Fibrolamellar hepatocellular carcinoma

- Heterogeneously enhancing, large, lobulated mass with hypointense central scar and radial septa
  - Calcification and necrosis are common (> 50%)
  - Nodal metastases (> 50%)
  - Vary from 5-20 cm (mean: 13 cm)
  - "Satellite" nodules are often present
- Slow-growing tumor that usually arises in normal (noncirrhotic) liver
- Better prognosis than conventional HCC but still locally invasive and frequently metastatic

General Features

- **Best diagnostic clue**
  - Heterogeneously enhancing, large, lobulated mass with hypointense central scar and radial septa
- **Location**
  - Intrahepatic (80%)
  - Pedunculated (20%)
- **Size**
  - 5-20 cm (mean: 13 cm)
- Key concepts
  - Slow-growing tumor that usually arises in normal (noncirrhotic) liver
    - » May occur with underlying cirrhosis (< 5% of cases)
  - "Satellite" nodules are often present
  - Characteristic microscopic pattern
    - » Eosinophilic malignant hepatocytes containing prominent nuclei
  - Absence of pathologic markers (e.g., inclusions of α-fetoprotein bodies) that are present in conventional HCC
  - Better prognosis than conventional HCC but still locally invasive and frequently metastatic

Fibrolamellar hepatocellular carcinoma

- No specific risk factors
- Usually no underlying cirrhosis or liver disease
  - Hepatitis and cirrhosis may be present (< 5%)
- Healthy young adult with large liver mass.
- May have gynecomastia or other estrogenic effects.
- **Age**
  - Adolescents/young adults
    - » Age range of 5-69 years (mean: 23 years)
  - Most patients present in 2nd-3rd decade of life

CT

- **NECT**
  - Mass
    - » Well-defined contour
    - » Hypoattenuating and heterogeneous
  - Central scar and septa: Markedly hypodense
  - Calcification and necrosis are common (> 50%)
  - Hemorrhage is rarely seen
- **CECT**
  - Arterial phase
    - » Mass: Heterogeneous and hyperattenuating (80%)
  - Portal phase
    - » Mass: Iso-/hypoattenuating
  - Delayed phase (10 min)
    - » Mass: Isodense
    - » Scar and septa: Hyperdense
  - Malignant features
    - » Biliary and vessel invasion
    - » Nodal metastases (> 50%)
      - Porta hepatis and cardiophrenic nodes
    - » Lung metastases

MRI

- **T1WI**
  - Mass: Heterogeneous and slightly hypointense
  - Scar and septa: Hypointense
- **T2WI**
  - Mass: Heterogeneous and hyperintense
  - Scar and septa: Hypointense
- **T1WI C+**
  - Arterial and portal phases
    - » Intense heterogeneous enhancement of mass, not scar
  - Delayed phase
    - » Mass: More homogeneous enhancement
    - » Scar and septa: Delayed partial enhancement

Check List

- **Consider**
  - Differentiate FLC from FNH and conventional HCC
  - FLC simulates FNH due to presence of central scar in both tumors
- **Image Interpretation Pearls**
  - FLC: Bigger, more heterogeneous mass, frequently with calcified central/eccentric scar and features of malignancy (vessel/biliary obstruction, nodal invasion, and lung metastases)
  - Scar on T2WI: Hypointense (FLC), hyperintense (FNH)
  - Large, heterogeneous, hypervascular tumor in young adult

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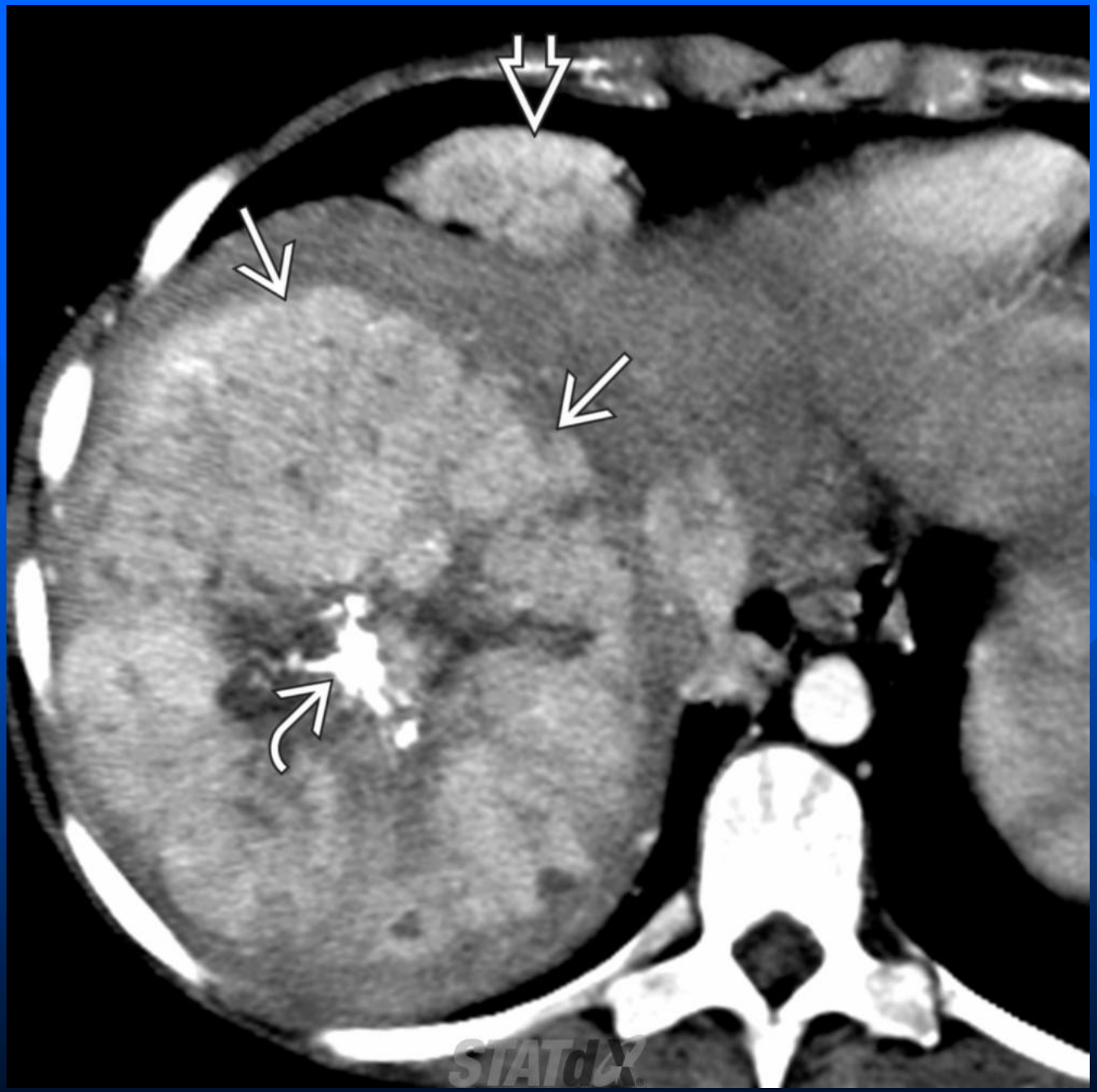
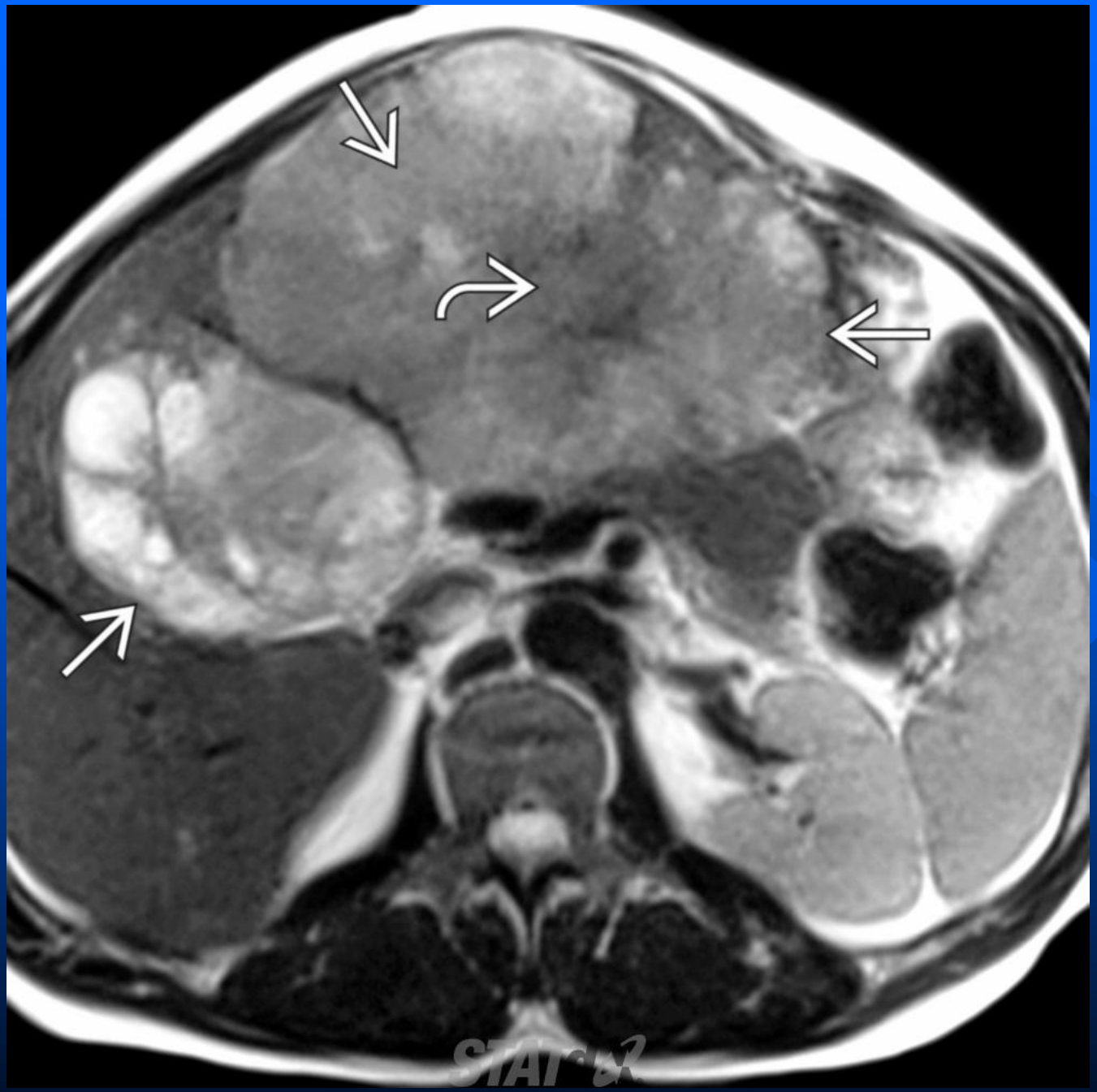
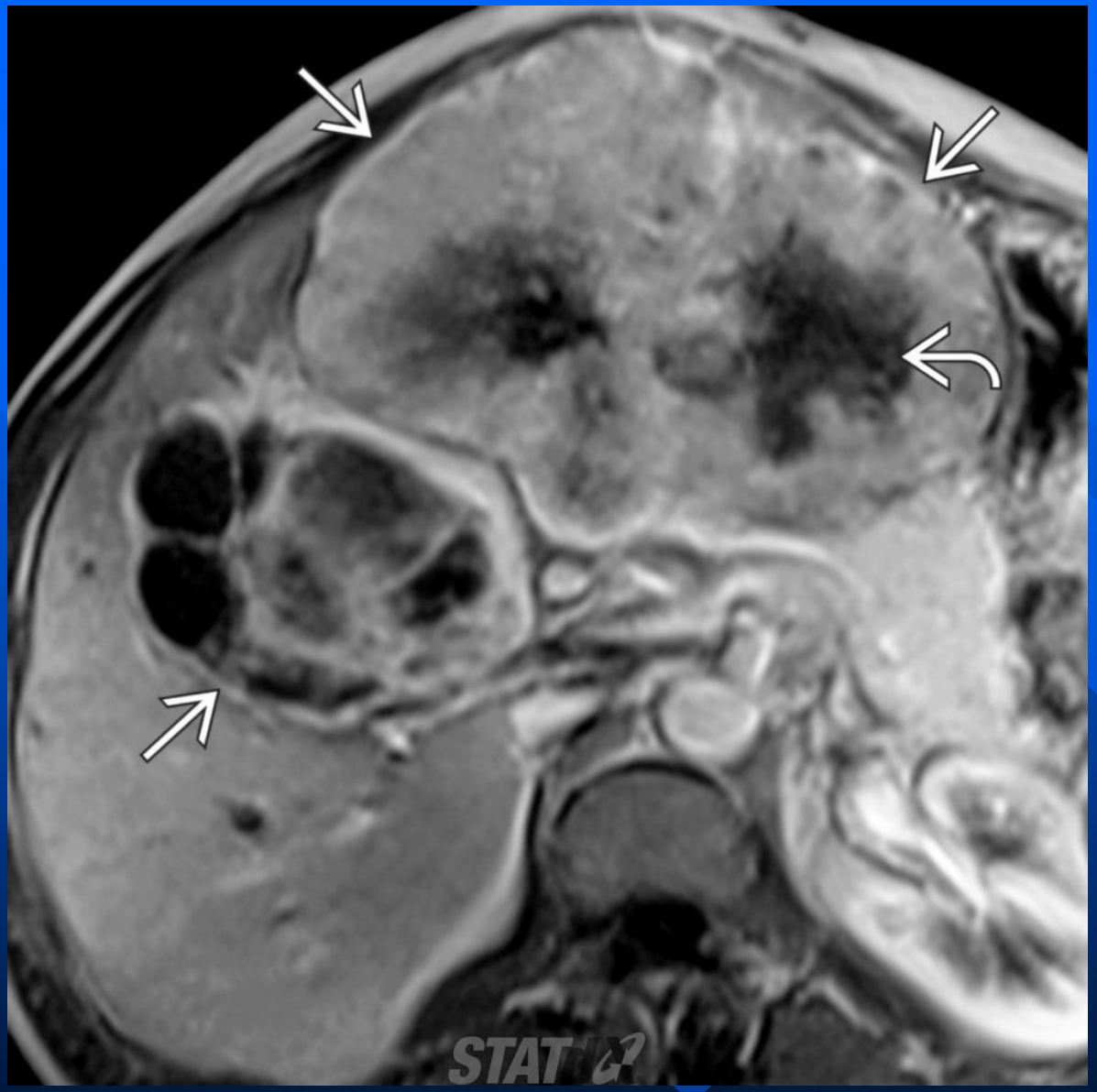
- Large
- Central scar is lower T2, may calcify
- Arterial phase enhancement is more irregular with radiating septations
- Central scar shows delyaed irregular enhancement.



Axial CECT in a 15-year-old boy shows a large, heterogeneous, hypervascular mass (white solid arrow) with a large, calcified central scar (white curved arrow) and cardiophrenic lymphadenopathy (white open arrow). In a young person, these findings are essentially diagnostic of fibrolamellar carcinoma (FLC)



T2WI MR in a 25-year-old man with early satiety and weight loss shows a large, heterogeneous, bilobed mass (white solid arrow) that is hyperintense to the liver. Note the hypointense central scar (white curved arrow).



Axial arterial phase T1 C+ MR in the same patient shows well-demarcated, lobulated, heterogeneous, and relatively vascular, dominant and satellite masses (white solid arrow). The fibrous scars (white curved arrow) show little enhancement on this phase of imaging but would often demonstrate delayed persistent enhancement in fibrolamellar carcinoma.

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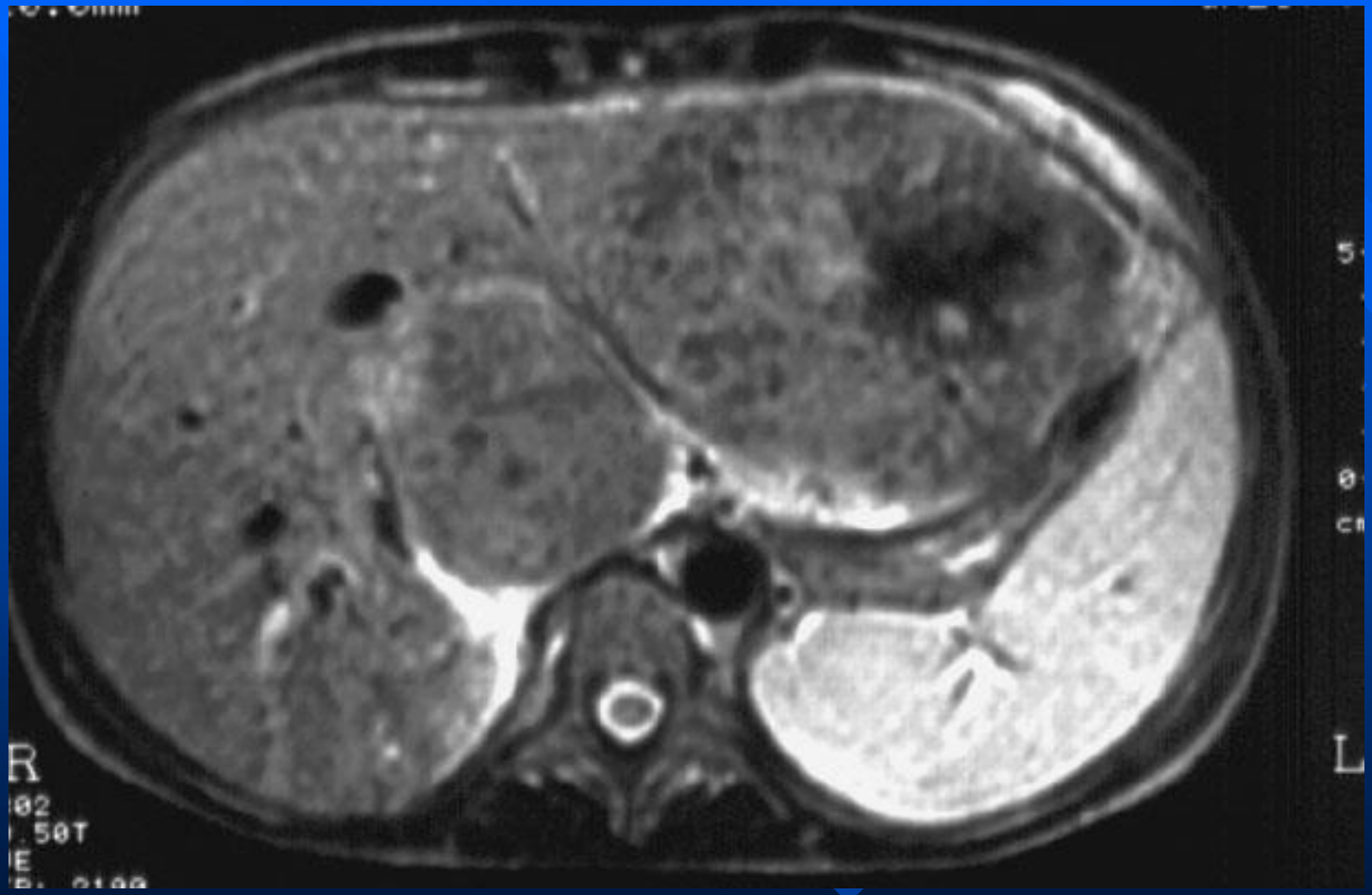

T2 Scar is dark

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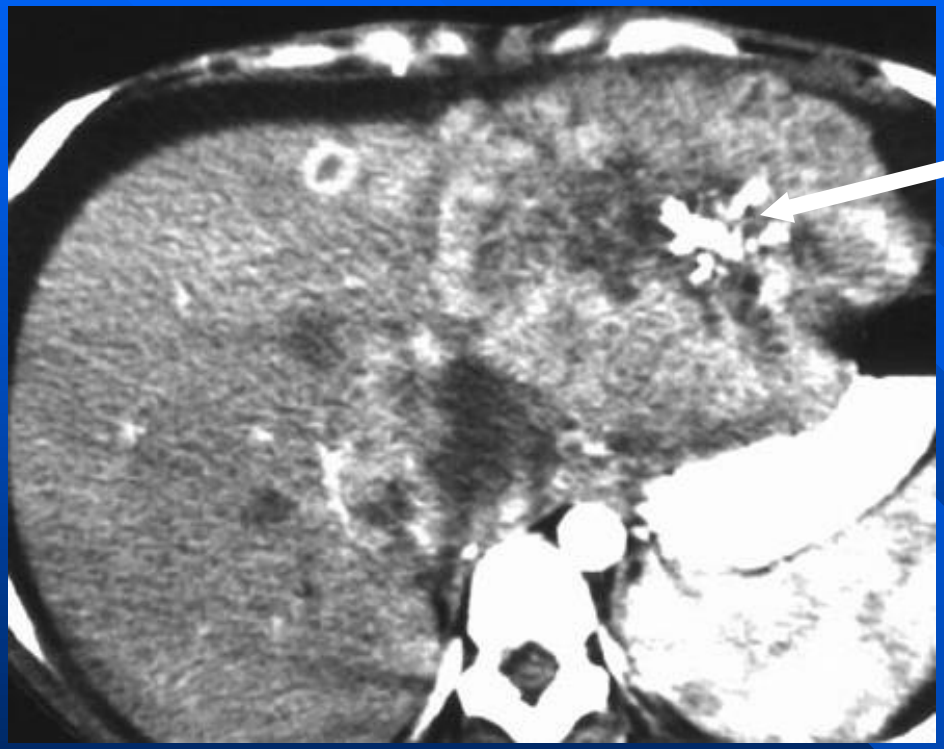

Ca+