A case of painless jaundice

A.A. Alkhatib¹*, J.M. Collins², A.A. Moss³, A. McCullough⁴, M.E. Harrison¹

Departments of 'Medicine, Division of Gastroenterology, ²Radiology, ³Surgery, and ⁴Pathology, Mayo Clinic, Scottsdale, Arizona, USA, *corresponding author: tel.: 801-590-8736, e-mail: khatibamer@yahoo.com

CASE REPORT

A 64-year-old geologist with Crohn's disease presented with fatigue, anorexia, nausea and painless jaundice. His serum carcinoembryonic antigen was 7.2 ng/ml (normal o-3 ng/ml) and the calcium 19-9 was 965 U/ml (normal <55 U/ml). Ultrasound and computed tomography (CT) scan of the abdomen revealed diffusely dilated bile ducts, with a 3 cm filling defect distending the distal common bile duct (CBD) (*figure 1*). Endoscopic retrograde cholangio-pancreatography (ERCP) showed a large filling defect in the distal CBD (*figure 2*). Biliary brushings and biopsies revealed large oval to polygonally shaped nuclei with very prominent nucleoli, surrounded by attenuated cytoplasm.

Figure 1. Imaging studies including right upper quadrant ultrasound, CT of the abdomen and MRI of the abdomen showing a mass within the common bile duct





WHAT IS YOUR DIAGNOSIS?

See page 418 for the answer to this photo quiz.

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ANSWER TO PHOTO QUIZ (PAGE 414) A CASE OF PAINLESS JAUNDICE

DIAGNOSIS

Primary malignant melanoma was diagnosed by cells staining positive for S100 and HMB45, and negative for pancytokeratin, hepatocyte-specific antigen, CK7, and CD68 (*figure 3*). BRAF mutation was detected with V600E mutation.

Further evaluation including magnetic resonance imaging of the abdomen did not detect melanoma distinct from the common bill duct (CBD) mass (*figure 1*). Dermatological and ophthalmological evaluation showed no evidence of cutaneous or ocular melanoma. MRI of the brain and abdomen, CT of the chest, abdomen and pelvis and CT PET scan showed no other lesions.

The patient underwent Whipple surgery in which the melanoma was seen to be confined to the common biliary duct (*figure 4*). The patient recovered uneventfully.

The majority of melanomas originate from the skin. However, a minority arise from other noncutaneous organs including uvea and the retina.^T Whenever melanoma is encountered in the bile duct, it usually presents as secondary melanoma.^{Tr2} Nonetheless, ten cases of primary melanoma of the bile duct have been reported.^{Tr4} Primary melanoma of the bile duct originates from the melanocytes that exist in the biliary system. This was shown by

Figure 3. Haematoxylin and eosin showing melanoma, ductal epithelium and pigment



Magnification is x 200 in main picture and x 600 in inset.

Figure 4. Gross pathology of the melanoma within the common bile duct



Ricci *et al.* who reported a case of primary melanoma of the gallbladder. In this case, they found ultrastructural evidence of two distinct benign and malignant melanocyte populations.⁴ The majority of the ten described cases of primary melanoma of the bile duct have been reported in males with an 8:2 male to female ratio. The ages of the patients range from 30 to 58 years with the majority in their fourth or fifth decades. Most of the cases present as a solitary polypoid lesion. Outcomes of primary melanoma of the bile ducts are variable. Half of reported cases of primary biliary melanoma underwent Whipple surgery. Three of these patients were disease-free after ten months, including one who was disease-free six years after Whipple surgery.¹⁻⁴

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