Intrahepatic Cholangiocarcinoma With Ductal Plate Malformation Pattern – A Case Report

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Case Report

A female in her fifth decade presented for further work-up of deranged liver function test results. Magnetic resonance imaging (MRI) of the liver showed a 25 x 24 mm subcapsular lesion in Segment 4b.

A segmental liver resection was performed. The macroscopic specimen contained a 25 x 22 mm lesion with vaguely demarcated outlines and a firm, white to pink cut surface.

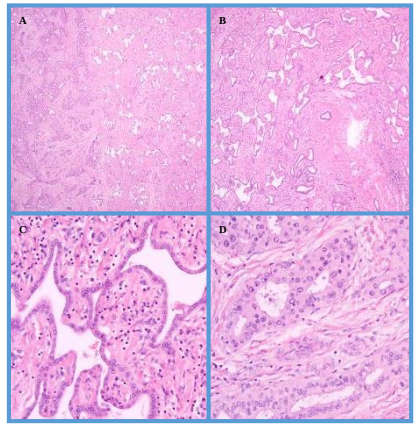

Figure 1 - The tumour contained areas of iCCA classic small duct type, and iCCA with DPM pattern (A. 20x) The DPM pattern included dilated tubules with polypoid projections (B. 40x and C. 200x). The classic small duct type showed greater cytological atypia (D. 200 x)

Sections showed a tumour with two distinct morphologies (Figure 1). Most of the tumour exhibited cribriform and tubular architecture, formed by cells with abundant dense eosinophilic cytoplasm, large rounded nuclei, coarse chromatin, and prominent nucleoli. Elsewhere, the tumour was comprised of dilated tubular structures with polypoid projections, lined by small cuboidal cells with relatively uniform rounded nuclei. These two tumour morphologies appeared to merge imperceptibly, and were reported as combined cholangiocarcinoma - cholangiocarcinoma with ductal plate malformation pattern.

Discussion

Intrahepatic cholangiocarcinoma (iCCA) can assume multiple histological appearances and, as such, a number of variants have been described. However, different terms have been used to describe similar lesions. The WHO Classification of Digestive System Tumours, 5th Edition (2019), appears to consolidate the classification of these lesions by dividing intrahepatic cholangiocarcinoma into small duct type and large duct type. Variants of small duct type include cholangiolocarcinoma and cholangiocarcinoma with ductal plate malformation pattern (Table 1).

The 5th Edition discourages the following terminology: cholangiocelluar carcinoma, cholangiolocellular carcinoma, and combined HCC-CCA with stem-cell features.

Table 1 - WHO Classification of Digestive System Tumours, 4th and 5th Editions

4th Edition (2010)	5th Edition (2019)
Intrahepatic cholangiocarcinoma (iCCA)	
Small duct	Small duct
No subtypes listed	Classic
	Cholangiolocarcinoma
	With ductal plate malformation
Large duct	Large duct
Combined hepatocellular carcinoma - cholangiocarcinoma	
With stem-cell features	*No subtypes listed*
Typical	
Transitional	
Cholangiocellular	
Cholangiolocellular	

iCCA - small duct type (classic)

iCCA small duct type tends to affect peripheral aspects of the liver. iCCA small duct type exhibits glandular architecture, formed by small to medium-sized cells with atypical but small rounded nuclei with inconspicuous nucleoli. Mucus secretion is usually absent.

iCCA - cholangiolocarcinoma

Cholangiolocarcinoma is characterised by malignant tubular structures with an arborizing or antler-like configuration. It shows a better overall survival and longer time to recurrence.

iCCA - with ductal plate malformation pattern

The ductal plate is an embryonic structure formed by a double layer of epithelial cells arranged as a ring around the portal tract blood vessels. It arises from a layer of hepatoblasts at the limiting plate, and undergoes remodelling to form the intrahepatic bile ducts. The histological manifestations of a ductal plate malformation are listed in Box 1. iCCA with ductal plate malformation pattern is a cholangiocarcinoma formed by malignant tubular structures with a resemblance to a ductal plate malformation.

Box 1 - Histological Features of Ductal Plate Malformation

- von Meyenberg complex
- Dilated duct containing bulbous papillary projection
- Circular lumen with a central fibrovascular core
- Multiple interrupted lumina scattered around a central fibrovascular core

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