Features of Focal Nodular Hyperplasia on Multiple Imaging Modalities

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Outline

• Introduction to focal nodular hyperplasia (FNH)
  – Pathogenesis of FNH
  – Pathology of FNH
  – Epidemiology and typical clinical presentation

• Patient’s initial presentation

• Imaging
  – US
  – CT
  – MRI +/- Gd-BOPTA
  – DISIDA

• Patient’s clinical course

• FNH management in general
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FNH: What is it?

- Non-malignant hepatic tumor

Mitros, Frank A. Focal Nodular Hyperplasia Gross. uptodate.com
FNH: Pathogenesis

• Hyperplastic response to hyperperfusion by anomalous arteries
Brief Review: Normal Structure of Hepatic Lobule

Digestive System. Droualb.faculty.mjc.edu
FNH: Pathology

- Central **stellate scar** contains an abnormally large artery in the middle.
- Fibrotic tissue surrounds the abnormal artery, along with portal veins and bile ducts.
- Hepatocytes are normal.
- FNH lesions contain **sinusoids** and Kupffer cells (specialized macrophages), both of which are absent in hepatocellular adenoma (HA).

Mitros, Frank A. *FNH Trichrome. upToDate.com*
FNH: Epidemiology and Clinical Presentation

• Found predominantly in female between age 20-50.
• Patient taking OCPs tend to have larger and more vascular masses.
• Associated with hereditary hemorrhagic telangiectasia (Osler-Weber-Rendu disease) and hepatic hemangiomas.
• 90% solitary
• Two-thirds to three-fourths of patients are identified incidentally. Can present with abdominal discomfort or palpable mass.
Brief Review: Cross-Sectional Anatomy of the Abdomen
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Our Patient: Presentation

- 32F presenting w/ 5 days of episodic, crampy RUQ pain associated w/ n/v.
  - Episodes last a few hours
  - No fevers/chills
- PMH: Obesity, Hereditary hemorrhagic telangiectasia (HHT) → epistaxis requiring cauterization
- Family hx: HHT
- Social hx: 1 beer/wk, denies tobacco and recreational drugs
- Physical Exam:
  - Vitals stable
  - Abdomen soft, palpable liver edge, slightly tender. No hepatomegaly appreciated.
- Labs: AST 45
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#1 differential for crampy RUQ pain = biliary disease

RUQ Ultrasound

Examples of other ddx:
- Hepatitis
- Retrocecal appendicitis
- PUD
- Pancreatitis
- Liver abscess
- Liver, pancreatic, or biliary tract cancer
- RLL pneumonia
Our Patient: RUQ U/S

Sagittal RUQ U/S

- Liver: Homogeneous w/o focal lesion, or intra-hepatic duct dilatation
- Gallbladder: thin-walled and unremarkable without stones or pericholecystic fluid.
Our Patient: Work-Up (2)

Negative US

CT Abd/Pelvis

Numerous other ddx:
- Retrocecal appendicitis
- Hepatitis
- PUD
- Pancreatitis
- Liver abscess
- Liver, pancreatic, or biliary tract cancer
- RLL pneumonia
C+ axial abd CT: Portal venous phase

• At least 10 rounded hyper-enhancing lesions measuring up to 3.2cm
  • These lesions were isoechoic to liver parenchyma on U/S.
Ddx for liver mass

- FNH
- Hepatic adenoma
- HCC
- Hemangioma
- Metastatic cancer
- Cyst, eg. congenital, echinococcal (hydatid), polycystic kidney disease
- Hepatic (Amebic/Pyogenic) abscess
Ddx based on CT and presentation

- FNH
- Hepatic adenoma
- Hemangioma
- HCC
- Metastatic cancer—no primary CA, low risk
- Cyst, eg. congenital, echinococcal (hydatid), associated w/ polycystic kidney disease—Low attenuation on CT C+
- Hepatic abscess (Amebic/pyogenic)—low attenuation on CT C+
Cyst and hepatic abscess are ruled out because they have low attenuation on CT with contrast. Let’s explore how the other ddx appear on CT.
Ddx based on CT

• FNH
• Hepatic adenoma
• HCC
• Hemangioma
Companion Pt 1: HCC on arterial phase CT

- Main blood supply is the hepatic artery (normal liver parenchyma has 20/80 arterial/portal blood supply)

C+ axial abd CT: arterial phase
- Diffuse enhancement of a large mass in the right lobe
Companion Pt 1: HCC on portal venous phase CT

- Portal venous phase CT: rapid washes out and become iso/hypo-dense
  - Heterogenous enhancement due to fibrosis, fat, necrosis and calcifications.

C+ axial abd CT: Portal venous phase
- Heterogeneous mass, hypo-attenuating compared to adjacent liver parenchyma
- Septa hyper-attenuating because of contrast retention in the fibrotic tissue
Ddx based on CT

- FNH
- Hepatic adenoma
- HCC
- Hemangioma
Companion Pt 2: Hemangioma on arterial phase CT

C+ axial abd CT: arterial phase
- discontinuous, nodular, peripheral enhancement
Companion Pt 2: Hemangioma on portal venous phase CT

C+ axial abd CT: portal venous phase
- progressive enhancement w/ centripetal fill in
Companion Pt 2: Hemangioma on delayed phase CT

Morgan, Matt A., and Yuranga Weerakkody. Radiopaedia

C+ axial abd CT: delayed phase
- Continued centripetal fill in
Ddx based on CT

- FNH
- Hepatic adenoma
- Hemangioma
  - CT findings not characteristic, also should be seen on US
- HCC
  - Low risk, CT findings not characteristic, also should be seen on US
It is very important to differentiate FNH and hepatic adenoma!

- **FNH**
  - no evidence of malignant transformation
  - Very rarely symptomatic and need resection
- **Hepatic adenoma**
  - Associated w/ malignant transformation, spontaneous hemorrhage and rupture
- **Note:** 20% of FNH can be associated with other benign tumor and vascular malformation.
Hepatobiliary Gd Contrast Agent

- Dual route of elimination (renal and hepatobiliary excretion)
- FNH is hyper/iso-intense on delayed phase because functional hepatocytes and biliary ducts
- Hepatic adenoma is hypo-intense on delayed phase because hepatocytes and biliary ducts are not functional
- Gd-DTPA
  - 92% specificity for FNH
- Gd-BOPTA
  - Differentiation of FNH vs HA:
    - Sensitivity 96.9%, specificity 100%, PPV 100%, NPV 96.4% and overall accuracy 98.3
Our Patient: MRI T2

Axial MRI T2
• Slightly hyper-intense lesions
Our Patient: MRI T2

Coronal MRI T2

- Slightly hyper-intense lesions
Our Patient: MRI T1 early arterial phase

Axial MRI T1 early arterial phase w/ Gd-BOPTA (slightly delayed)

- Hyper-intense lesions (lack central scarring—atypical)
Our Patient: MRI T1 late arterial phase

Axial MRI T1 late arterial phase w/ Gd-BOPTA (slightly delayed)
- Slightly hyper-intense lesions
Our Patient: MRI T1 portal venous phase

Axial MRI T1 portal venous phase w/ Gd-BOPTA (slightly delayed)
- Slightly hyper-intense lesions
Our Patient: MRI T1 equilibrium phase

Axial MRI T1 equilibrium phase w/ Gd-BOPTA (slightly delayed)

- Slightly hyper-intense lesions
Our Patient: MRI T1 2hr delay

Axial MRI T1 2hr delay w/ Gd-BOPTA
- Hyper-intense lesions (atypical)
Our Patient: 2\textsuperscript{nd} Presentation

- 1 month later, pt presented again w/ RUQ pain.
  - Severity: 10/10
  - Radiates to R flank, shoulder and R arm
  - Labs were unremarkable

- More workup to rule out cholecystitis
  - RUQ U/S: unremarkable, no evidence of cholecystitis
  - DISIDA
Now let’s look at some selected images from our patient’s DISIDA scan
Our Patient: DISIDA (1)

• Liver
Our Patient: DISIDA (2)

- CBD
Our Patient: DISIDA (3)

- Gallbladder
- Duodenum
Our Patient: DISIDA (2) inverted

- FNH lesion w/ increased uptake

- FNH has functional hepatocytes and biliary ducts.
  - may be photon deficient in 60% of cases
- Hepatic adenoma takes up the tracer but doesn’t excrete it.
- Not specific enough to differentiate FNH vs adenoma
- HCC takes up tracer in 50% of the case
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Our Patient: Clinical Course

• Because of patient’s demographics, history, and pain following CCK administration (for DISIDA), pt underwent cholecystectomy.

• Gallstones were present on pathology which were not seen on imaging.

• Pain significantly improved

• Initial presentation may have been due to atypical FNH or cholecystitis
  • The sensitivity of ultrasound in the detection of acute cholecystitis is 95% and the specificity is 78-80%.
  • This case of FNH is atypical: the lack of central scarring in many lesions, the large number of nodules and pain
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FNH: management in general

- No evidence for malignant transformation for FNH in general.
- Follow up with Gd-DTPA at 3-6 months to confirm stability of the lesions
- If stable, no long-term routine follow-up is required.
Our Patient: management

• Because this patient had atypical lesions, additional follow up was recommended.
Take-home points: FNH

• Found predominantly in females between age 20-50.

• Associated with hereditary hemorrhagic telangiectasia

• 90% solitary

• 2/3 to 3/4 of lesions are identified incidentally. Can present with abdominal discomfort or palpable mass.

• Must be differentiated from hepatic adenoma

• 20% of cases are associated with other benign lesions
Take-home points: FNH on Imaging

– US: Typically hypoechoic w/ central scar, can be variable

– CT: Typically isodense on portal venous phase, atypical in this case.

– MRI: use hepatobiliary Gd contrast. Early enhancement. **Hyper-/iso-intense on 2hr delay.** Can be atypical and lack **central scar.**
  • Gd-BOPTA is more specific than Gd-DTPA

– DISIDA: normal or increased uptake and excretion (may be photon deficient in as many as 60% of patients). Not used to differentiate FNH from adenoma.
Browse the following slides for the appearance of *typical* FNH lesions on various imaging modalities
Typical FNH: U/S

Transverse RUQ U/S

- Hypoechoic mass
- Central scar

Focal nodular hyperplasia and hepatic adenomas. ultrasoundcases.info
Typical FNH: CT

CT C+: Arterial phase
- Hyperdense mass
- Central scar

CT C+: Portal venous phase
- Isodense mass
Typical FNH: MRI Gd-BOPTA

Axial MRI T1, Gd-BOPTA, 3hr delay

- Hyper-enhancing mass
- Central scar

Kruskal, Jonathan B. Focal nodular hyperplasia of the liver seen on Gd-BOPTA scan. Uptodate.com
References


References

- *Digestive System*. *Droualb.faculty.mjc.edu*. [http://droualb.faculty.mjc.edu/Lecture%20Notes/Unit%206/Spring%202006%20Digestive%20System%20with%20Figures.htm](http://droualb.faculty.mjc.edu/Lecture%20Notes/Unit%206/Spring%202006%20Digestive%20System%20with%20Figures.htm)
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