Imaging of hepatic hypervascular tumors & clinical implications
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Von Meyenburg complexes mimicking metastatic disease at laparotomy for focal nodular hyperplasia
A 44-year-old woman presented with symptoms of fatigue and increasing abdominal discomfort. MR imaging with the hepatobiliary contrast Gd-EOB-DTPA (Primovist®) was performed showing a 6cm lesion in segment 2/3 of the liver typical for focal nodular hyperplasia. Because of severe complaints attributed to the lesion, the patient was scheduled for resection. At laparotomy multiple small white lesions were found throughout the liver with enlarged loco-regional lymph nodes. Macroscopically, the findings were consistent with widespread metastases and the surgeon felt compelled to determine the nature of these lesions before continuing resection. Final diagnosis revealed multiple bile duct hamartomas and an FNH lesion as expected.

During a routine liver resection for the benign liver tumor FNH, the unexpected findings of multiple small lesions made us doubt the benign nature of the large liver tumor as we feared for widespread metastases throughout the liver. Fortunately the small lesions were in time diagnosed as hamartomas and resection of the FNH lesion could proceed as planned.

PRESENTATION

A 44-year-old woman presented with symptoms of fatigue and increasing abdominal discomfort. Patient history included hypothyroidism (medically treated with Thyrax) and a cholecystectomy. Diagnostic imaging was performed and a 6cm focal nodular hyperplasia (FNH) was diagnosed in segment 2/3 of the liver. Laboratory results showed elevated yGT 228 U/L (normal <40) and alkaline phosphatase 141 U/L (normal 40-120). Tumormarkers were normal: carcinoembryonic antigen 1.6μg (normal <5.5) and alpha-fetoprotein 4μg (normal <7). Additional MR imaging with the hepatobiliary contrast Gd-EOB-DTPA (Primovist®) was performed showing a lobulated hypervascular 6cm lesion in segment 2/3 of the liver (Figure 1A). The hypodense area within the lesion was consistent with a central scar, which is typical for FNH. Furthermore, the hepatobiliary phase 20 minutes post injection of contrast showed accumulation of contrast within the lesion, also consistent with FNH. Hepatocellular adenoma and (fibrolamellar carcinoma) were considered. However, the typical central scar made hepatocellular adenoma less likely. The lesion did not show typical findings for malignancies including ‘wash-out’: loss of contrast during veno-portal phase of dynamic imaging.

TREATMENT

Because of severe complaints attributed to the lesion, the patient was scheduled for resection of segment 2/3. At laparotomy multiple small white lesions (<0.5 cm) were found throughout the liver (Figure 2A) with enlarged loco-regional lymph nodes. Macroscopically, the findings could be consistent with widespread metastases or granulomatous hepatitis and the surgeon felt compelled to first determine the nature of these lesions before continuing resection. Frozen section examinations of liver biopsies revealed bile duct hamartomas, whereas the lymph nodes revealed only reactive changes, without signs of malignancy. Final histopathological diagnoses revealed an FNH (5,5 x 6cm; Figure 2B, Figure 3A), of which the diagnosis was confirmed with an additional GS staining, showing a typical ‘map-like’

**Figure 1**
A: Shows the FNH lesion in segment 2/3 of the liver on the transverse Tw arterial image of a 44-year-old woman (white arrowhead). The hypointense central area within the lesion is consistent with a central scar, which is typical for FNH (Gd-EOB-DTPA, Primovist®). B & C: Show the transverse MR Tw images with multiple small subcapsular hypointense lesions (white arrows). On other series of MR imaging only the largest two were visible and were described as cystic lesions.

**Figure 2**
A: View of the liver at laparotomy showing the tumor (FNH) in the left-lateral segments (2/3) and multiple small greyish white lesions throughout the liver parenchyma mimicking multiple metastases.
B: Cross-sectional image of the lesion in segments 2/3, showing the fibrotic central area, consistent with the central scar seen on pre-operative imaging (arrow).
staining pattern (Figure 3B) [1]. The other lesions consisted of multiple bile duct hamartomas (Von-Meyenburg complexes) in the surrounding liver parenchyma (Figure 4) and reactive changes in the resected lymph nodes. No signs of malignancy, granulomatous hepatitis, tuberculosis, or sarcoidosis were found. After resection, the patient was relieved of her symptoms. However, during follow-up the patient developed a cicatrical hernia, for which she underwent surgical repair.

**DISCUSSION**

FNH are benign liver lesions with no risk of bleeding or malignant transformation [1]. Therefore, FNH are only considered for resection if discomfort perceived by the patient is severe, other possible causes of complaints are excluded, and risk of surgery versus complaints is well evaluated. The exact etiology of FNH is unknown, but one hypothesis is vascular damage resulting in a hyperplastic and fibrotic response. The etiology of Von-Meyenburg complexes, or bile ducts hamartomas, is better known and is thought to be a developmental malformation with persistence of the ductal plate configuration. [2, 3]. The two cystic lesions larger than 5mm were depicted on pre-operative MR imaging and were described as simple cysts (Figure 1). However, like in most cases with hamartomas, the majority of hamartomas in our patient was small and only in retrospect detected as hyperintense irregular lesions on T2w series and low signal intensity on T1w series of the MR (Figure 1). FNH and hamartomas are most likely to co-exist based on pure chance and no causal relation between both entities has been substantiated in literature. However, the co-existence might cause confusion as it mimics metastatic disease.

**LEARNING POINTS & TAKE HOME MESSAGE**

- FNH is a rare, benign liver tumor which may give rise to symptoms, relieved by excision of the lesion.
- Characteristic features of FNH on imaging are a central scar, without signs of wash-out of the lesion on veno-portal phase of dynamic imaging.
- Because bile duct hamartomas are usually small, they are often not detected on cross-sectional imaging studies.
- Bile duct hamartomas may mimick metastatic disease of the liver.
- Bile duct hamartomas may coincide with any other type of liver tumor, benign or malignant. An association of bile duct hamartomas with FNH is not apparent.
Case study references

24 Seo HI, Jo HJ, Sim MS, Kim S. Right trisegmentectomy with thoracoabdominal approach after transarterial embolization for giant hepatic hemangioma. World J. Gastroenterol. 15(27), 3437-3439 (2009).

Case study 4