

Fibrolamellar Hepatocellular Carcinoma: Is it Rare in Morocco?

H Abid, FZ Hamdoun*, N Lahmidani, M El Yousfi, D Benajah, M El Abkari, N Aqodad and SA Ibrahim

Department of Hepato-Gastro-Enterology, University Hospital Hassan II - Fez, Faculty of Medicine - Fez, University Sidi Mohammed Ben Abdelal - Fez, Morocco

***Corresponding Author:** FZ Hamdoun, Department of Hepato-Gastro-Enterology, University Hospital Hassan II - Fez, Faculty of Medicine - Fez, University Sidi Mohammed Ben Abdelal - Fez, Morocco.

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Abstract

Introduction: Fibrolamellar hepatocellular carcinoma (FLHCC) is a relatively rare malignant hepatic tumor distinct from traditional hepatocellular carcinoma in terms of demographic, radiological and prognosis features. We report a series of patients carrying fibrolamellar hepatocellular carcinoma through which we will identify the characteristics of this rare tumor.

Methods: This is a retrospective study including patients with fibrolamellar hepatocellular carcinoma between January 2010 and June 2019. The study focused on socio-demographic, clinical, imaging data, histological, treatment options and survival. Biopsy was reserved when the diagnostic was uncertain.

Results: During the study period, we recruited 7 patients with FLHCC. The average age was 44.7 years [19; 57]. The sex ratio F/M was 1.33. The circumstances of discovery of the disease were dominated by the feeling of heaviness in the right hypochondrium in 57.14%. The average alpha fetoprotein level was 26.16 ng/ml [2.3; 87.8]. Imaging was performed for all of our patients. The mean diameter of the largest lesion was 7.3 cm. More than two thirds of our patients underwent a regular hepatectomy. Only one patient was started on Sorafenib. The decline was 32 months. Survival at 5 years was 71.42%.

Conclusion: Fibrolamellar hepatocellular carcinoma seems to have a better prognosis when diagnosis and treatment are done early. The 5-year survival in our series was 71.42%.

Keywords: *Fibrolamellar Hepatocellular Carcinoma; Hepatocellular Carcinoma, Prognostic*

Introduction

Fibrolamellar hepatocellular carcinoma (FLHCC) is a variant of hepatocellular carcinoma (HCC). It is a relatively rare malignant hepatic tumor distinct from traditional hepatocellular carcinoma in terms of demographic, radiological and prognosis features. It comprises approximately 1 - 9% of all HCCs with an age adjusted incidence rate estimated at 0.02 per 100,000 [1]. The term 'fibrolamellar' is derived from the histological presence of thick fibrous collagen bands surrounding the tumor cells [2]. Most cases of FLHCC are diagnosed in persons with no or minimal liver disease and seems to have a better prognostic, while the majority of HCC cases are diagnosed in patients with significant liver damage, usually cirrhosis with poor prognosis.

We report a series of patients carrying FLHCC through which we will identify the characteristics of this rare tumor.

Methods

This is a retrospective series including patients with FLHCC without liver disease between January 2010 and June 2019. The study focused on socio-demographic, clinical, imaging data, histological, treatment options and survival. Biopsy was reserved when the diagnostic was uncertain.

The results were analyzed using the Statistics Package for Social Science software (SPSS version 20) for Windows. Quantitative variables were expressed as average \pm standard deviation. Qualitative variables were expressed in number and percentage (%).

Results

During the study period, we recruited 7 patients with fibrolamellar hepatocellular carcinoma. The average age was 44.7 years [19; 57]. The sex ratio F/M was 1.33 with female predominance. The circumstances of discovery were dominated by the feeling of heaviness in the right hypochondrium in 57.14% followed by the pain in the right hypochondrium in 42.8%. The average alpha fetoprotein level was 26.16 ng/ml [2.3; 87.8]. It was negative in 42%.

Abdomen CT scan and MRI, including an unenhanced phase followed by an intravenous contrast-enhanced hepatic arterial phase, a portal venous phase, and a delayed phase, were performed for all of our patients. They have objectified large heterogeneous well-defined lesions with a lobulated outline, calcifications and central stellate scar were seen in 85% (Figure 1). The extension assessment allowed the discovery of pulmonary metastases in one case. The mean diameter of the largest lesion was 7.3 cm [2.3 cm - 18.23 cm].



Figure 1: Hepatic MRI showing large heterogeneous well-defined lesions with a lobulated outline and central stellate scar.

All the files were discussed in a multidisciplinary concertation meeting. Most of our patients underwent a regular hepatectomy (85.7%) and only 14.2% were started on a systemic chemotherapy based on Sorafenib due to the presence of pulmonary metastasis at the time of diagnosis. The decline was 32 months. Survival at 5 years was 71.42%: One patient died in the postoperative period from hemorrhagic shock another patient recurred after 5 months in the postoperative period.

The characteristics of patients with fibrolamellar hepatocellular carcinoma in our series are summarised in table 1.

	N (%)
Middle age	44.7 years [19; 57]
Sex ratio (H/F)	1,33
Clinicals symptoms	
• Heaviness in the right hypochondrium	4 (57.14%)
• Pain in the right hypochondrium	3 (42.8%)
The average Alpha foetoproteinerate	26.16 ng/ml [2.3; 87.8]
The average size of the tumor	7.3 cm [2.3 cm - 18.23 cm]
Treatment	
• Surgery	6 (85.71%)
• Sorafenib	1 (14.28%)
The decline	32 months
Survival at 5 years	5(71.42%)

Table 1: The characteristics of patients with fibrolamellar hepatocellular carcinoma in our series.

Discussion

Several studies have looked at fibrolamellar hepatocellular carcinoma and have shown its difference from traditional hepatocellular carcinoma. Compared to HCC, patients with FLHCC are more likely to be younger with no or minimal liver disease and seems to have a better prognostic, while the majority of HCC cases are diagnosed in patients with significant liver damage, usually cirrhosis with a poor prognosis.

In the study of Hashem B., *et al.* [3], the mean age of diagnosis of FLHCC was 39 years with female predominance which joins our finding. Liver resection and liver transplantation are the two potentially curative surgical treatment options [4]. The proportion of patients receiving potentially curative therapy (resection, transplantation) was 41.7% in the study of Hashem B., *et al.* [3] vs 85.7% in our series who underwent a regular hepatectomy.

The role of chemotherapy in the management of FLHCC is not clear as these tumors tend not to be chemo-responsive. Some response has been observed with platinum based and interferon alpha 2b based regimens [5]. Few reports have also suggested the use of gemcitabine-based regimens [6]. Maniaci., *et al.* [5] shown that patients who received multimodality therapy for recurrence had median overall survival of 9.3 years with two patients showing at least partial response to cisplatin and fluorouracil. The use of chemotherapy in our series did not show any benefit in terms of survival: only one patient was put on Sorafenib, the overall survival did not exceed 5 months.

According to the literature, overall survival appears to be better in FLHCC. A review of 17 cases in the study of Epstein BE., *et al.* [7] with non resectable metastatic FLC, reported a median survival of 57 months. The overall survival at 5 years in Hashem B., *et al.* [3] was 31.8% vs 71.42% in our series.

Conclusion

Fibrolamellar hepatocellular carcinoma is different from traditional hepatocellular carcinoma and seems to have a better prognosis when taken care of early. The 5-year survival in our series was 71.42%.

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