Focal Nodular Hyperplasia:
A Case Report of Rare Multiple Ruptures of a Common Liver Tumour in a Single Patient

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ABSTRACT
Focal nodular hyperplasia (FNH) is one of the three most common benign solid liver tumours along with hemangiomas and adenomas. FNH is considered a vascular abnormality that usually follows an uneventful course after accidental discovery on CT or MRI for an unrelated medical problem and rarely requires any treatment. These lesions are stable in nature with minimal risk of rupture and essentially no risk for malignant degeneration. The general recommendations for an asymptomatic FNH are observation only, regardless of size of the mass. However, the consequences of a ruptured liver mass can be very serious as abdominal bleeding may be catastrophic, so accurate diagnosis is essential. Here we present the only known case of a patient with multiple FNH nodules and subsequent rupture of two of the lesions; the first treated with a left hepatectomy and the second with embolization. A discussion of the management of the ruptured tumours follows and highlights how little evidence is available for the treatment of multiple ruptures of FNH or for properly risk stratifying patients.

KEYWORDS: focal nodular hyperplasia, ruptured tumor, liver resection and radiofrequency ablation

INTRODUCTION
Benign liver tumours are predominantly found in women with the most common usually being categorized as one of the following: hemangioma, adenoma, or focal nodular hyperplasia (FNH). Out of these three, adenomas are most notable for their risk of rupturing and malignant degeneration. FNH is characterized by its benign course, and generally no treatment is recommended. Hemangiomas follow a similar benign course, and again, observation only is recommended. FNH is the second most common benign solid liver tumour, makes up 8% of all primary hepatic tumours, and is present in up to 3% of the general population. However, spontaneous rupture and subsequent bleeding is very rare.

The pathophysiology of FNH is not well understood although it is thought to be caused by polyclonal hyperplasia of liver cells as a result of locally enhanced blood flow due to vessel malformations. Lesions that typically present are well-circumscribed with a central scar and are noted most often during X-ray computed tomography (CT) on the arterial phase contrast rather than venous, distinguishing FNH from adenoma. It is important to use imaging to diagnose FNH so as not to miss a more serious diagnosis of a potential malignancy. Using magnetic resonance imaging (MRI) after gadolinium administration, the lesions are hyperintense but then become isointense on later images. The MRI will also show the characteristic central scar of FNH more readily than CT and will often also demonstrate sulfur colloid imaging uptake by Kupffer cells, which does not have the sensitivity and specificity to confirm or refute FNH but is usually not seen in malignancy. Unlike adenomas, FNH does not seem to increase in oral contraceptive users but can occur more often in older women. Classically, these lesions remain stable in size, do not rupture, and do not have malignant potential.

CASE REPORT
Here we present a case report of a 37-year-old First Nations woman with multiple FNH lesions who first presented to the Emergency Department (ED) with right upper-quadrant abdominal pain and a vague history of back, flank, and abdominal pain of two months duration. Upon ultrasound, a 4 cm solid lesion was detected in
the left lobe of the liver along with a smaller lesion in the same region. A triphasic CT was performed which showed multiple lesions in her liver seen only in the arterial phase with the largest lesion being 5 cm. The lesions were thought to be consistent with FNH, adenoma, or hepatocellular carcinoma.

This patient had no history of Hepatitis B or C infection or significant alcohol intake. Her past medical history was unremarkable except for a hospitalization for a Caesarean section and a previous laparoscopy for ovarian pain.

At one-month follow-up, CT scanning, MRI, and nuclear medicine imaging were done. The imaging was highly consistent with FNH including arterial phase hypervascularity, the presence of a central scar, and concordance with sulfur colloid uptake indicating the presence of Kupffer cells within the lesions. These features together are considered pathognomonic of FNH. The largest nodule was in liver segment 7/8 and measured 4.9 cm X 3.5 cm. In segment two, another large nodule measuring 3.8 cm X 3.2 cm was present as well as several other smaller scattered lesions. See Figure 1 for a biphasic CT scan showing typical dynamic phase imaging of FNH. The arterial phase image (1A) demonstrates early arterial enhancement and the portal phase image (1B) demonstrates portal venous washout. See Figure 2 for a contrast-enhanced MRI that also demonstrates similar features as the CT scan with arterial enhancement (2A) and portal venous washout (2B).

Further laboratory work was unremarkable and negative for Hepatitis A Virus, Hepatitis B Surface Antigen, and antibodies to Hepatitis C Virus (anti-HCV). Alpha fetoprotein levels were also normal. Her pain was attributed to irritable bowel syndrome.

The patient continued to have right upper-quadrant pain for several more months when she presented with sudden increased abdominal pain. Imaging demonstrated evidence of hemoperitoneum and rupture of one of her liver masses. She was taken to the operating room on urgent basis where a left hepatectomy was performed. Pathology confirmed the diagnosis of FNH with a ruptured 7.5 cm nodule. Her postoperative course was uneventful. Specimens were sent to pathology with the results shown in Figure 3 and Figure 4, both demonstrating the classic pathology for FNH: a central scar and surrounding non-dysplastic hepatocytes. A subsequent abdominal sonogram was done that showed three new lesions in the right lobe of the liver with the rest of the exam unchanged.

At her follow-up appointment six months later, she was still complaining of intermittent aches and pains. Discussions ensued regarding the utility of prophylactic embolization or ablation of her remaining lesions. This was not recommended due to lack of evidence and follow-up imaging in six months was the recommended course of action.

The patient remained stable for another six months when she again presented to the ED for recurrence of right upper-quadrant
nodoses. Complications related to FNH resulting in ruptured FNH, and the invasive nature of these treatments as preventative measures. The consequences of a ruptured FNH nodule can be very serious for patients, especially those who may not have immediate access to tertiary centres if surgical intervention is needed. Patients that have a high risk of rupture may be considered for prophylactic ablation, embolization, or surgical excision could be considered in high-risk patients. The obvious question is, “What constitutes high-risk?” as no data exists to define this group of patients.

CONCLUSION

The consequences of a ruptured FNH nodule can be very serious for patients, especially those who may not have immediate access to tertiary centres if surgical intervention is needed. Patients that have a high risk of rupture may be considered for prophylactic hepatic resection, ablation, or embolization where appropriate. However, the benefits of these procedures must be weighed against the generally low incidence of ruptured nodules in patients with FNH, and the invasive nature of these treatments as preventative measures. Complications related to FNH resulting in ruptured nodules are rare. As a result, it is unclear as to which patients with FNH require more frequent follow-up versus those whose nodules will never rupture. Surveillance protocols for FNH patients are required so that risk factors for a rupture can be identified. The paucity of literature on this topic makes it difficult to provide specific recommendations that are evidence-based. Patients have to be individualized in their approach to therapy, risk factors, and potential benefits.