

von Meyenburg Complex

Manoj Gopal Madakshira^a , Tushar Pandey^b , Ashim Das^a 

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A partial medical autopsy was carried out on a 50-year-old gentleman with a clinical diagnosis of acute severe necrotizing pancreatitis. The gross evaluation confirmed the presence of extensive necrotizing pancreatitis. However, the capsular and cut surface of the liver revealed the presence of subcapsular

small bile stained lesions measuring 1 to 3 mm in diameter (Figure 1a, b). On histology, these bile stained lesions were seen to represent micro-hamartomas composed of a collection of dilated varying sized ducts lined by cuboidal epithelium in a fibrotic stroma. The cells lining these ducts were positive for

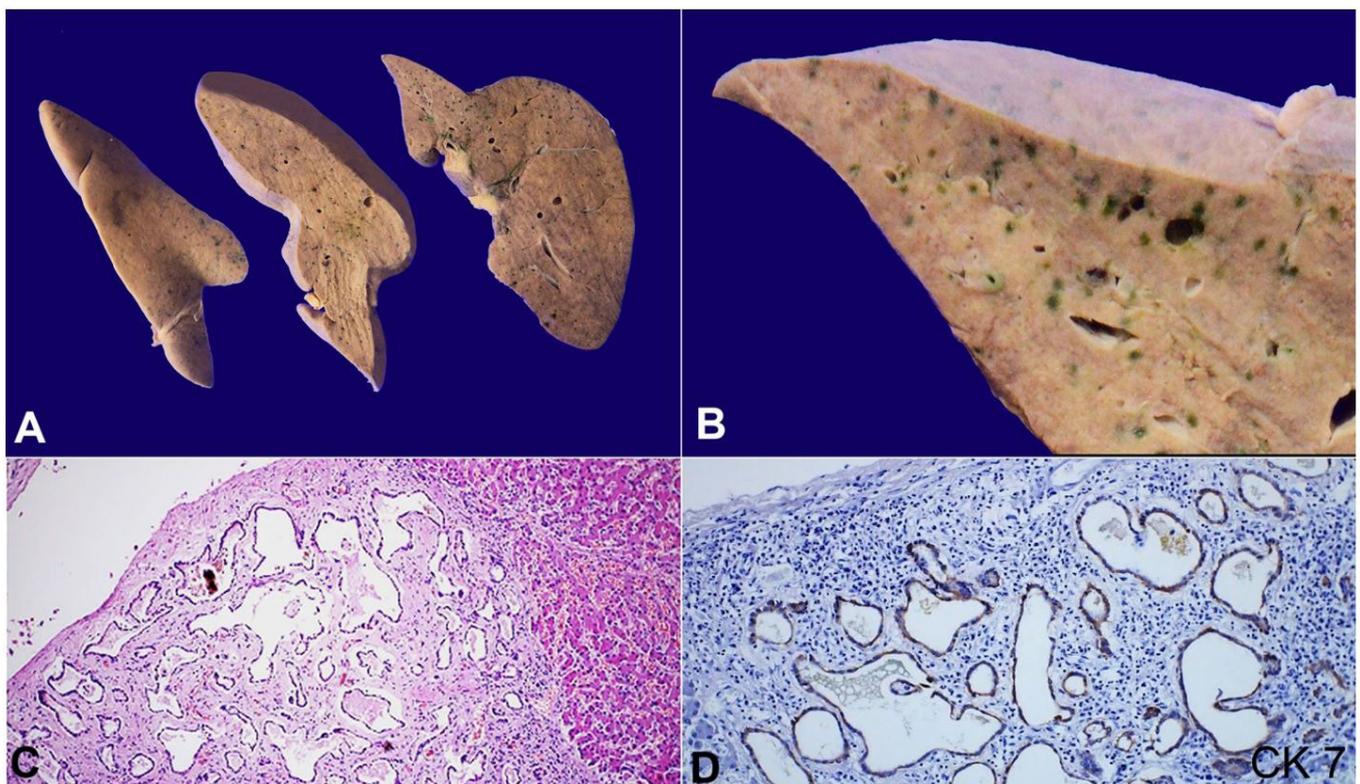


Figure 1. **A** and **B** - Cut surface of the liver show presence of multiple bile stained lesions measuring 3 to 15 mm in diameter in subcapsular location. **B** A closer view of the subcapsular bile stained lesions; **C** and **D** – Photomicrographs of the liver shows mis-shaped dilated bile ducts lined by cuboidal cells, embedded in a fibrotic stroma (H&E, 200X) and cuboidal cells lining the ducts show cytoplasmic staining for Cytokeratin 7.

^a Postgraduate Institute of Medical Education and Research, Department of Histopathology. Chandigarh, India

^b Postgraduate Institute of Medical Education and Research, Department of Pathology. Chandigarh, India.

Cytokeratin 7, confirming them to be of bile duct origin (Figure 1c, d). The subcapsular location of these lesions and characteristic histomorphology confirmed them to represent Von Meyenburg complexes (VMC).

VMC is a hamartomatous lesion and has been reported in 5.6% of autopsies.¹ These micro-hamartomas are a part of the spectrum of ductal plate malformations which also include congenital hepatic fibrosis, Caroli's disease, and adult polycystic disease.² Embryonic ductal plate develops as a rounded structure between the primitive hepatocytes and portal mesenchyme.³ The ductal plate malformation is a result of an aberrant persistence of the embryonic ductal plate at varying levels of the biliary tree – smaller interlobular bile ducts are affected in VMC, while larger interlobular bile ducts are affected in Caroli's disease.^{2, 4} The defect lies at the stage of morphogenesis of bile duct formation which has its molecular basis in ciliogenesis and cellular polarization orchestrated by hepatocyte nuclear factors 1 and 6 β .⁵

The importance of VMCs is in their simulation of neoplastic lesions. Utilizing the services of imaging and liver biopsy are essential to confirm the diagnosis of VMCs.⁶ There have also been anecdotal reports of malignant transformation of VMCs to cholangiocarcinoma. However, the low incidence of malignancy in case of VMCs suggest the importance played by secondary factors such as alcoholism, drugs, or infections in carcinogenesis.⁷

The authors retain an informed written consent for publication, and the manuscript is in accordance with the Institutional Ethics committee requirements.

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Correspondence

Prof Ashim Das

Post Graduate Institute of Medical Education and Research - Department of Histopathology

Research Block A Sector 12, Chandigarh, India

Pin Code: 160012

Phone: +91 9872223744

ashim126@gmail.com

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