CASE REPORT

Von Meyenburg complex and complete ductal plate malformation along with Klatskin tumour: a rare association

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SUMMARY

Von Meyenburg complexes (VMCs), or bile duct microhamartomas, are among the constellation of defects of ductal plate malformation. These present as multiple small intrahepatic cysts and are diagnosed incidentally. Association of intrahepatic VMCs with a bile duct cancer has rarely been reported. We describe a case of a 53-year-old man presenting with obstructive jaundice. Biochemistry and radiology gave a provisional diagnosis of a resectable Klatskin tumour. The patient underwent right hepatectomy with common bile duct and caudate lobe excision. The histopathological examination demonstrated intrahepatic VMCs with complete ductal malformation and malignancy at the hilum.

BACKGROUND

Von Meyenburg complexes (VMCs) are congenital malformations that occur because of ductal plate dysgenesis.¹ Their incidence is 5.6% in adults.² These lesions are typically composed of multiple cysts, each being less than 1.5 cm in size.³ They are incidentally detected in patients who undergo surgery for an unrelated condition.¹ When multiple, these cysts are usually mistaken for metastasis.^{4 5} Most of these lesions occur intrahepatically, however, extrahepatic microhamartomas have also been reported.⁶ We describe a rare case of VMC in a man who presented to us with features suggestive of Klatskin tumour. There was intrahepatic biliary hamartoma with complete ductal plate malformation and hilar adenocarcinoma on histopathological examination. This association has rarely been reported in the literature. The rare associations of these two entities make this case a unique presentation.

CASE PRESENTATION

A 53-year-old man, who neither smoked nor consumed alcohol, presented to our surgical outpatient department, with yellowish discolouration of the eyes and dark coloured urine for 1 month. He had experienced abdominal pain in the right upper quadrant for the past 2 weeks. This was associated with passage of clay coloured stools. There was no history of fever, oliguria, or loss of weight or appetite. On examination, the patient was deeply icteric. He had hepatomegaly 3 cm below the costal margin on palpation. Per rectal examination was unremarkable. He did not have any clinical findings

to suggest metastasis (ascites, left supraclavicular lymph node).

INVESTIGATIONS

Main biochemical findings were serum glutamic oxaloacetic transaminase-165 U/L, serum glutamic pyruvic transaminase-134 U/L, alkaline phosphatase-1033 U/L, total bilirubin/direct bilirubin—9.5/6.7 mg%, total protein/albumin—5.5/3.3 g% and carbohydrate antigen 19.9–4.3 IU (N=0–37 IU). Viral serology was negative. MR cholangiopancreatography (MRCP) imaging of this patient demonstrated dilated bilateral intrahepatic bile duct radicles with collapsed gallbladder. There was presence of a 2.5 by 2.0 cm heterogeneous lesion with altered intensity at the porta. The common bile duct was not visualised. The liver was normal on T1 and T2-weighted images. An impression of Klatskin tumour was rendered. Contrast-enhanced CT (CECT) scan of the abdomen with abdominal angiography confirmed the presence of ill-defined soft tissue with enhancement at the confluence of the right and the left hepatic ducts. Bilateral intrahepatic biliary radicles were dilated and the confluence was non patent ([figure 1](#)). There was no anatomical abnormality of the abdominal vasculature.

TREATMENT

With the impression of Klatskin tumour, the patient was prepared for right hepatectomy and caudate lobe excision. The left lobe of the liver was drained with a percutaneously placed biliary drain. With this evaluation, the patient underwent right hepatectomy with common bile duct excision, caudate lobe excision and left duct hepaticojejunostomy. Intraoperatively, there were no features of metastases; the gall bladder was not distended. There was a mass at the porta that was extending into the right duct. There was neither abnormal vasculature nor a biliary anatomy. On histopathological examination, gross examination of the liver was unremarkable except for the capsular congestion. There was a 1.2 cm by 1 cm whitish area at the porta hepatis. The gall bladder was grossly normal. On microscopic examination, multiple sections from the hilum revealed the proliferation of the biliary ductules with extensive fibrosis in and around areas. There was evidence of adenocarcinoma ([figure 2](#)) with perineural as well as intra-neural invasion ([figure 3](#)). There was evidence of extensive large bile duct obstruction changes in the

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Figure 1 Contrast-enhanced CT showing hilar block with proximal intra hepatic biliary radical dilatation.

liver. The liver showed maintained architecture. Extended portal tracts secondary to fibrosis were noted. Multiple VMCs (figure 4) with complete ductal plate dysgenesis were also seen (figure 5). The overall picture was suggestive of Klatskin tumour with sclerosing variant with free resection limits along with VMC and complete ductal plate malformation.

OUTCOME AND FOLLOW-UP

The patient had the expected postoperative course except for a small collection in the subhepatic space, which was drained percutaneously. He was discharged in satisfactory condition and was doing well on follow-up.

DISCUSSION

Von Meyenberg complexes were first reported in 1918. These lesions are detected incidentally during laparotomies for unrelated conditions or at autopsy.¹ VMCs are the most common variant of hepatic fibrocystic disease.⁶ They are among the constellation of defects of ductal plate dysgenesis that includes choledochal cyst, adult autosomal dominant polycystic liver disease, Caroli's disease and Caroli's syndrome.¹

VMCs are diagnosed both radiologically as well as pathologically. Ultrasound of the liver detects multiple small hypoechoic/hyperechoic foci distributed evenly throughout the liver. MRI reveals multiple small lesions that are hypointense on T1-weighted images and hyperintense on T2-weighted images.

Figure 2 Photo microphotograph of the cholangiocarcinoma.

Figure 3 Photo microphotograph of perineural invasion.

CECT shows small hypodense lesions usually distributed throughout the liver.^{3 5} Non-uniform and larger size of the liver metastasis on radiology can help differentiate VMCs from hamartomas.^{3 5} However, in this indexed case, radiology was not helpful in the diagnosis of VMC.

On histopathological examination, these complexes are whitish in colour and can vary from 1 mm to 1.5 cm in size. Microscopically, they consist of multiple, small to moderate sized ductules. These ductules are dilated and separated by fibrocollagenous stroma. The lining epithelium of these ductules is made up of flattened, cuboidal cells. They have well-circumscribed, round to oval nuclei. VMCs are present on the periphery of the portal tracts. They contain multiple cystic spaces filled with eosinophilic granules and inspissated bile that differentiates these lesions from normal bile ducts.^{3 7 8}

VMCs typically mimic intrahepatic metastasis. Their presence along with an extrahepatic malignancy pose a challenge to clinicians to prove these masses to be non-metastatic lesions.

Varying presentations of these malformations have been reported in the literature. These include incidental findings on abdomen ultrasound or features of infection and cholangitis.⁹ Cholangiocarcinoma developing from asymptomatic VMC has also been reported.¹⁰⁻¹² The malignancy arising from a long-standing VMC has been reported to be of low metastatic potential.⁹

Figure 4 Photo microphotograph of the von Meyenburg complex.

Figure 5 Photo microphotograph of complete ductal plate malformation.

The natural history of these hamartomas is scarce in the literature. Radiological examination along with histopathology of the lesion helps diagnose this rare entity.

The association of VMCs with bile duct cancer has been limited to a few case reports. On an extensive search of PubMed, this association was reported in 14 cases up to year 2004.¹³ VMC along with complete ductal malformation with a

bile duct cancer is still rarer. The associations of these pathologies provide an insight aiding the clinician to not misdiagnose these benign entities as intrahepatic metastasis when they are present on the surface of the liver. However, the liver surface was grossly normal in this index case.

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Learning points

- ▶ Von Meyenburg complexes (VMC) is a rare entity that is difficult to diagnose preoperatively.
- ▶ Association of VMC and bile duct cancer is yet rarer.
- ▶ VMC is essentially a histopathological diagnosis.
- ▶ A high index of suspicion is required for diagnosing VMC, as other conditions such as metastasis can mimic VMCs in clinical practice.

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