Case Report:

Focal Nodular Hyperplasia in children presenting as acute Cholecystitis

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Abstract:

In this case report, a female child of 10 years old presented with pain epigastrium and with sonographic features of acute cholecystitis. Later on, the focal mass in the liver was diagnosed by Magnetic Resonance Imaging (MRI) examination and proved by the biopsy as focal nodular hyperplasia (FNH), a benign focal response of the liver and a hepatic mass. Although focal nodular hyperplasia is rare mass in children however, Computed Tomography (CT) scan / Magnetic Resonance Imaging (MRI) examination are essential for the diagnosis and can obviate the need for the biopsy and unnecessary surgery.

Key words: Focal nodular hyperplasia, ultrasound, MRI.

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Introduction:

The focal nodular hyperplasia (FNH) was first described by Edmondson in 1956. This is the second common benign tumour after the haemangioma and accounts for 8% in adults and 2% in children (1). The pathogenesis is uncertain, although its association with oral contraceptives in females and chemotherapy / radiotherapy has been described in the literature. Mostly it is solitary and measure 5 cm in diameter, however large sizes up to 15 cm has been reported. Grossly, FNH is described as lobulated, non-capsulated benign hepatic lesion with a central star-like scar (2). It is asymptomatic most of the time and detected incidentally by imaging or at autopsy. However, very large masses presents with abdominal pain due to pressure effects.

Case presentation:

A 10 year old girl, presented to the emergency with sudden onset of abdominal pain, mainly in the epigastric region without history of vomiting or abdominal distension. On physical examination, mild hepatomegaly was present with normal vitals. The liver function and viral serology tests were unremarkable. Ultrasound (US) abdomen revealed a well defined subcapsular hyperechoic mass in the right lobe of liver, measuring 6.48 x 7.44 cm. (Fig.1) with evidence of acute cholecystitis. A diagnosis of haemangioma with cholecystitis was made by US examination. After a few days, epigastric pain subsided with antibiotic course and further US examination showed normal gall bladder with persistent hepatic mass. Further characterization of the mass with MRI examination revealed a solitary subcapsular mass in the right lobe of liver, measuring 6.5 x 7.5 cm with a central stellate scar. The mass was iso-intense on T2W images and displayed decreased signal on T1w images. The postcontrast images revealed intense enhancement of the mass in arterial phase with delayed enhancement of the central scar (Fig.2 & 3). A diagnosis of focal nodular hyperplasia was made by MRI examination. After few days biopsy of the lesion confirmed the diagnosis. Histological analysis showing hyperplastic parenchyma with a central fibrous scar, containing a proliferation of small bile ducts, thickened vessels.

Discussion:

Focal nodular hyperplasia (FNH) is not a true neoplasm, but it probably represents a local hyperplastic response of hepatocytes to a congenital vascular anomaly (3). FNH contains all the normal constituents of the liver but in an abnormally organized fashion. Focal nodular hyperplasia is a localized, well defined focal lesion within an otherwise normal liver. Liver function tests are usually within normal range.

In most patients FNH is incidentally discovered during US or CT scan examinations as solitary mass of < 5 cm in size. Large mass is rare to occur and may present with abdominal pain and mass. FNH occurs commonly as a solitary lesion, but multiple lesions may occur. Although FNH usually has no clinical significance, recognition of FNH is important to avoid unnecessary surgery, biopsy, and follow-up imaging. Malignant transformation of FNH has not been reported.

The US features of FNH are variable (4). The mass may appear hypo echoic, isochoric or hyper echoic. Central scar is only visible in few cases as hypo dense star like are. The Doppler examination may reveal central vascularise with peripheral spoke wheel patter.

In non-enhanced CT scan, FNH may be isodense or hypo dense and appears hyper dense in contrast enhanced arterial phase, becomes less dense in venous phase and appears isodense to the liver in delayed phase. The central scar is visible in 15-30% as hypo dense in the arterial phase and becomes hyper dense in the delayed phase due to contrast wash out (5).

In majority of cases, FNH appear as iso to hypo-intense in T1 images and hyper-intense on T2 images with variable signal in the central scar. The gadolinium enhanced MR images reveal hyper-intense mass in arterial phase which becomes iso-intense in venous and delayed phases. Late and prolong enhancement of central scar occasionally seen (6).

Although the CT scan and MRI findings are typical in FNH with presence of central scar and typical enhancement patter, however in difficult cases with differential diagnosis of adenoma and carcinoma, biopsy is necessary.

This case presentation of FNH in a female child provides an opportunity to the clinicians and radiologists to gain confidence for the diagnosis of FNH by the characteristic appearance of FNH on imaging and obviating the need for the biopsy and unnecessary surgery.

This case of FNH in this child was associated with the acute cholecystitis. This could be an incidental finding or has some relation with acute cholecystitis. So far no case has been reported in the literature with this correlation. It's yet to prove the correlation between the FNH with the acute cholecystitis. This case presentation can open the new doors for further research in this direction.

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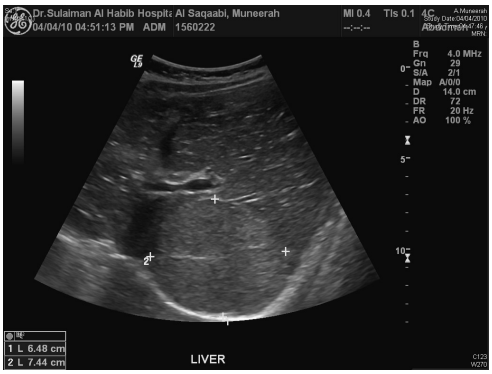

Fig.1: US examination showing a hyperechoic mass in the right lobe of liver.

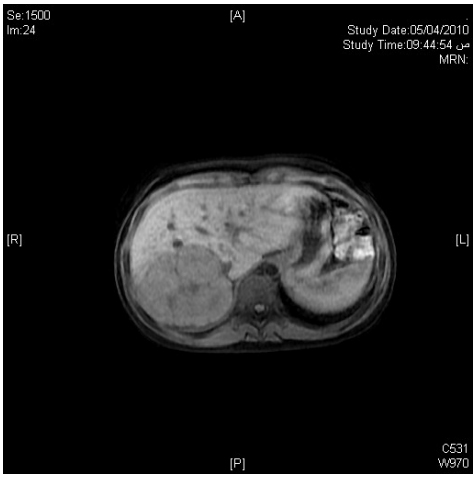

Fig. 2 MRI examination displaying decreased signal in the mass with very low signal of the central stellate scar.

Fig. 3: Post contrast MRI examination showing delayed enhancement of the central scar of the hepatic mass.