Focal nodular hyperplasia (FNH) of the liver is uncommon in adults and rare in children. It is a benign and usually asymptomatic lesion. The hepatic tumor is often found incidentally on routine physical examination. Establishing the diagnosis of FNH is difficult because there are no universally accepted diagnostic imaging criteria [1, 2]. We present a case of FNH of the liver together with a review of the literature and discussion of the etiology, clinical features, diagnosis, and treatment of this disease.

Case Report

A 7-year-old girl was referred to National Taiwan University Hospital for further evaluation of an asymptomatic abdominal mass found on a routine physical examination at school. The non-tender mass was palpable at the right lower margin of the liver with an extension to 10 cm below the right costal margin. There was no maternal history of exposure to exogenous steroids during pregnancy. The results of all laboratory studies were normal, and the serum α-fetoprotein concentration was less than 20 ng/mL. Hepatitis B surface antigen (HBsAg) and anti-hepatitis C virus (HCV) antibody were negative. Sonography demonstrated a heterogeneous hepatic tumor about 10 cm in diameter, which was more echogenic than the normal liver. Magnetic resonance (MR) imaging showed a large heterogeneous hepatic tumor in the right lobe with iso-signal intensity on T1- and T2-phase images, and mild enhancement after gadolinium diethylenetriaminepentaacetic acid (Gd-DTPA) injection. The inferior vena cava and first branches of the portal vein were compressed to the left and upwards (Fig. 1).

A malignant hepatic tumor was suspected and surgical intervention was arranged. On laparotomy, a circumscribed, hypervascular mass with a nodular surface was found in the right hepatic lobe near the gallbladder (Fig. 2). The rest of the hepatic surface was smooth. No other lesion was detected in the abdominal cavity. The brownish tumor was totally removed, along with the gallbladder.

Pathologic examination revealed a 10 x 9 x 5-cm mass with a central area of collagenous scarring that had fibrous septa radiating from it (Fig. 3). Microscopically, the fibrous septa contained dilated vessels and proliferating bile ductules. In various areas, lymphocytes were abundant. The proliferating hepatocytes did not show any nuclear abnormality. The gross and microscopic appearance of the lesion was suggestive of FNH. The postoperative course was smooth except for fluid accumulation in the subhepatic area, detected on the 16th postoperative day. Percutaneous computed tomography (CT)-guided drainage was performed, and the patient was discharged on the 21st day. She remained well 2 years after surgery.
Focal Nodular Hyperplasia in Children

**Fig. 1.** Magnetic resonance imaging shows a heterogeneous hepatic tumor (arrow) in the right lobe with iso-signal intensity on T1- and T2-phase images, and mild enhancement after gadolinium diethylenetriamine pentaacetic acid injection. The inferior vena cava and first branches of the portal vein are compressed to the left and upwards.

**Discussion**

Primary hepatic tumors are relatively rare during childhood, with a reported incidence of 0.4 to 1.9 per million children each year [3, 4]. FNH represents less than 2% of pediatric hepatic tumors [4], and occurs in all age groups, but it is found most commonly in women (80%–95% of cases) in the third and fourth decades of life [5, 6]. Approximately 15% of cases occur in children (0–16 years). The majority of patients are asymptomatic (50%–80% of cases) [6, 7]. The diagnosis is often made incidentally during a physical examination, or fortuitously during an operation, an autopsy, or radiologic investigation. Symptomatic patients often complain of an abdominal mass or abdominal pain [1, 6].

FNH is a benign and well-circumscribed hepatic tumor. Its gross features typically consist of a central stellate scar with radiating septa. The stellate scar and all the radiating septa contain blood vessels, bile ducts, and lymphocytes. Microscopically, the proliferating cells are practically identical to the surrounding hepatocytes [2, 8]. Because of the excellent blood supply, hemorrhage, necrosis, and infarction are extremely unusual. FNH lesions vary in size, and are usually less than 5 cm in diameter [2]. Lesions may be single or multiple. Histologically proven FNH presenting with a malignant evolution has not been reported. However, cases of FNH lesions have been reported in association with hepatocellular carcinoma [9]. FNH can also occur in association with other hepatic lesions, such as hepatic adenoma [10]. Similar to FNH, partial nodular transformation (PNT) is also a focal nodular benign lesion of the liver. However, PNT is mainly localized to the parenchyma surrounding the hepatic hilum, and can cause noncirrhotic portal hypertension [11]. Our patient had a single and unusually large tumor (10 cm in diameter) in the right lobe of the liver. No portal hypertension was found in the preoperative examination or during the surgical intervention. The pathologic findings showed typical features of FNH and no other associated hepatic lesions were found.

The exact etiology of FNH is unknown. FNH probably represents a local hyperplastic response of the hepatic parenchyma to a preexisting vascular anomaly [12], although there is a strong association between the use of oral contraceptives and hepatic adenoma. The association of FNH with the contraceptive pill remains controversial; it has been reported in 50% to

**Fig. 2.** A circumscribed, hypervascular mass with a nodular surface (arrow) is present in the right hepatic lobe near the gallbladder.

**Fig. 3.** Pathologic examination reveals a central area of collagenous scarring (arrow) that has fibrous septa radiating from it (hematoxylin & eosin, x 40).
75% of women [8, 13]. In addition, when FNH is associated with oral contraceptives, the lesions tend to be larger and symptoms are more common [13, 14]. A possible tumorigenic effect of oral contraceptives is supported by the clinical observation that FNH regresses in patients who discontinue the pill [15]. However, some authors suggest that FNH is not caused by or even associated with the use of oral contraceptives [2, 16]. No oral contraceptives had ever been used by our patient or her mother.

Establishing the diagnosis of FNH is difficult because there are no universally accepted diagnostic imaging criteria. The presence of a homogeneous tumor with a central scar on ultrasonography, CT, or MR imaging may suggest the diagnosis of FNH [2]. Normal or increased uptake on a technetium (Tc)-99m sulfur colloid scan is extremely helpful in supporting the diagnosis of FNH. However, only half of all cases of FNH demonstrate uptake of Tc-99m sulfur colloid scan [2]. The benign nature of such hepatic tumors can be certified only by an open liver biopsy or resection of the tumor. In our patient, FNH was suspected but not diagnosed preoperatively because there was no obvious central tumor scar on ultrasound or MR imaging, and because of the rarity of FNH in children. Radionuclide scan was not performed because FNH was not the major suspected diagnosis and a cold lesion on sulfur colloid imaging could represent any space-occupying lesion within the liver, including FNH [2]. Because of the possibility of malignancy of such a large hepatic tumor, we performed hepatectomy to completely remove the tumor. Although subhepatic effusion complicated the postoperative course, it was resolved by percutaneous CT-guided drainage.

The correct treatment for patients with FNH remains debatable. The management possibilities for FNH include conservative follow-up or excision of the tumor [17]. Vascular intervention (embolization or ligation of the hepatic artery) has also been reported to be an option [18]. Recently, Reymond et al reviewed the follow-up data and outcomes of 31 pediatric case reports of FNH, and found that the outcome appears to be good for both those with observation after operative liver biopsy and those with resection [1]. Whichever method is chosen, regular follow-up with ultrasound or radiologic images is mandatory.

References