

Diagnosis of patients with fibrolamellar carcinoma: a Dutch nationwide study

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Background & objective

Fibrolamellar carcinoma (FLC) is a rare primary liver cancer characterized by abundant eosinophilic cytoplasm and lamellar fibrotic bands. Adequate diagnosis is important for prognosis and treatment. The current study describes the diagnosis of fibrolamellar carcinoma in a Dutch historical cohort.

Methods

Adult patients diagnosed with FLC between 1990 and 2020, with pathology slides and clinical data available, were included through the Netherlands Cancer Registry and Automated National Pathological Anatomy Archive. Two expert hepatopathologists revised histopathology and immunohistochemistry (CD68 and CK7).

Results

In total, 52 adult patients, 25 (48%) male, diagnosed with FLC were included. Biopsies were available for 30 patients (58%) and resection specimens in 22 patients (42%). Upon expert review, in nine patients (17%) diagnosis FLC was unequivocally confirmed. Patients diagnosed with unequivocal FLC had a mean age of 26 years. Four additional lesions harbored characteristics of both FLC and conventional hepatocellular carcinoma (HCC). Three patients exhibited morphological features suggestive of FLC, yet with negative CD68 staining.

In the remaining 36 patients diagnosis was revised in cholangiocarcinoma (CCA, n=6, 12%), conventional HCC (n=25, 48%) or HCC/CCA (n=5). The lesions identified as conventional HCC were of steatohepatic (n=11), scirrhous (n=9), and chromophobe (n=5) subtypes.

Conclusion

The presence of fibrotic bands in steatohepatic and scirrhous HCC can lead to misdiagnosis of FLC as conventional HCC. This could have important treatment consequences as there is a tendency towards surgical treatment of FLC if feasible. Contrarily, evidence supporting the efficacy of systemic treatments for FLC remains limited. All in all, our Dutch historical cohort underlines the challenging diagnosis of FLC and emphasizes the critical role of expert review in accurate diagnosis.

Topic: Digestive Diseases Pathology - Liver/Pancreas