International Journal of Medical Science and Clinical Research Studies

ISSN(print): 2767-8326, ISSN(online): 2767-8342

Volume 03 Issue 02 February 2023

Page No: 209-211

DOI: https://doi.org/10.47191/ijmscrs/v3-i2-11, Impact Factor: 5.365

A Case of Fibrolamellar Hepatocellular Carcinoma at the High Specialty Hospital of Veracruz, Mexico

Jessica Ariatna Carreto Navarrete[1], Estanislao Antonio Calixto[2], Pedro Arredondo Ruiz[3], David Sandoval Sánchez[4], Josué Isaac Olarte García[5], Charit Nayelly Chang Cruz[6], Alfredo Bautista de la Cruz[7]

[1,2,4,5,6,7]Department of Internal Medicine. High Specialty Hospital of Veracruz SESVER. Veracruz, Mexico

[3]Department of Oncology. High Specialty Hospital of Veracruz SESVER. Veracruz Mexico

ABSTRACT

Fibrolamellar hepatocellular carcinoma is a primary hepatic malignancy, primarily affecting adolescents and young adults without underlying liver disease. We present the case of a 22-year-old male farmer who presents burning pain in the epigastrium with an intensity of 2/10 on the VAS scale, on palpation with the presence of a tumor in the right hypochondrium with ultrasound of the liver and bile ducts with evidence of liver mass in the segment VIII, in addition to exploratory laparoscopy with the discovery of multiple implants in 70% of the liver surface. In the histopathological analysis with morphological findings compatible with fibrolamellar carcinoma. In the laboratory with AST 356 UI/L, ALT 205 UI/L, GGT 930 U/L, LDH 343 UI/L AF 638 UI/L, for which treatment was started with atezolizumab 1200 mg and bevacizumab 1000 mg. Continuing to follow up. Fibrolamellar hepatocellular carcinoma is a variant of hepatocellular carcinoma, which usually presents with non-specific symptoms. The imaging approach is fundamental in the diagnosis, being the anatomopathological confirmation. Despite being low frequency tumors, they should be suspected in all young patients with a palpable mass in the liver.

KEYWORDS: Fibrolamellar, Hepatocellular, Carcinoma

ARTICLE DETAILS

Published On:
11 February 2023

Available on:
https://ijmscr.org/

INTRODUCTION

Fibrolamellar hepatocellular carcinoma (FCH) is a rare primary liver malignancy (accounting for less than 1% of all diagnosed primary liver tumors), primarily affecting adolescents and young adults without underlying liver disease (cirrhosis or hepatotrophic virus infection). It receives the term fibrolamellar for the thick bands of intratumoral fibrous collagen[1].

The most important prognostic factors are the stage of the disease at the time of diagnosis and the resectability of the tumor, with an average survival of 7 years in stages I and II, 4.5 years in stage III and 2 years in stage IV; in patients who are not candidates for surgery, a half-life of 1 year is reported, and 5 years with surgical treatment[2].

BACKGROUND

The incidence of hepatocellular carcinoma (HCC) has increased worldwide over the past 20 years and is expected to increase by 2030 in some countries, including the United States, while in other countries, such as Japan, the incidence has begun to decline[2,3].

In 2012, liver cancer was the fifth most common cancer in men (554,000 new cases) and ninth in women (228,000 new cases) and the second most common cause of cancer-related death (746,000 estimated deaths) all over the world[4].

Less than 1% of primary liver tumors in the US and 5.8% of liver tumors in Mexico are CHF. However, incidence rates are relatively homogeneous among various racial and ethnic groups throughout the world[4,5].

The DNAJB1-PRKACA fusion gene drives the pathogenesis of this unique tumor in more than 95% of cases. It was first described by Edmondson in 1956 in the pediatric population along with conventional hepatocellular carcinoma (HCC)[6].

The age-adjusted global incidence between 2000 and 2016 was 0.02 per 100,000 per year. In general, men have a higher incidence compared to women (0.03 vs 0.02 per 100,000 per year)[7]. In a large series of databases, the average age at diagnosis was 39 years, and 60% of patients were diagnosed under 40 years of age. It has a unique pathogenesis (more

stable genome, fewer somatic mutations, and lower levels of promoter methylation), with distinct histologic, clinical, and epidemiologic features compared to conventional HCC[8]. A case is presented at the third level of care.

CLINICAL CASE

A 22-year-old male working as a peasant, with prolonged exposure to pesticides and insecticides, in addition to alcohol consumption of more than 100 grams at a rate of 4 times a month and methamphetamine use for the past 3 years. He presented burning pain in the epigastrium with an intensity of 2/10 on the VAS scale, which subsidized with food intake, on palpation with the presence of a tumor in the right hypochondrium that caused hyperalgesia. He was evaluated by the general surgery service who performed ultrasound of the liver and bile ducts and simple abdominal tomography (Fig. 1A) with evidence of a liver mass in segment VIII, as well as exploratory laparoscopy (Fig. 1B) with the discovery of multiple implants in 70% of the liver surface. Histopathological analysis (Fig. 1C) reveals bands of dense connective tissue and scan reactive-type liver tissue, morphological findings compatible with fibrolamellar carcinoma. The oncology department requests a panendoscopy that reports small baveno esophageal varices and laboratory results with hemoglobin 11 mg/dL, platelets 534,000 mm3, leukocytes 7.6 mm3, PT 12.9 sec, TTP 33.4 sec, INR 1.1, glucose 84 mg/dL , albumin g/ dL, creatinine 1.02 mg/dL, BUN 16 mg/dL, AST 356 IU/L, ALT 205 IU/L, GGT 930 U/L, LDH 343 IU/L, and alkaline phosphatase 638 IU/L per which started treatment with atezolizumab 1200 mg and bevacizumab 1000 mg. Continuing in follow-up, conditioned by an adequate response to chemotherapy.

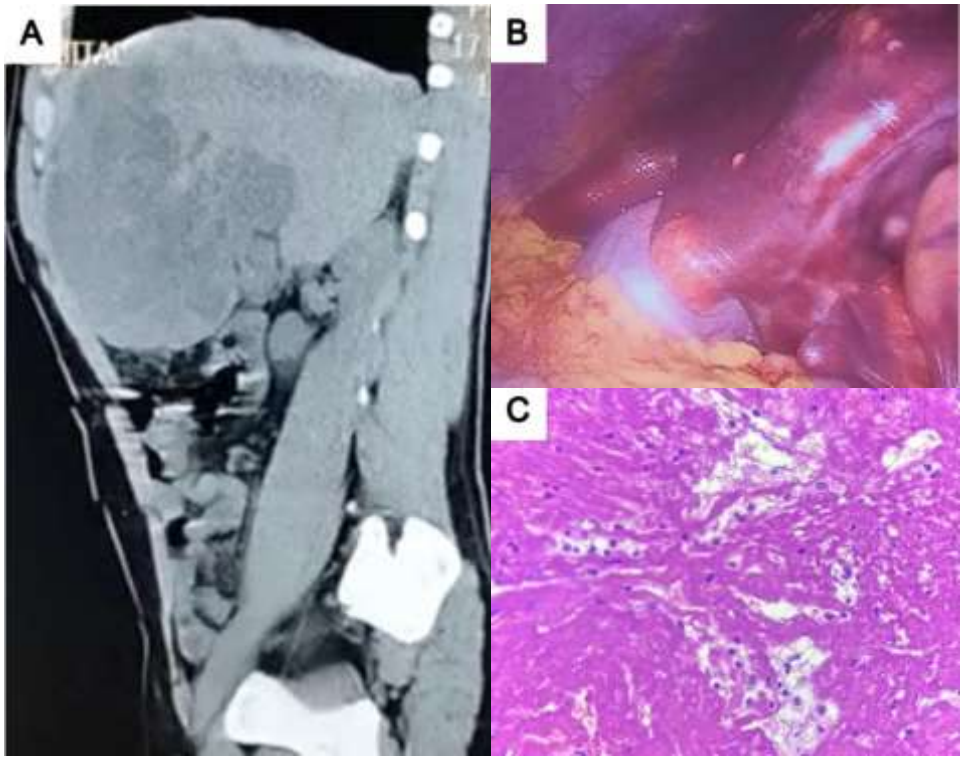

Figure 1. A) Simple abdominal tomography in sagittal section with hypodense intrahepatic lesion. B) Liver surface with multiple nodular lesions. C) Cells with granular cytoplasm in some areas and clear in others, central nuclei, mild atypia, and scarce mitosis, which form tubular structures.

DISCUSSION

Fibrolamellar hepatocellular carcinoma commonly debuts with non-specific symptoms. The average age of presentation is 20 years, being clinical stage IV at the time of diagnosis in 50% of cases. No gender predominance[1,2,4]. No previous liver disease. Frequently hepatitis B and C are negative, as well as in alpha-fetoprotein they are not usually elevated3. Regarding the pathology, they are usually a large tumor and in cords separated by bands of collagen fibers. Distant metastases occur in 30% of patients[4,6].

The imaging approach is fundamental in the diagnosis, being the anatomopathological confirmation. In the tomography, the characteristic findings are the presence of large lesions and multiple densities, with lobulated edges. In more than 60% of cases, they present a large central scar, with a diameter greater than 2 cm and the presence of fibrous septa. Calcifications can be observed in 50% of cases. Tumor necrosis and intratumoral haemorrhage are less frequent[7,8,9].

Lymphatic metastases are more frequent and occur in 50% of cases. Lymphadenopathy is common in the hepatic hilum and in the hepatoduodenal ligament, in some cases it is observed in the retroperitoneum, being a highly relevant prognostic factor. Intrahepatic satellite lesions, biliary obstruction and vascular invasion are factors associated with a worse prognosis[9,10]. With less response to chemotherapy; It can be useful in advanced cases.

Surgical resection, with a high rate of recurrence at ten years. Recurrences are frequent and range between 33% and

100%[11]. Prognostic factors are the stage of the disease, multiplicity, tumor thrombosis, lymphovascular invasion, metastasis, and whether or not a complete surgical resection was performed[12]. The DNAJB1-PRKACA fusion gene has been linked to the pathogenesis of this unique tumor in more than 95% of cases, this being the cornerstone for the development of future therapies[13].

CONCLUSION

CHF is an entity with high mortality despite timely treatment. Given this scenario, the study and evidence of these cases allow us to evaluate the degree of response. Despite being low frequency tumors, they should be suspected in all young patients with a palpable mass in the liver.

CONFLICTS OF INTEREST

None reported.

REFERENCES

I. Mayo SC, Mavros MN, Nathan H. Treatment and Prognosis of Patients With Fibrolamellar Hepatocellular Carcinoma; a National Perspective. J Am Coll Surg. 2014;218(2):196-205.
II. Petrick JL, Kelly SP, Altekruse SF. Future of Hepatocellular Carcinoma Incidence in the United States Forecast Through 2030. J Clin Oncol 2016;34: 1787–1794.
III. White DL, Thrift AP, Kanwal F. Incidence of Hepatocellular Carcinoma in all 50 United States, from 2000 Through 2012. Gastroenterology 2017;152: 812–820.e5.
IV. Serag HBE, Dávila JA. ¿Es el Carcinoma Fibrolamelar Diferente del Carcinoma Hepatocelular. Un estudio basado en la población de EE. UU. Hepatología 2004;39:798-803.
V. Eggert T, McGlynn KA, Duffy A. Carcinoma Hepatocelular Fibrolamelar en los EE. UU., 2000-2010: Un Informe Detallado sobre la Frecuencia, el Tratamiento y el Resultado basado en la Base de Datos de Vigilancia, Epidemiología y Resultados Finales. United European Gastroenterol J 2013;1: 351-7.
VI. Nasr JA, Villalobos LG, Nuncio J, Maldonaldo H, Bornstein-Quevedo L. Fibrolamellar Hepatocellular Carcinoma in Mexican Patients. Pathol Oncol Res. 2002;8: 133-7.
VII. Ramai D, Ofosu A, Lai JK, Gao ZH, Adler DG. Fibrolamellar Hepatocellular Carcinoma: A Population-Based Observational Study. Dig Dis Sci. 2020 Feb 12.
VIII. Ganeshan D, Szklaruk J, Kundra V, Kaseb V, Rashid A, Elsayes KM. Imaging Features of Fibrolamellar Hepatocellular Carcinoma. AJR Am J Roentgenol 2014; 202(3): 544-52
IX. Eggert T, McGlynn KA, Duffy A, Manns MP, Greten TF, Altekruse SF. 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. 2013;1: 351-7.
X. Ang CS, Kelley RK, Choti MA, Cosgrove DP, Chou JF, Klimstra D, Clinicopathologic characteristics and Survival Outcomes of Patients with Fibrolamellar Carcinoma: Data From the Fibrolamellar Carcinoma Consortium.
XI. Kaseb AO, Shama M, Sahin IH, Nooka A, Hassabo HM. Prognostic Indicators and Treatment Outcome in 94 Cases of Fibrolamellar Hepatocellular
XII. Carcinoma.Oncology. 2013;85(4):197.
XIII. Stipa F, Yoon SS, Liau KH, Fong Y. Outcome of Patients With Fibrolamellar Hepatocellular Carcinoma. Cancer. 2006; 106(6): 1331.
XIV. Tomasini MD, Wang Y, Karamafrooz A, Karamafrooz A, Li G, Beuming T, et al. Conformational Landscape of the PRKACA-DNAJB1 Chimeric Kinase, the Driver for Fibrolamellar Hepatocellular Carcinoma. Sci Rep 2018; 8(1):720.