

Pedunculated focal nodular hyperplasia

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Case report

A 12-year-old female was admitted to the Cardiothoracic Surgery Unit for surgical correction of tetralogy of Fallot. On routine pre-operative examination a large, firm, freely mobile intra-abdominal mass was detected, of which the patient was unaware.

An abdominal US revealed a large, well-defined, lobulated mass of heterogenous echogenicity, with central hypoechoic branching linear areas and a small speck of calcification. The mass was continuous with the inferior edge of medial segment of left lobe of liver through a narrow pedicle, with no surrounding liver parenchyma. The pedicle contained dilated vascular channels (Fig. 1). Color Doppler examination revealed these vascular channels to be a branch of left hepatic artery, and a vein draining directly into the left hepatic vein. These vessels could be traced into the centre of the mass, from where branches radiating to the periphery of the mass could be seen. Rest of the hepatic parenchyma and other intra-abdominal organs were normal.

Contrast enhanced CT scan of the abdomen revealed a large well-defined, lobulated, enhancing mass with central hypodense branching areas containing enhancing blood vessels and calcification (Fig. 2). Communication between the inferior edge of the left lobe of liver and mass through a pedicle containing dilated tortuous vascular channels could be made out.

On the basis of these radiological findings a diagnosis of pedunculated liver tumor was made. Further, characteristic features, viz; a central branching area (scar) containing blood vessels and calcification, helped us make a diagnosis of pedunculated focal nodular hyperplasia (FNH).

All radiological findings were confirmed at surgery – the pedicle was ligated and the mass excised in-toto. Histopathology was confirmatory for FNH. Interestingly, after excision of the mass, an appreciable decrease was noted in the degree of the patient's cyanosis.

Discussion

FNH is classically described in middle-aged females and is usually asymptomatic. The mass is usually small (average 4 cm diameter; maximum reported size 20 cm). A central scar containing fibrous tissue, vascular channels and calcification is often present (60%) and is pathognomonic [1].

Sonography is sensitive for detection of FNH, but the echo-pattern of these tumors is variable and non-specific – thus differentiation with other benign and malignant liver tumors is not possible on the basis of US alone [1]. Our experience suggests that a combination of Color Doppler and 2D US imaging can help make a specific diagnosis by demonstrating the characteristic pattern of vascular supply, with major feeder blood vessels and their branches contained within the central hypoechoic scar.

CT appearances of FNH are non-specific with a variable attenuation pattern on both non-contrast and contrast enhanced scans, though a hypodense lesion on precontrast scan which becomes

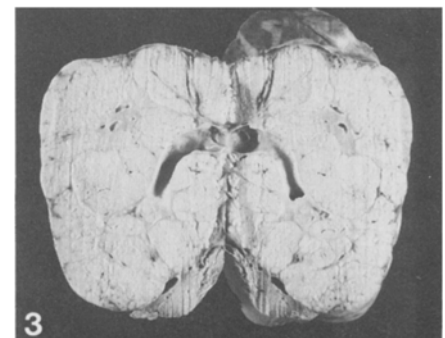
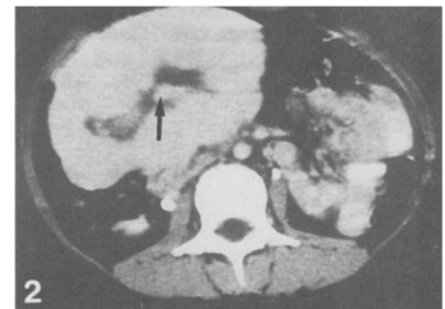
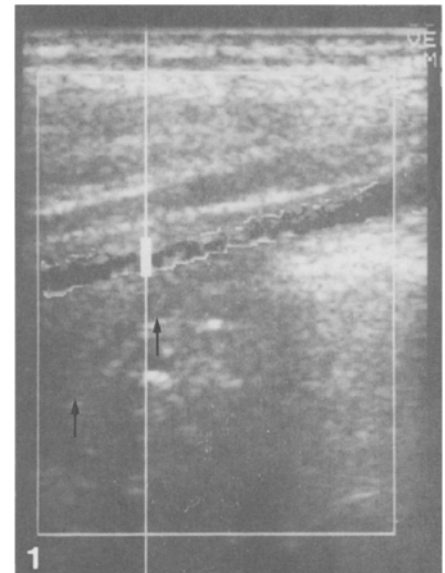


Fig. 1. Longitudinal right paramedian scan. Pedicle is seen containing dilated blood vessel (Cursor in vessel draining into left hepatic vein) and connecting left lobe of liver (arrows) to mass (not seen in scan)

Fig. 2. Enhanced CT scan at L3 vertebral level. Homogeneous well defined encapsulated mass with central branching hypodense scar. Blood vessels (arrow) seen within scar

Fig. 3. Cut surface of excised mass. The mass is divided by broad fibrous trabecular strands into lobules of varying sizes. The fibrous septae are converging towards a central area of fibrosis which contains large blood vessels

hyperdense with IV contrast is typical [1].

We have not come across any previous reference of a radiologically documented case of pedunculated FNH, though in a large series of pediatric patients, there was a mention of two patients with pedunculated FNH [2]. Pedunculated hepatocellular carcinomas have been reported in 0.3% to 2.4% of all patients with hepatocellular carcinoma [3]. Many explanations have been postulated for the pedunculated appearance of hepatic masses, e. g., masses arising in congenitally displaced hepatic lobules in Glisson's capsule, ectopic liver tissue, accessory lobes, etc. [3].

Since FNH usually arises superficially, just under the capsule of liver and from

the inferior edge, it is not surprising that a pedunculated mass can result, though it is rare.

Also, an association with a congenital cardiac anomaly (tetralogy of Fallot in this patient) has not been reported previously, though a coincidental co-existence is possible. The decrease in cyanosis following excision of the mass may be explained on the basis of a hypervascular tumor with a large amount of oxygen extraction.

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