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Case report

Voluminous fibrolamellar carcinoma in a young adult: A case report

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ABSTRACT

Introduction: Fibrolamellar carcinoma (FLC) is a rare primary liver cancer, typically presenting as a solitary nodule in young adults without underlying liver disease. Surgical resection is currently the only curative treatment.

Presentation of case: We report a 27-year-old woman with a 6-month history of moderate epigastric pain, right upper quadrant heaviness, and a 20-kg weight loss. Imaging studies (ultrasound, CT, and MRI) revealed an 11-cm mass in the right liver featuring a central scar and calcifications, highly suggestive of FLC. A liver biopsy confirmed the diagnosis. The patient subsequently underwent a right hepatectomy with en bloc resection of an adherent diaphragmatic collar and lymphadenectomy. Her postoperative course was uneventful, leading to discharge on postoperative day 10.

Discussion: FLC accounts for less than 1 % of primary liver tumors and is distinct from conventional hepatocellular carcinoma, primarily due to its occurrence in non-cirrhotic, younger patients. Characteristic radiologic findings include a well-circumscribed, large lesion with a central fibrous scar and occasional calcifications. Although the prognosis post-resection is generally favorable, recurrence rates exceed 60 %, emphasizing the need for aggressive surgical management and vigilant long-term follow-up.

Conclusion: FLC is a distinct clinical entity with improved surgical outcomes compared to classical hepatocellular carcinoma. Nonetheless, its high recurrence potential necessitates continued surveillance and further research to optimize treatment strategies.

1. Introduction

Fibrolamellar carcinoma (FLC) is a rare primary hepatic cancer. It is a rare, malignant entity, which appears as a single hepatic nodule and usually affects young adults and adolescents (1). Unlike standard hepatocellular carcinoma (HCC), fibrolamellar carcinoma is typically characterized by its occurrence in a younger population without history of underlying liver disease (2). FLC has no gender predominance. It represents less than 1 % of primary liver tumors (3), and has therefore been little studied. Surgical resection is the only curative treatment for FLC. Outcomes and prognosis of FLC are better than HCC, because of its younger onset and the absence of underlying liver disease (4).

We describe a case of voluminous FLC in a 27-years-old patient located in the right liver, without secondary lesion, which underwent right hepatectomy with resection of a diaphragmatic collar.

2. Presentation of case

We present a 27-years-old woman, with no significant pathological history who reported moderate epigastric pain with a feeling of heaviness in the upper right quadrant of the abdomen that has been evolving for 6 months associated with a weight loss of 20 k. The patient was in excellent general condition, without jaundice. Abdominal examination revealed hepatomegaly, without collateral venous circulation or ascites. Biology was normal, especially without disturbance of the hepatic tests. Alpha-fetoprotein was not increased. Abdominal ultrasound showed a 10 cm mass in the right liver with calcifications.

A complementary thoraco-abdominal computed tomography (CT) scan was performed showing a large tissue mass of the liver of 11 cm in diameter straddling segments VI, VII and VIII, spontaneously hypodense with a central scar more hypodense seat of calcifications (Fig. 1). This mass presents early intense enhancement at arterial time. It laminates

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the portal branch of the VII, spares the portal pedicle of the VIII, invades the right hepatic vein with development of communicants draining the posterior sector into the median hepatic vein which is respected. The mass is largely in contact with the retro hepatic inferior vena cava which is compressed but remains permeable. The rest of the liver is homogeneous, not dysmorphic and the portal trunk is permeable. Moreover, there were no secondary lesions.

Magnetic Resonance Imaging (MRI) of the liver showed a mass of 11 cm occupying the right liver. This mass was in T2 hypersignal, with a central scar in T2 hyposignal. The tumor was in T1 hyposignal without signal drop on the out of phase sequence. It presented an intense enhancement in arterial time without washout, with a center that remained hypovascularized. The rest of the liver is homogeneous in signal. The radiological aspect was strongly suggestive of a fibrolamellar carcinoma.

Liver biopsy was made, confirming diagnosis of FLC.

The patient was operated by laparotomy. Intraoperatively, the tumor had intimate contact with the right diaphragmatic dome and the right edge of the retro hepatic vena cava which is not invaded. A right hepatectomy via an anterior approach removing a diaphragmatic collar in monobloc (Fig. 2) was performed associated with an inter-aortico-caval, a retro portal and a hepatic pedicle lymphadenectomy. The diaphragmatic breach was sutured, and a chest drain was placed into the right pleural cavity at the end of the procedure. The postoperative course was uneventful, and the patient was discharged on hospital day 10 after surgery.

Gross examination showed the presence of a tumor formation measuring 8 × 5 cm of firm consistency, whitish and greenish colour, which is centered by a fibrous scar with a star-shaped appearance (Fig. 3A). A diaphragmatic collar of 6.5 × 7 cm was attached to the tumor (Fig. 3B). Histological examination (Fig. 4) showed a tumor of trabeculated architecture. The tumor cells were rounded and polygonal. Their cytoplasm was eosinophilic and granular. The nuclei were often enlarged with a prominent nucleolus. The stroma was abundant, fibrous and hyalinized. No vascular emboli, peri-nervous sheathing or lymph node invasion was seen. Moreover, the resection margins were clear, as well as the course of the retro hepatic vena cava.

3. Discussion

Liver cancer is the 6th most common cancer in the world in terms of incidence, and the 3rd most common cause of cancer death (5). Fibrolamellar carcinoma is a rare form of HCC accounting for less than 1 % of all primary liver cancers, compared to 60–80 % for classical HCC. This entity was first described by Edmonsson in 1956 (6) as a variant of hepatocellular carcinoma (HCC). It occurs in a younger population than HCC, without gender predominance and on a typically healthy liver without underlying hepatitis or cirrhosis. Due to the lack of specific symptoms or screening test, this tumor is frequently diagnosed at advanced stage. The most common symptoms in patients with FLC are abdominal mass, a sensation of heaviness in the abdomen, pain or weight loss.

The hepatic biological tests may be normal or modestly disturbed. Injected abdominal computed tomography (CT) and magnetic resonance imaging (MRI) of the liver are the gold standard for diagnosis. FLC is usually characterized by well-defined, large single tumor with lobulated margins, a central scar (33-60 %) with fibrous septa showing hypointensity on T2-weighted MR images, and isolated, fine calcifications in or near the central scar. In a series of 21 cases, Chagas and al. showed that the majority of patients (80 %) had a single nodule on radiological examination and the median tumor size is 120 mm, with extremes from 6.6 to 19 cm (7). Biopsy, either fine needle aspiration (FNA) or Trucut (TRU-CUT) biopsy, can provide a definitive diagnosis. In the diagnosis of FLC, tumor markers are not very useful. Alpha fetoprotein (AFP), traditionally the marker of HCC, is little or not increased in FLC (8). At present, there are no specific biomarkers for FLC. Histologically, cytokeratin 7 and epithelial membrane antigen may be useful in differentiating fibrolamellar carcinoma from hepatocellular carcinoma. On the basis of immunohistochemistry, fibrolamellar carcinoma appears to show both hepatocellular and biliary differentiation (9). In our case, immunohistochemistry was not performed due to the unavailability of reagents.

Recent advances have revealed that fibrolamellar carcinoma is genetically distinct from conventional hepatocellular carcinoma. A highly specific molecular hallmark of FLC is the DNAJB1-PRKACA fusion gene, which results from a 400-kb deletion on chromosome 19. This fusion is found in nearly 100 % of FLC cases and is absent in other liver tumors, reinforcing its diagnostic and potentially therapeutic significance. Although the exact mechanism through which this fusion drives oncogenesis remains under investigation, its consistent presence underscores FLC's unique biology and supports its classification as a distinct clinical entity. Ongoing studies are exploring its potential as a therapeutic target and as a biomarker for diagnosis or disease monitoring.

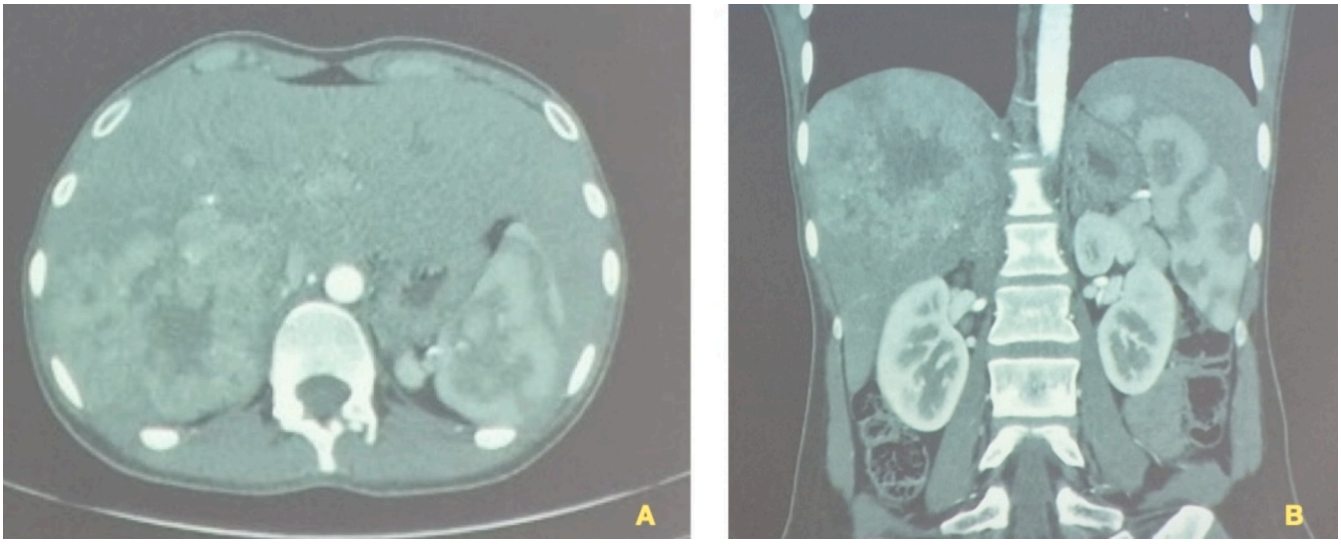

Fig. 1. Abdominal Computed Tomography in axial (A) and coronal (B) planes showing a large tissue mass of the liver of 11 cm in diameter, straddling segments VI, VII and VIII, which presents early intense enhancement at arterial time.

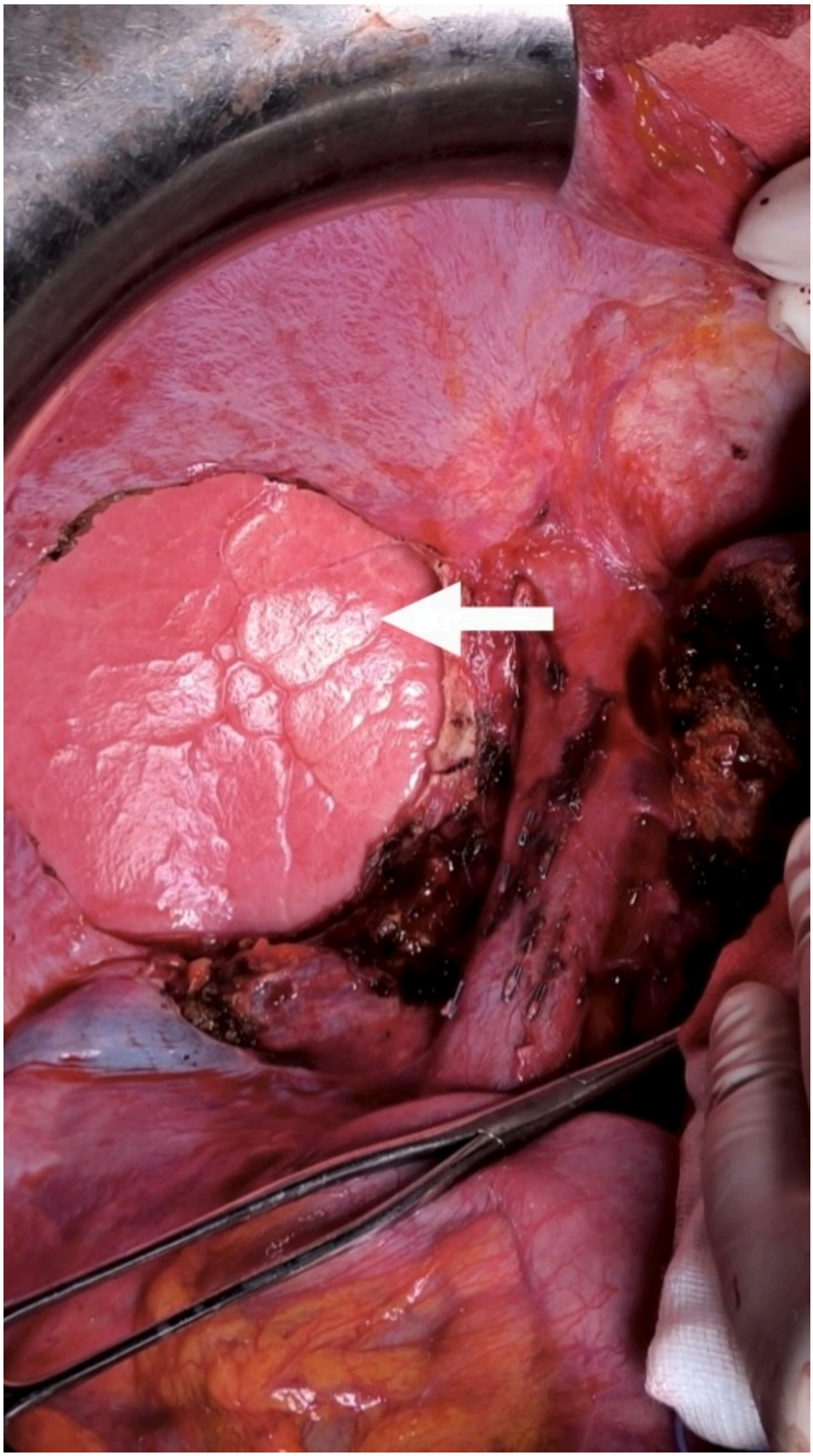

Fig. 2. Intraoperative view after right hepatectomy, lymphadenectomy and removing of a diaphragmatic collar. The diaphragmatic breach (white arrow) was sutured at the end of the procedure.

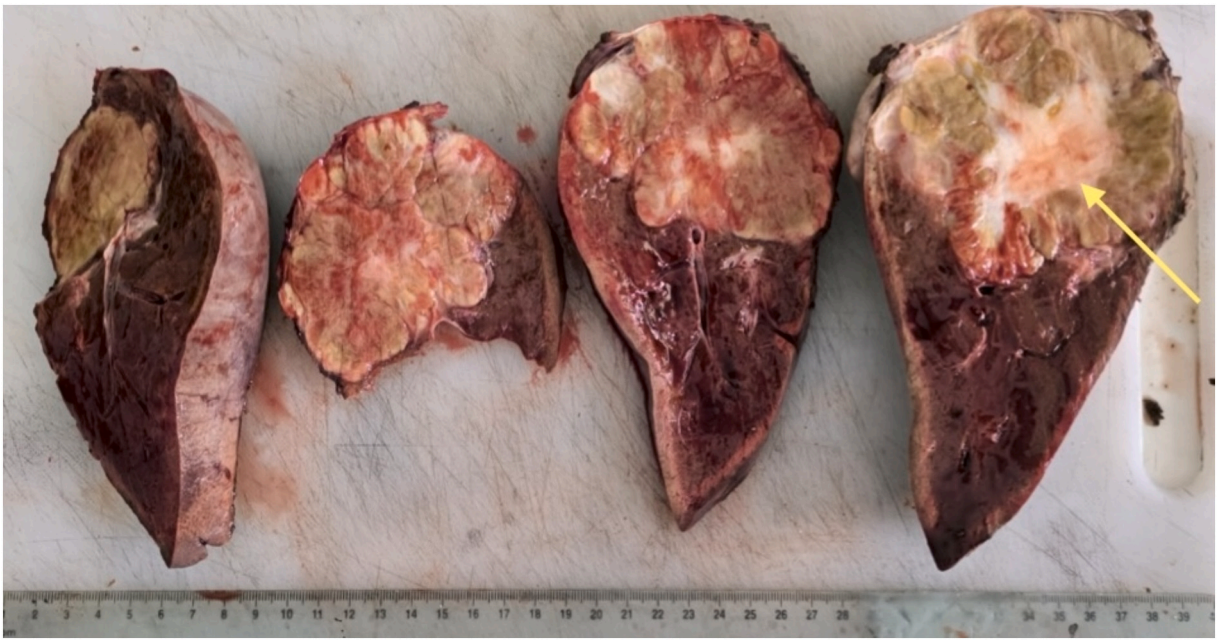

Fig. 3A. Gross examination showing the whitish/greenish tumor measuring 8 × 5 cm centered by a characteristic fibrous scar (yellow arrow). (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

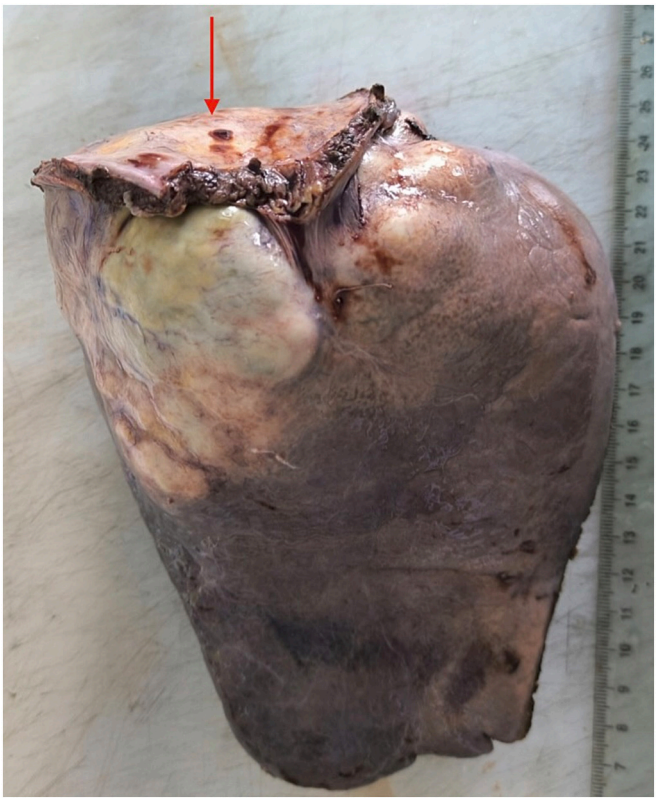

Fig. 3B. Gross examination showing the diaphragmatic collar attached to the tumor (red arrow). (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

At present, surgery with lymphadenectomy of the hepatic pedicle is the first line and the only curative treatment for fibrolamellar carcinoma. If it is an unresectable tumor that conforms to the Milan criteria, an orthotopic liver transplantation can be performed. In a retrospective American study, Mayo et al. (4) compared the long-term prognosis between FLC and HCC. They showed that the median overall survival (OS) for patients who have undergone surgical treatment for FLC was 75 month which was better than the median OS of 43 months for patients with HCC. However, certain factors such as age at diagnosis, or the quality of the underlying liver may be involved in these differences. A large American case series (10) reported that there was no statistically significant difference in 5-year survival after surgery between FLC and HCC if the age of the patient at diagnosis was less than 40 years and the underlying liver was healthy.

However several studies have reported a high rate of tumor recurrence after surgical resection of more than 60 % (11), worsening the prognosis because at present there is no clearly established treatment for unresectable forms.

While surgical resection remains the cornerstone and only curative approach for FLC, high recurrence rates post-resection necessitate consideration of systemic therapeutic strategies. Currently, no standardized adjuvant therapy has been established due to the rarity of the disease and limited clinical trials. However, emerging research is investigating molecularly targeted therapies, particularly those that inhibit the PKA signaling pathway, given the DNAJB1-PRKACA fusion's role. Immunotherapy and multi-kinase inhibitors, commonly used in HCC, are also being evaluated in small cohorts of FLC patients, although responses have been variable.

4. Conclusion

Fibrolamellar carcinoma was first described by Edmonson in 1956 as a variant of HCC. Today it is considered as a distinct form of primary liver cancer. Prognosis after surgical treatment is better than other conventional HCC due to the younger age at diagnosis and the absence of history of underlying liver disease. When the tumor is resectable, we must not hesitate to be aggressive surgically. Nevertheless, it remains a tumor with a high potential for recurrence requiring increased long-term follow-up. Further studies and clinical trials are needed to establish structured guidelines.

Consent for publication

Written informed consent was obtained from legal authorized

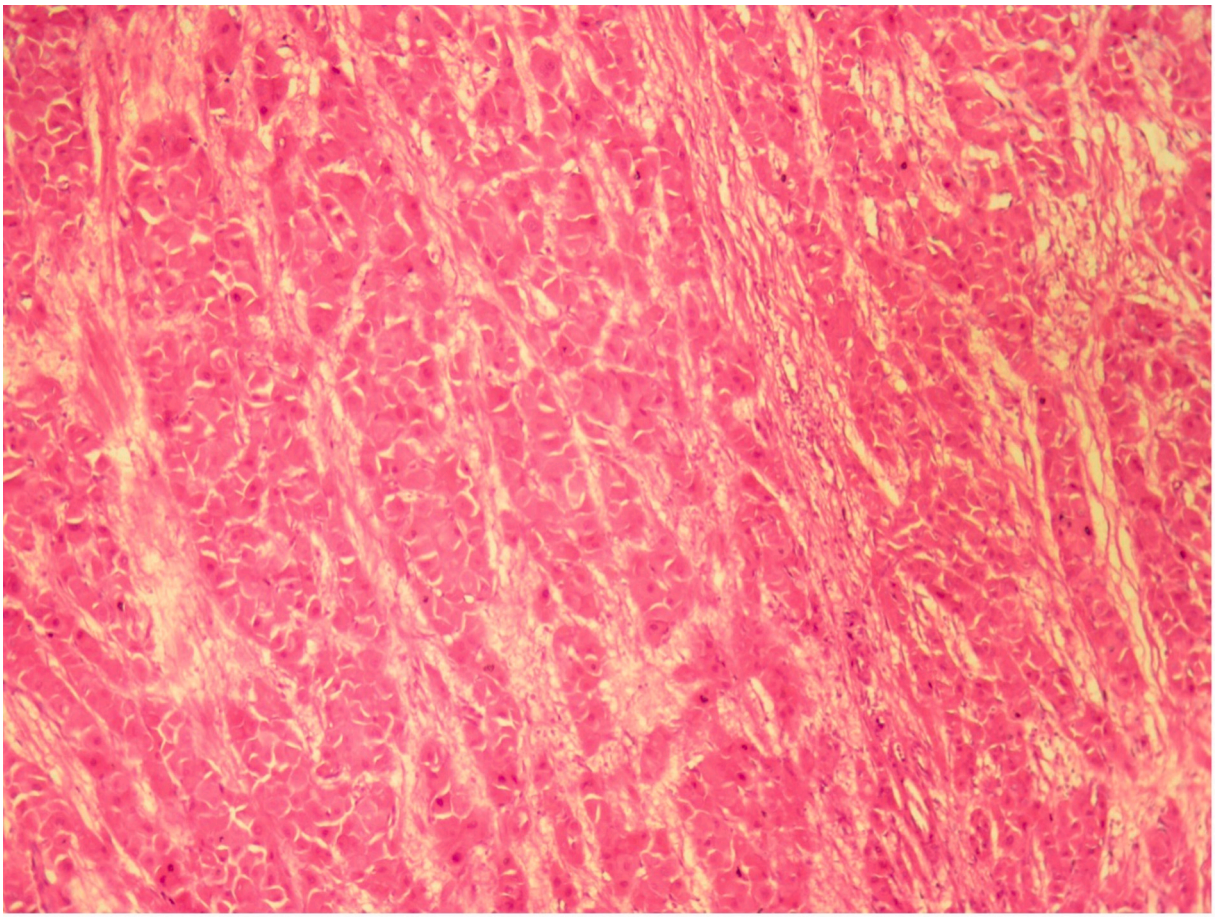

Fig. 4. Trabeculae and cords of neoplastic cells with abundant oncocytic cytoplasm in a background of dense collagen bundles (Hematoxylin-Eosin x 200).

representatives before the study. On request, a copy of the written consent is available for review by the Editor-in-Chief of this journal.

Ethical approval

Ethical approval is exempt/waived at our institution.

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Declaration of competing interest

The authors declare no competing interest.

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This work has been reported in line with the SCARE criteria [12].

Availability of data and materials

This published article includes all the required data.

References

[1] J.R. Craig, R.L. Peters, H.A. Edmondson, M. Omata, Fibrolamellar carcinoma of the liver: a tumor of adolescents and young adults with distinctive clinico-pathologic features, Cancer 46 (2) (15 juill 1980) 372–379.
[2] R.L. Meyers, Tumors of the liver in children, Surg. Oncol. 16 (3) (nov 2007) 195–203.
[3] H.B. El-Serag, J.A. Davila, Is fibrolamellar carcinoma different from hepatocellular carcinoma? A US population-based study, Hepatology 39 (3) (mars 2004) 798–803.
[4] S.C. Mayo, M.N. Mavros, H. Nathan, D. Cosgrove, J.M. Herman, I. Kamel, et al., Treatment and prognosis of patients with fibrolamellar hepatocellular carcinoma: a national perspective, J. Am. Coll. Surg. 218 (2) (févr 2014) 196–205.
[5] H. Sung, J. Ferlay, R.L. Siegel, M. Laversanne, I. Soerjomataram, A. Jemal, et al., Global cancer statistics 2020: GLOBOCAN estimates of incidence and mortality worldwide for 36 cancers in 185 countries, CA Cancer J. Clin. 71 (3) (2021) 209–249.
[6] H.A. Edmondson, Differential diagnosis of tumors and tumor-like lesions of liver in infancy and childhood, AMA J. Dis. Child. 91 (2) (févr 1956) 168–186.
[7] A.L. Chagas, L. Kikuchi, P. Herman, R.S.S.M. Alencar, C.M. Tani, M.A. Diniz, et al., Clinical and pathological evaluation of fibrolamellar hepatocellular carcinoma: a single center study of 21 cases, Clinics (Sao Paulo) 70 (3) (mars 2015) 207–213.
[8] S. Liu, K.W. Chan, B. Wang, L. Qiao, Fibrolamellar hepatocellular carcinoma, Am. J. Gastroenterol. 104 (10) (oct 2009) 2617–2624 (quiz 2625).
[9] S.C. Ward, J. Huang, S.K. Tickoo, S.N. Thung, M. Ladanyi, D.S. Klimstra, Fibrolamellar carcinoma of the liver exhibits immunohistochemical evidence of both hepatocyte and bile duct differentiation, Mod. Pathol. 23 (9) (sept 2010) 1180–1190.
[10] T. Eggert, K.A. McGlynn, A. Duffy, M.P. Manns, T.F. Greten, S.F. Altekruse, Fibrolamellar hepatocellular carcinoma in the USA, 2000-2010: a detailed report on frequency, treatment and outcome based on the surveillance, epidemiology, and end results database, United European Gastroenterol J 1 (5) (oct 2013) 351–357.
[11] P. Herman, A.L. Chagas, M.V. Perini, F.F. Coelho, G.M. Fonseca, V.A.F. Alves, et al., Surgical treatment of fibrolamellar hepatocellular carcinoma: an underestimated malignant tumor? Hepatobiliary Pancreat. Dis. Int. 13 (6) (15 déc 2014) 618–621.
[12] C. Sohrabi, G. Mathew, N. Maria, A. Kerwan, T. Franchi, R.A. Agha, The SCARE 2023 guideline: updating consensus Surgical CAse REport (SCARE) guidelines, Int. J. Surg. Lond. Engl. 109 (5) (2023) 1136.