Malignant masquerade at the hepatic hilum

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A 69-year-old lady presented with fever with chills and rigors and surgical obstructive jaundice of 1 days duration. Her surgical history was significant for open cholecystectomy and choledocholithotomy. Also she received anti tubercular treatment for intestinal tuberculosis. Physical examination revealed icterus and a healed right subcostal scar, and laboratory values showed obstructive jaundice. Computed tomography revealed dilated intrahepatic biliary radicals (IHBRD) with a soft tissue mass at the hepatic duct confluence with mild thickening of the hepatic ducts and proximal CBD with significant narrowing of proximal CBD. The Soft main portal vein (MPV) and encasing the right hepatic artery (RHA) tissue mass was abutting the right portal vein (RPV) and MPV and encasing RHA (Fig. 1) and MRCP showed type 4 biliary block and bilobar IHBRD

What is the Diagnosis?

- A. Hilar cholangiocarcinoma
- B. Post cholecystectomy benign biliary stricture
- C. Carcinoma gall bladder with obstructive jaundice
- D. Tubercular stricture of CBD

Strictures at the hepatic hilum do not always represent malignancy. Eight to 13% of patients with proximal biliary strictures are benign following histology of the resected specimen [1]. Hadjis et al [2] coined the term ‘Malignant masquerade’ to emphasize that hilar stricture whether benign or malignant may not be distinguishable based on the clinical and radiological features. Management decisions are generally based on imaging, as biopsy is generally

non-diagnostic. Nakayama et al [1] reported a series of 14 benign proximal bile duct strictures who had fibrosis of the bile duct with an inflammatory infiltrate suggesting that its a spectrum of disease with some cases of idiopathic fibrosis and others a *form fruste* of sclerosing cholangitis. The differentials include granulomas, benign idiopathic stenosis, cholangiomitosis, Mirizzi’s syndrome. As some of these cases require endoprosthesis or palliative surgery without histology there’s a risk of mismanagement of such patients based on wrong diagnosis. Based on imaging, a lengthy segment of extrahepatic bile duct stricture with irregular margins and asymmetric narrowing suggests cholangiocarcinoma whereas a short segment with regular margin and symmetric narrowing suggests benign cause [3].

This patient represents an unusual example of malignant masquerade (Fig. 2). She underwent bile duct excision followed by reconstruction of biliary tree (roux-en-y hepaticojejunostomy). The histopathology showed diffuse fibrosis with no evidence of malignancy or granulomas.

Fig. 1 CT scan showing soft tissue mass in porta extending into left hepatic duct with IHBRD

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Fig. 2 Thick walled dilated bile duct adherent to RHA

In conclusion, all proximal strictures with sectoral duct separation may not be malignant. A careful reassessment based on clinical and radiological findings should be done before contemplating major surgery. However, in the case of a high-grade biliary stricture surgical exploration will still be required.

References

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