was slightly larger. A dynamic CT scan of the liver disclosed a lobulated tumor with several expanded feeding vessels in the right hepatic lobe. On arterial and portal venous phase imaging, the tumor continued to exhibit a brightly enhanced early phase and early washout (Fig. 1). Next, we invited a pediatrician to perform the needle biopsy in our hospital; however, the diagnosis was still indeterminate after the histologic examination. Thus, we decided to intervene surgically to obtain a definite pathologic diagnosis. During surgery, a well-defined mass was palpated in the right posterior lobe of the liver. The texture of the mass was hard, and the diameter was approximately 7 cm. Combined with the pre-operative imaging examination, the intra-operative diagnosis was FNH. Thus, we decided to perform a partial hepatectomy.

On the gross pathologic examination of the surgical specimen, the tumor was shown to be enlarged in the right posterior liver, well-defined, yellow-to-tan in color, and 11 × 8 × 7 cm in size, with numerous small nodes on the surface of the liver tumor (Fig. 2). Microscopically, we observed a huge non-encapsulated tumor (10.5 × 8 × 6.5 cm), which consisted of a proliferation of bile ducts and a central stellate scar containing tortuous blood vessels. The pathologic findings were consistent with FNH (Fig. 3). Post-operatively, the child was recovering well at the 6-month follow-up visit.

3. Discussion

Among pediatric primary liver neoplasms, 57% are malignant and 43% are benign [5]. FNH belongs to benign in children with liver diseases. and FNH is usually diagnosed between 2 and 5 years of age [7]. Nevertheless, case reports involving children >5 years of age are still rare worldwide.

FNH is an uncommon diagnosis in children, and is often found incidentally [8–12]. The etiology of these lesions is unclear, with possibilities including vascular or environmental factors [10,13,14]. Like other benign liver tumors, small lesions can be asymptomatic incidental findings. Larger lesions eventually present with numerous symptoms, the most frequent of which is abdominal pain. On examination, the child usually presents with a right upper abdominal mass [15]. Although normal levels of AFP suggest a benign lesion, laboratory tests are usually unremarkable [16]. In this case report, the 6-year-old boy had no obvious abdominal symptoms when he came to our hospital and the physical examination just showed mild hepatomegaly and the tumor-marker test of AFP is normal (0.7 ng/ml).

FNH of the liver, which is a nodular, non-encapsulated, occasionally multifocal (15%-20%) liver mass, most frequently occurs in the left liver lobe [17]. Abdominal ultrasound is often used as the initial diagnostic imaging modality. Ultrasound is non-specific and FNH typically appears as a homogeneous, well-circumscribed lesion that can be isoechoic, hypoechoic, or hyperechoic [11]. The typical central scar is slightly hyperechoic, but is often difficult to visualize on ultrasound (20% of cases) [18]. Ungermann et al. [19] showed that contrast-enhanced ultrasonography can be the final diagnostic method for FNH >3 cm in size with a typical spoke-wheel vessel structure. If this phenomenon is not present and the central scar is not visible, a specific diagnosis of FNH cannot be based solely on contrast-enhanced ultrasound findings. The tumor has characteristics which are more specific and can aid in the diagnosis on CT scan after intravenous contrast enhancement. On pre-contrast CT scans, FNH is often seen as s focal hypo- or iso-dense mass. Some statistics have shown that a central hypodense scar is observed in only one-third of cases [18]. FNH typically demonstrates uniform enhancement with IV contrast administration and enhances more than normal liver tissue. In most cases (89%–100%), the lesion enhances rapidly during the arterial phase of contrast-enhanced CT because of the prominent arterial supply to FNH [20]. In our case, the typical central scar was not noted on ultrasound imaging which was taken two years ago; however, the central scar was seen on CT. Nevertheless, the diagnosis of CT was uncertain, which was compatible with the pathologic results.

Thus far, no histologic reports have shown that FNH undergoes malignant progression; however, cases of FNH lesions associated with hepatocellular carcinoma (HCC) do exist [21–23]. Petsas et al. [23] reported a case of HCC arising within a large FNH. The patient underwent pre-operative CT-guided core needle biopsy that revealed FNH; however, the final pathologic diagnosis disclosed FNH and HCC. Based on this case, we understand that although radiographic imaging could have a particular finding, liver biopsy or resection might be necessary to establish the diagnosis effectively. Makhlouf et al. [24] reported that it is necessary to have