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# Multifaceted Imaging of Fibrolamellar Carcinoma: A Rare Subtype of Hepatocellular Carcinoma

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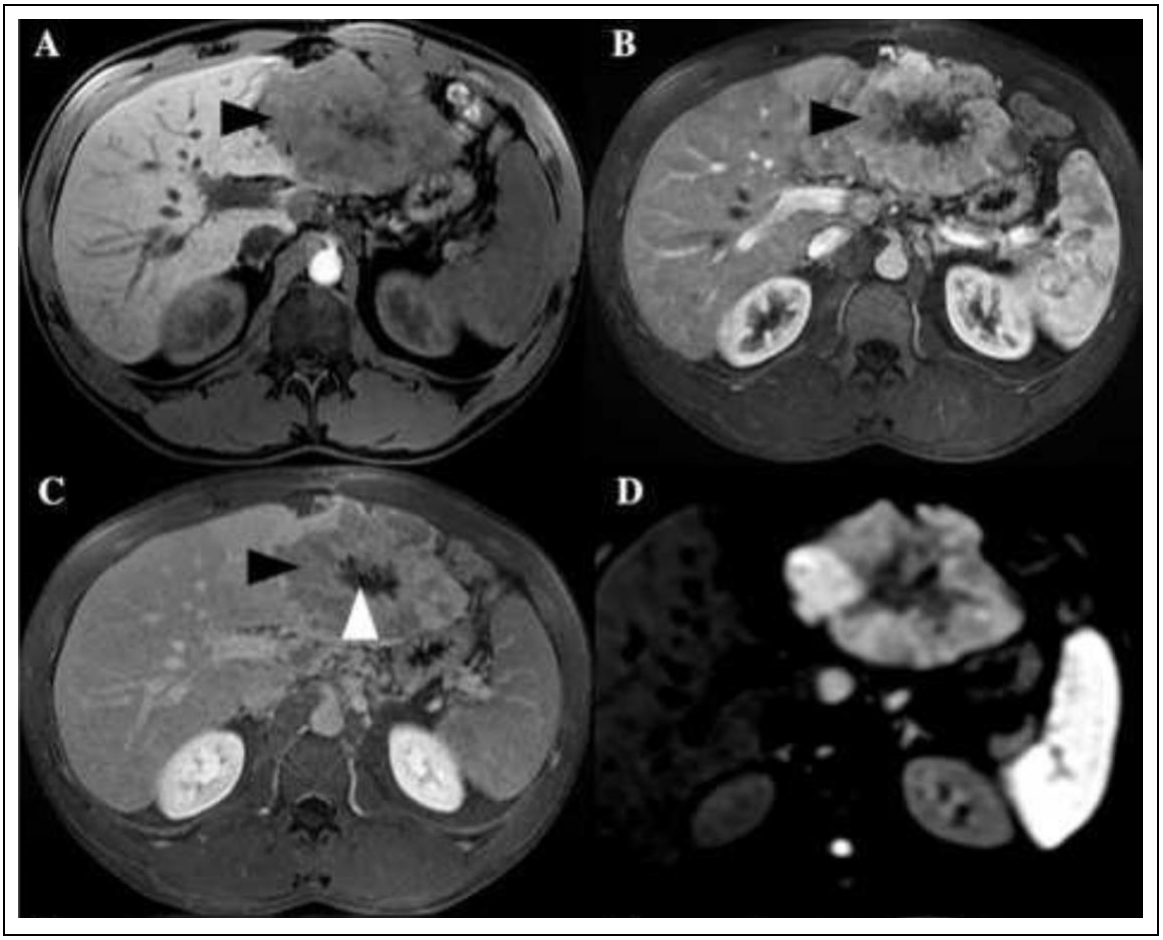

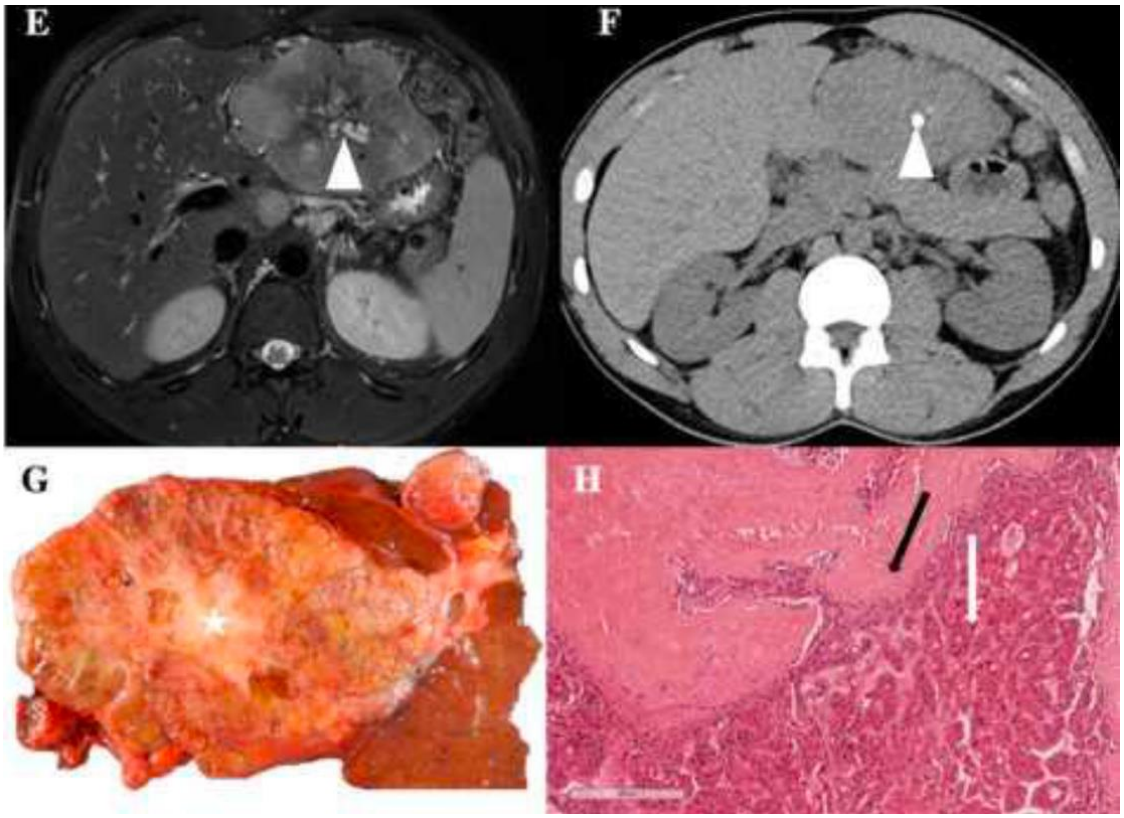


**Figure 1:** Fibrolamellar hepatocellular carcinoma in a 29-year-old male patient. The outer portion (black arrowhead) exhibited hypo-intensity compared to the liver on T1-weighted imaging **(A)** with arterial phase hyper-enhancement **(B),** washout on delayed phase **(C),** moderate hyper-intensity on T2-weighted-imaging **(D)** and showed marked hypersignal on diffusion b=800s/mm$^2$ **(E).** The inner portion (white arrowhead) displayed heterogeneity on T2-weighted imaging and lacked contrast enhancement, while calcification was observed on CT scan **(F).** This central area was clearly visible in the macroscopic specimen (star) **(G)** Microscopic examination **(H**, hematein-eosin-saffron coloration) showed compact clusters of large eosinophilic cells (black arrow) with prominent macro nucleoli embedded in a dense fibrous stroma (white arrow) made of collagen arranged in thin parallel lamellae.

## Clinical Image

A 29-year-old male patient with no pre-existing medical conditions presented with epigastric discomfort. Diagnostic investigations revealed a 12 cm hepatic mass located in the left lobe. Magnetic resonance imaging (MRI) showed no features of chronic liver disease and evidenced a focal lesion with two distinct components. The outer portion exhibited marked restriction of diffusion, arterial phase hyperenhancement (injection of extracellular gadolinium-based contrast agent, gadoteric acid), followed by washout. In contrast, the central part was heterogeneous and showed no enhancement. On computed tomography (CT), central calcifications were present. Another lesion was present in segment one, but there was no vascular invasion or any lesions outside the liver.

The patient subsequently underwent a partial hepatectomy. A gross pathological examination of the tumor confirmed the dual components detected by imaging. The histopathological assessment 14 revealed the outer portion to be composed of large eosinophilic hepatoid cells with prominent nucleoli organized in irregular trabeculae, while the central part featured a lamellar fibrous stroma. These investigative results supported a diagnosis of fibrolamellar carcinoma (FLC), a rare subtype of hepatocellular carcinoma (HCC), accounting for less than 1% of cases. First characterized histologically, FLC is distinguished by unique clinical, histological, and molecular attributes [1]. The etiopathogenesis of FLC remains unclear, with most diagnoses occurring in younger patients without a history of liver disease or significant liver fibrosis. Notably, in contrast to other HCC subtypes, lymph node metastasis is commonly observed [2].

Its pathological diagnosis can be challenging, given that classical HCC (i.e., Not Otherwise Specified [NOS]), emerging from chronic liver disease, may present areas of FLC. A consensual cut-off value defining the minimal tumor area percentage with FLC morphology required for the diagnosis is yet to be agreed upon. However, differentiating pure FLC (FLC homogeneity throughout the tumor) from mixed FLC (tumors demonstrating both FLC and NOS-HCC characteristics) is important as they appear to be two distinct entities. The pure FLC variant shows unique molecular oncogenic abnormalities with the presence of DNAJB1–PRKACA translocation, which was observed in this case [1]. Compared to patients with mixed FLC, individuals with pure FLC tend to be younger, exhibit lower serum alpha-fetoprotein levels, higher lymph node involvement rates, and improved overall survival rates post-surgical resection [3].

**Keywords:** Fibrolamellar hepatocellular carcinoma; Magnetic resonance imaging; Liver Neoplasms / genetics; Liver Neoplasms / pathology

**Abbreviations:** CT: Computed Tomography; FLC: Fibrolamellar Carcinoma; HCC: Hepatocellular Carcinoma; MRI: Magnetic Resonance Imaging; NOS: Not Otherwise Specified

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