

## Focal nodular hyperplasia (FNH) of liver in children

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### Abstract

**Introduction:** Focal nodular hyperplasia (FNH) is one of the rare benign tumors of liver and second common benign tumour after the haemangioma in adult<sup>1</sup>. It is mostly seen in young middle age women .The aetiology of this tumour is unknown; however congenital vascular malformation or vascular injury to the hepatocytes has been suggested as the underlying mechanism for hepatocellular hyperplasia<sup>2,3</sup>. Its relation with OCP is still controversial<sup>2</sup>.

The tumor is asymptomatic most of the time and detected during routine physical examination or incidentally during radiological investigation. Large lesion presented with abdominal mass and abdominal pain due to pressure effects over surrounding tissue. Only Symptomatic lesion needs treatment. Bleeding and torsion can occur but malignant transformation never occur in this disease<sup>4</sup>.

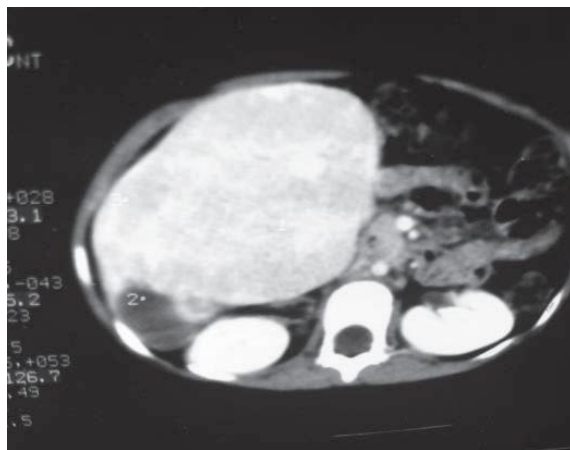
FNH is very rare in pediatric population.

Here we report two case of such rare case in pediatric population. Hepatic resection was undertaken in both patients with excellent outcome.

### Case 1

A 6-year-old girl was admitted for evaluation of abdominal pain and palpable abdominal lump. During examination liver was palpable 3 cm below the costal margin. Her LFT and coagulation profiles, tumor markers ( $\alpha$ -fetoprotein and  $\beta$ -HCG) were within normal limit. USG of abdomen shows heterogenous mass occupying both lobes of liver suggestive of hepatoblastoma. This sonographic finding was further confirmed by contrast enhanced CT scan of the abdomen which also revealed malignant liver mass suggestive of hepatoblastoma occupying IVB and V segment of liver (fig 1) . Non anatomical resection of liver mass was done with 1 cm clear margin. Mass was about 12\*10\*8 cm lobulated, pedunculated and arising from segment IV and V of liver.

Histopathology revealed focal nodular hyperplasia atypical type.

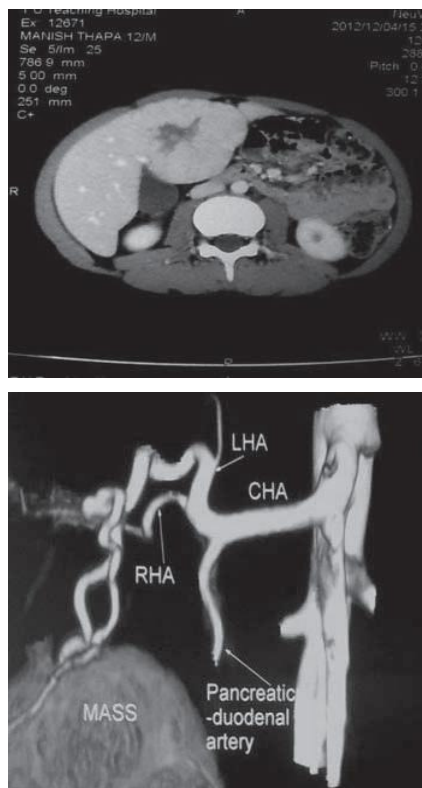


**Figure 1:CECT abdomen shows heterogenous mass occupying segment IVB and V of liver**

## Case 2

A 12-year-old boy was presented with on and off right upper abdominal pain and on examination liver was palpable 5 cm below the costal margin, otherwise patients was normal. Patient was admitted for evaluation of liver mass. His liver function tests were within normal limit except slightly raised alkaline phosphatase. Viral serologic tests for hepatitis B, and C were negative. Abdominal ultrasound showed heterogenous mass occupying segment IV B of liver suggestive of hepatoblastoma with differential diagnosis of hepatocellular carcinoma. In contrast to USG, contrast enhanced CT scan showed large well-circumscribed homogeneously enhancing mass that was isodense to liver with classic low-attenuation central scar and large feeding artery. Non anatomical resection of segment IV B was done. The gross appearance was that of lobulated, well circumscribed masses, showing a central scar with fibrous septa running to the periphery and partially demarcating nodular structures

Histopathology revealed focal nodular hyperplasia: Typical type.



**Figure 2: CECT showing liver mass with central scar, and feeder artery**

## Discussion

FNH is the second most common benign tumor of the liver and characterized by abnormally organized normal hepatocytes in the form of nodular mass, with a fibrous septa that extend out from a central scar<sup>5</sup>.

The abnormally organized normal liver cells is due to congenital vascular anomalies, result in an increased arterial flow to the hepatocytes. Some hepatotoxic drug such as busulphan, malfalan and radiotherapy are considered as the risk factors for FNH<sup>6</sup>. Smoking also plays an important role in pathogenesis of FNH<sup>7</sup>. The arterial supply in FNH is from the hepatic artery and the venous drainage is into the hepatic veins. FNH does not contain portal venous supply.<sup>8</sup>

Sometimes the disease is associated with other vascular anomalies, malformation, and agenesis of the portal vein, hepatic vein, and capillaries. Studies suggest that angiopoietin-1 (Ang-1) gene and angiopoietin-2 (Ang-2) are involved in angiogenesis in FNH with increase in Ang-1/Ang-2 ratio as compared to normal liver<sup>9</sup>. Sometime chromosomal abnormality is also associated with focal nodular hyperplasia<sup>10</sup>. Solitary occurrence is more common<sup>11</sup>. These lesions are either found incidentally during physical examination or during routine imaging. Twenty to fifty percentages of the patients presented with right upper quadrant pain which is due to pressure effect<sup>12</sup>. FNH can complicate by spontaneous rupture and haemorrhage, torsion, but these complications are rare. It does not undergo malignant transformation<sup>4</sup>.

FNH are of two types, typical and atypical. Typical lesions characterised by a tumour with well defined margins, prominent central scar with radiating fibrous septae. A large central artery is usually present with centrifugal flow. In contrast to typical FNH, atypical FNH lacks the central scar and central artery, and is thus difficult to distinguish it from other lesions on gross inspection and imaging. Lesion are heterogenous and pseudocapsule is present. Central scar is present but does not enhance on imaging. Nodules can grow and disappear and new nodules can appear even after resection.

Here one of the cases was typical and other case was atypical.

Some authors also describe the atypical FNH into telangiectatic, mixed hyperplastic and adenomatous, large cell hepatocellular atypia variant.

For the optimal evaluation of FNH, a helical CT scan with a 4-phase study should be performed. This evaluation should include nonenhanced, arterial, portal venous and delayed phase examinations.

During the arterial phase, FNH lesion shows an intense enhancement, where as central scar shows delayed enhancement. This is due to presence of abundant myxomatous stroma in the scar. During the portal venous phase there is decreased enhancement of the lesion, resulting in the lesion being isoattenuating to the liver, with gradual diffusion of the contrast material into the central scar in delayed phase. Twenty percents of the scar demonstrates enhancement on delayed phase.<sup>13</sup> Contrast enhanced MRI is equally effective as contrast enhanced CT scan for diagnosing FNH. Technetium (Tc) 99m sulphur colloid scan positive is seen in 80% of lesions, and is helpful in distinguishing them from other hepatic neoplasm.<sup>13</sup>

As Treatment is not required for asymptomatic FNH so, accurate diagnosis is essential in preventing unnecessary intervention and treatment. However, surgery is needed if the lesions is symptomatic, increasing in size or sign of intralesional hemorrhage on investigation.

## Conclusion

FNH is rare benign tumor in children, with no malignant potential and very small risk of complication (rupture, haemorrhage) and thus are usually treated conservatively. However Symptomatic lesion needs surgical intervention.

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