CASE REPORT

A liver lesion in primary sclerosing cholangitis

Sujata Biswas,¹ Neil Rajoriya,¹ Lai Mun Wang,² Jane Collier¹¹Translational Gastroenterology Unit, John Radcliffe Hospital, Oxford, UK²Department of Cellular Pathology, John Radcliffe Hospital, Oxford, UK**Correspondence to**Dr Sujata Biswas,
sujatabiswas07@gmail.com

Accepted 9 March 2015

SUMMARY

Primary sclerosing cholangitis is associated with an increased risk of cholangiocarcinoma; appropriate surveillance should be undertaken for this. Our case illustrates an unexpected liver lesion in such a patient, which would not have been detected with imaging alone. In suspected cancer, if a patient is not a candidate for liver resection, liver biopsy should be considered in case there is a treatment option for an alternative cause of the liver lesion. This is a careful decision to be made due to the risk of tumour seeding compromising cure and needs to be discussed via the hepatobiliary multidisciplinary team.

BACKGROUND

Primary sclerosing cholangitis (PSC) is associated with an increased risk of cholangiocarcinoma; appropriate surveillance should be undertaken for this. We present a case of a liver lesion, found on MRI, that was presumed to be cholangiocarcinoma, but which at resection was found to be an unexpected liver lesion.

If a patient is not a candidate for liver resection and has a malignant-appearing liver lesion on imaging, biopsy should be considered. This may yield an alternative liver pathology to cholangiocarcinoma, which could result in a different management plan.

CASE PRESENTATION

A 59-year-old man with PSC underwent a routine MR cholangiopancreatogram (MRCP). His medical history included colectomy with ileoanal pouch formation for ulcerative colitis, cholecystectomy and haemophilia C. His only medication was ursodeoxycholic acid 600 mg two times a day.

An MRCP 2 years previously showed extensive PSC with a dominant stricture of the distal common bile duct. There had been no interval change from a prior MRCP at that time, and normal CA19-9, so the decision had been made to monitor his progress with serial imaging.

INVESTIGATIONS

The MRCP showed no interval change in the intrahepatic duct beading and stricturing; however, there was a new 2 cm T2 hyperintense lesion in segment VII (figure 1). CA19-9 was again normal at 15 U/mL. Alkaline phosphatase was stable at 1043 IU/L and bilirubin 18 µmol/L. Platelet count was 305×10^9 L, prothrombin time 13.2 s and albumin was 40 g/L.

Figure 1 Coronal MRI.

TREATMENT

A presumed diagnosis of cholangiocarcinoma was made and the patient was discussed in the HPB (hepatopancreatobiliary) multidisciplinary meeting regarding the options of surgical resection, transplant assessment or biopsy. Staging CT scan showed no extrahepatic lesions. Alternative causes of the liver lesion such as hepatocellular carcinoma or hypervascular metastasis were considered but deemed to be unlikely given the context.

On the multidisciplinary team's recommendation the patient underwent surgical resection. Portal pressure measurements were made to assess the degree of portal hypertension prior to surgery; hepatic venous wedged pressure was raised at 20 mm Hg. The patient underwent open liver resection of segments VI and VII. The histology is shown in figure 2.

OUTCOME AND FOLLOW-UP

Histology surprisingly showed malignant melanoma. The background liver was consistent with PSC. On further examination, the patient had multiple cutaneous naevi with a very dark 5 mm lesion on the calf, which had been present for several

Figure 2 Liver histology.

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To cite: Biswas S, Rajoriya N, Wang LM, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2014-207685

years and were unchanged. These were not thought to be primary melanomas and skin biopsies were benign. Staging CT scans showed no metastases elsewhere. Six months on from surgery the patient is well and has had no adjuvant therapy.

DISCUSSION

It is not clear whether the liver lesion was a primary or secondary melanoma, but given the absence of other lesions it seemed likely to be primary. Primary liver melanoma has been described only in case reports¹ and to the best of our knowledge it has not been reported in PSC. The case reports similarly report no extra-hepatic lesions and the optimal management has not been established given its rarity.

In all such cases, surgery should be considered via the multidisciplinary meeting as per national guidelines² before biopsy to avoid the potential risk of tumour seeding. Biopsy of a malignant liver lesion could compromise the long-term survival of a patient if the aim is for curative resection, ablation or transplant.

In this situation, however, the diagnosis of melanoma would not have been made if the patient had not been a surgical candidate and had not undergone biopsy. It is worth considering the role of biopsy in patients who are not fit for surgical resection in case there is an alternative diagnosis to cholangiocarcinoma. Liver melanoma is rare, but if risk factors are present or skin lesions are noticed, the diagnosis of liver melanoma should be considered. Biopsy can help in confirming the nature of the lesion and guiding chemotherapy if curative treatments cannot be undertaken.

Learning points

- ▶ Primary sclerosing cholangitis carries an increased risk of cholangiocarcinoma and patients should undergo regular surveillance imaging.
- ▶ If a liver lesion is found on imaging, HPB (hepaticopancreaticobiliary) multidisciplinary discussion should take place as to whether resection is appropriate.
- ▶ The point that this case illustrates is that if the patient is not a surgical candidate, liver biopsy should be considered to exclude other causes of liver lesions that may be amenable to alternative treatment.

Contributors SB wrote the article. LMW supplied the histopathological images. NR and JC revised the article.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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