CASE REPORT

Von Meyenburg complexes mimicking metastatic disease at laparotomy for focal nodular hyperplasia

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SUMMARY

A 44-year-old woman presented with symptoms of fatigue and increasing abdominal discomfort. MRI with the hepatobiliary contrast Gd-EOB-DTPA (Primovist) was performed showing a 6 cm lesion in segment 2/3 of the liver typical for focal nodular hyperplasia (FNH). 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 locoregional 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.

BACKGROUND

During a routine liver resection for the benign liver tumour focal nodular hyperplasia (FNH), the unexpected findings of multiple small lesions made us doubt the benign nature of the large liver tumour 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.

CASE PRESENTATION

A 44-year-old woman presented with symptoms of fatigue and increasing abdominal discomfort. Patient history included hypothyroidism (medically treated with thyroxine) and a cholecystectomy.

INVESTIGATIONS

Diagnostic imaging was performed and a 6 cm FNH was diagnosed in segment 2/3 of the liver. Laboratory results showed elevated γ GT 228 U/L (normal <40) and alkaline phosphatase 141 U/L (normal 40–120). Tumour markers were normal: carcinoembryonic antigen 1.6 μ g (normal <5.5) and α -fetoprotein 4 μ g (normal <7). In addition MRI with the hepatobiliary contrast Gd-EOB-DTPA (Primovist) showed a lobulated hypervascular 6 cm 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 min postinjection of contrast showed accumulation of contrast within the lesion, also consistent with FNH.

DIFFERENTIAL DIAGNOSIS

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 venoportal phase of dynamic imaging.

TREATMENT

Because of severe symptoms attributed to the lesion, the patient was scheduled for resection of segment 2/3.

Figure 1 (A) The focal nodular hyperplasia (FNH) lesion in segment 2/3 of the liver on the transverse T1-weighted 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 and C) The transverse MR T2-weighted images with multiple small subcapsular hyperintense lesions (white arrows). On other series of MRI only the largest two were visible and were described as cystic lesions.

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OUTCOME AND FOLLOW-UP

At laparotomy multiple small white lesions (<0.5 cm) were found throughout the liver (figure 2A) with enlarged locoregional 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 (5.5×6 cm; figures 2B and 3A), of which the diagnosis was confirmed with an additional glutamine synthetase staining, showing a typical 'map-like' 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 cicatricial 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 symptoms are excluded, and risk of surgery versus symptoms 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

Figure 2 (A) View of the liver at laparotomy showing the tumour focal nodular hyperplasia 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 preoperative imaging (arrow).

Figure 3 (A) H&E-stained sections (×4) of the lesion in segments 2/3 composed of nodules, formed by monomorphic hepatocytes and separated by fibrotic bands in which solitary dystrophic arteries (black arrows), inflammation and ductular reaction is seen. (B) Immunohistochemical staining (×4) of the lesion for glutamine synthetase, showing a typical 'map-like' pattern consistent with focal nodular hyperplasia.

lesions larger than 5 mm were depicted on preoperative MRI 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

Figure 4 H&E-stained sections (×4) of the whitish lesions found throughout the liver parenchyma showing fibrous stroma in which dilated bile ducts (black arrows) are present, lined by cuboidal epithelium and occasionally filled with bile plugs, typical of hamartomas or 'Von Meyenburg complexes'.

hyperintense irregular lesions on T2-weighted series and low-signal intensity on T1-weighted 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

- ▶ Focal nodular hyperplasia (FNH) is a rare, benign liver tumour 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 venoport phase of dynamic imaging.
- ▶ Because bile duct hamartomas are usually small, they are often not detected on crosssectional imaging studies.
- ▶ Bile duct hamartomas may mimic metastatic disease of the liver.
- ▶ Bile duct hamartomas may coincide with any other type of liver tumour, benign or malignant. An association of bile duct hamartomas with FNH is not apparent.

Contributors MB and TMVG were responsible for patient care, gathering of the data and follow-up of the patient. SSP evaluated imaging and contributed to text and images in the manuscript. JV evaluated histopathology and contributed to text and images in the manuscript.

Competing interests None.

Patient consent Obtained.

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REFERENCES

- 1 Maillette de Buy WL, Terpstra V, Beuers U. Focal nodular hyperplasia and hepatic adenoma: epidemiology and pathology. *Dig Surg* 2010;27:24–31.
- 2 Principe A, Lugaresi ML, Lords RC, *et al*. Bile duct hamartomas: diagnostic problems and treatment. *Hepatogastroenterology* 1997;44:994–7.
- 3 Redston MS, Wanless IR. The hepatic von Meyenburg complex: prevalence and association with hepatic and renal cysts among 2843 autopsies [corrected]. *Mod Pathol* 1996;9:233–7.

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