INTRODUCTION

Focal nodular hyperplasia (FNH) represents an uncommon benign liver tumour of well-circumscribed liver parenchyma. Focal nodular hyperplasia usually occurs in non-cirrhotic livers. A central fibrous scar with prominent arterial branches is typically observed at the histological examination [1]. The etiology of FNH is still unclear. In this report, we present the case of a 31-year old woman who developed multiple FNH within two years after insertion of a portocaval shunt. These findings are able to support the hypothesis that abnormal hepatic blood flow, especially reduced portal flow, is involved in the pathogenesis of FNH.

CASE REPORT

A 31-year old woman was admitted to our clinic for further diagnostic work-up of a large suspect tumour located in the right liver lobe. The lesion was an incidental finding. The patient presented with recurring nausea, vomiting and pain in the limbs. Furthermore, she had complained of constant night sweating for three years. There was no weight loss or fever. Her past medical history included a fibroadenoma of the right breast two years earlier and lactose intolerance. She did not take oral contraceptives.

Based on these findings we decided to perform an extended right hemihepatectomy. Previous embolization of the right portal vein was necessary in order to induce a hypertrophy of the small remnant left lateral liver segments (340 ml of total 2400 ml). This intervention led to the formation of a lefthesid portobiliary fistula with a consecutive thrombosis of the left portal vein. Subsequently, an emergency laparotomy became inevitable due to the significant deterioration of the
Patient’s general condition and increasing impairment of the liver function. Intraoperative findings evidenced a huge solitary tumour and the complete occlusion of the portal vein. Thrombectomy of the intrahepatic portion of the left portal vein was unsuccessful whereas the extrahepatic portal vein appeared to provide sufficient blood. Therefore, a portocaval shunt was performed after an extended right hemihepatectomy.

Histopathology revealed a benign angiomyolipoma, which was removed completely. Postoperative course was prolonged by liver insufficiency and hepatorenal syndrome. Furthermore, a leakage of the right hepatic duct was bridged with endoscopic stenting. Twenty days after surgery the patient was discharged in a good condition.

As a follow-up examination, the patient underwent abdominal sonography scans at intervals of six months after the extended right hemihepatectomy. Two years after surgery, multiple liver lesions were found (Fig. 2A,B). The biopsy obtained from one of the detected nodules located in liver segment II diagnosed FNH. In the further course, the patient was monitored conservatively with closed-meshed MRI. Two years later a slight increase in the size of the nodules was observed. As a consequence of this finding, the patient underwent a second biopsy, which did not reveal a clear diagnosis. Thus, we decided to perform an atypical liver resection in order to collect a decent tissue sample. The samples obtained from segment II and III unequivocally revealed FNH. There were no signs of malignancy or recurrence of angiomyolipoma.

In the follow-up examinations, the patient’s general condition and liver function (except for a slightly prolonged prothrombin time) were reported to be good. Two years after the second operation, the follow-up did not show any alterations or signs of malignant transformation.

**DISCUSSION**

This case report describes the development of multiple FNH lesions following extended liver resection and the creation of a portocaval shunt in one patient. Based on the observations of this case, we would like to reconsider the pathogenesis of FNH. According to the literature, there is a strong relationship between altered hepatic circulation and the development of benign liver lesions. More specifically, radiological and histological investigations of FNH have displayed an imbalance of arterial and portal bloodstream with an enhanced arterial blood supply [1-3]. Other studies claim that hyperplastic nodules are the result of a physiological reaction to tissue or vascular injury [3-7]. Kumagai et al described FNH as a response to acquired arterial and/or venous thrombosis [8]. The consumption of birth control pills has not shown any clear association with FNH [1, 9] in contrast to adenomas, where an association has been evident [10].

The portal blood flow might represent a gate to numerous important hormones and transmitters originating from the...
gastrointestinal system. Therefore, the absence or even the reduction of this flow may result in alterations of the hepatic cell function, structure and regeneration [11, 12]. The observation that common hepatic tumors are associated with congenital absence of the portal vein (CAPV) [13] is able to support this hypothesis and also the results of the present study.

Several investigations in animal models established a possible causal connection between the occurrence of hyperplastic liver lesions and portosystemic shunts [14-17]. Based on histopathological investigations implemented by Rasenack et al [15] it was found that a complete portocaval shunt initially leads to focal nodular necrosis, followed by epithelial proliferation and finally leading to the formation of hepatic nodules. The observed effects turned out to be even stronger after simultaneous liver resection [17]. Moreover, there is evidence that modified portocaval shunts, with preserved pancreatic blood flow into the liver, do not lead to FNH [14, 15]. In this present case the portal-venous blood stream towards the liver was completely interrupted. This finding supports the hypothesis that factors from the gastrointestinal system, especially pancreatic hormones, transported via portal blood stream maintain function and regeneration of the liver parenchyma.

To date, there is very little information available regarding the association of liver tumours and acquired complete portocaval shunts in humans. Fukushima et al [18] reported a male patient developing multiple benign liver tumours highly suggestive of FNH after portocaval shunting in his childhood. However, in this special case, benign liver tumours developed more than 30 years after the portocaval shunting. Female gender and corresponding hormones may have led to an even more rapid nodular growth in our case. There are other studies reporting hyperplastic nodules after portocaval shunting in patients with glycogen storage disease-I, a disease which is normally associated with adenomas or malignant tumours [19, 20]. In our case, the surrounding liver parenchyma did not reveal any pathological findings. There were no signs of liver cirrhosis or evidence of any other underlying metabolic disease. Pre-existing vascular malformation, which might have been responsible for the induction of angiomyolipoma, could be excluded. Furthermore, the rare case of malignant changes in angiomyolipoma as described by Nguyen et al [21] was not evident. The lesions found in the present case could be clearly identified as FNH, excluding the possibility of the recurrence of angiomyolipoma.

CONCLUSION

This case report supports the hypothesis that portosystemic shunt and consecutive loss of direct hepatic blood flow might promote the formation of FNH. Since the differentiation of intrahepatic nodules is not possible in all cases by imaging only, biopsies should be performed in order to be able to identify potential malignancy at early stages. Therefore, it is compulsory to adhere to careful and frequent follow-up regimes.

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REFERENCES


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