Imaging of hepatic hypervascular tumors & clinical implications
Bieze, M.

Citation for published version (APA):

General rights
It is not permitted to download or to forward/distribute the text or part of it without the consent of the author(s) and/or copyright holder(s), other than for strictly personal, individual use, unless the work is under an open content license (like Creative Commons).

Disclaimer/Complaints regulations
If you believe that digital publication of certain material infringes any of your rights or (privacy) interests, please let the Library know, stating your reasons. In case of a legitimate complaint, the Library will make the material inaccessible and/or remove it from the website. Please Ask the Library: http://uba.uva.nl/en/contact, or a letter to: Library of the University of Amsterdam, Secretariat, Singel 425, 1012 WP Amsterdam, The Netherlands. You will be contacted as soon as possible.

UvA-DARE is a service provided by the library of the University of Amsterdam (http://dare.uva.nl)
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 could be 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 was expected.

**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' pattern.
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 aetiology of FNH is unknown, but one hypothesis is vascular damage resulting in a hyperplastic and fibrotic response. The aetiology 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 mimic 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.
References

Case study 1

24 Seo HI, Jo HJ, Sim MS, Kim S. Right trisegmentectomy with thoracoabdominal approach after tran
25 Lerner SM, Hiatt JR, Salamandra J et al. Giant cavernous liver hemangiomas: effect of operative ap
26 Singh RK, Kapoor S, Sahni P, Chattopadhyay TK. Giant haemangioma of the liver: is enucleation bet
27 Kayaglu HA, Hazinedaroğlu S, Özkan N, Yerdel MA. Surgical treatment of symptomatic cavernous
28 Belli G, D’Agostino A, Fantini C et al. Surgical treatment of giant liver hemangiomas by enucleation
using an ultrasonically activated device (USAED). Hepatogastroenterology. 57(54), 556-560 (2010).
29 Kuo PC, Lewis WD, Jenkins RL. Treatment of giant hemangiomas of the liver by enucleation. J. Am.
30 Choi J, Lee YJ, Hwang DW, Chon SH, Naggal A, Park KM. Surgical treatment of giant hepatic he
31 Karahasanoglu T, Altinli E, Ergunes Y, Ertem M, Numan F. Laparoscopic enucleation of giant liver
32 Lanthaler M, Freund M, Nehoda H, Laparoscopic resection of a giant liver hemangioma. J. Laparoen

Case study 4

1 Maillette de Buy WL, Terpstra V, Beuers U. Focal nodular hyperplasia and hepatic adenoma: epidemi
3 Redon MS, Waneson JR. The hepatic von Meyenburg complex: prevalence and association with he
4 Bieze M, Bennink RJ, El-Massoudi Y, et al. The use of 18F-fluoromethylcholine PET/CT in differenti
ating hepatocellular adenoma from focal nodular hyperplasia: prospective study of the additional value
5 Bieze M, Busch OR, Tanis PJ, et al. Outcomes of liver resection in hepatocellular adenoma and focal
nodular hyperplasia. HPB (Oxford) 2013 Mar 19 Epub.
cellular adenoma from focal nodular hyperplasia: prospective study of the additional value of gadox