strongly positive in 90 - 100% of cases reported to be ES/PNETs. All six reported cases of gastric ES/PNETs also revealed the diagnostic significance of CD99. Vimentin which is an intermediate filament that is present in most mesenchymal cell is also usually positive in ES/PNET cells. The six reported cases of gastric ES/PNETs also showed positive vimentin expression in tumor cells. In ES/PNET, various neural markers such as S100, chromogranin A, synaptophysin and neuron-specific enolase (NSE) shows variable immunohistochemical staining which may be due to the differing degrees of neuroectodermal differentiation in the individual tumors.

Surgical resection was possible in all six reported cases of gastric ES/PNETs. The current standard chemotherapeutical treatment for ES/PNETs is a regimen comprising the drugs: vincristine, doxorubicin, cyclophosphamide, ifosfamid and etoposide. Chemotherapy has a curative role in the management of localized ES/PNET but its role in metastatic disease is limited. Combined modality treatment comprising of surgery, multi agent chemotherapy and radiation therapy improves survival in metastatic ES/ PNET.

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Mucin hypersecreting bile duct neoplasm

Papillary growth can occur anywhere along the biliary tree. It can be anything from low-grade dysplasia to invasive carcinoma pathologically. These growths have been given various names such as biliary papillomatosis, mucin-producing cholangiocarcinoma, mucin-hypersecreting bile duct tumour, and biliary intraductal papillary mucinous neoplasm. They show a more favourable prognosis compared to non-papillary cholangiocarcinoma. The clinical entity intraductal papillary neoplasm of the bile duct (IPNB) was added to the 2010 World Health Organization (WHO) classification, and it includes intraductal papillary cholangiocarcinoma and its precursor lesions.

Case Report

A 62 years old gentleman presented with history of progressive jaundice for 4 months, associated with weight loss (18 kg in 4 months) and pruritus. Initial laboratory values included Hb of 10.9 g/dL, WBC of 4200/μL, albumin of 3.9 g/dL, total bilirubin of 10.3 mg/dL, direct bilirubin 8.2 mg/dL, AST/ALT of 30/19, alkaline phosphatase of 340 IU/L and CA 19-9 of 225. CECT abdomen and MRCP revealed intrahepatic biliary radicle dilatation (IHBRD) with heterogeneous enhancing soft tissue density at hilum extending along right and left
hepatic duct till the ostium of segmental duct, suggestive of cholangiocarcinoma type IV. The lesion was causing indentation of left portal vein. No intrahepatic or extrahepatic metastases were found. (Figure 1 and 2)

Left hepatectomy including caudate lobectomy with excision of extrahepatic biliary apparatus was done. Exploration did not reveal ascites or peritoneal metastatic nodule. The intraductal tumour in the common hepatic duct was found to be extending above bifurcation of hepatic ducts. Mucinous secretion were noted from tumour. The patient showed no postoperative complication and was discharged on POD 9.

The histopathological examination grossly revealed a intra luminal mucinous tumor measuring 7x2x2 cms in left hepatic duct. Cut surface revealed a mucinous jelly type tumor. Microscopic examination revealed tumour cells arranged in tubular glands and small nests floating in pool of mucin. (Figure 3) Intracytoplasmic mucin also seen. No lymphovascular or perineural invasion was seen. Resection margin of bile duct and hepatic parenchyma were found to be free of tumour cells.

Immunohistochemistry revealed tumour cells were focally positive for CK7 and negative for CK20/p53. CK19 was expressed diffusely and showed both cytoplasmic and membranous staining. Mib 1 proliferative index was 50% in the hotspots.

Patient is on regular followup on an out-patient basis with no evidence of recurrence.

Discussion

A bile duct neoplasm can be of flat or papillary type, the latter being called as IPMN-B. It is regarded as a counterpart of pancreatic intraductal papillary mucinous neoplasm (IPMN) as reported in various studies; they both arise and grow predominantly in ducts and have
papillary appearance. The difference between the two lies in respect to mucin secretion which is common in IPMN but is found in only one third of IPMN-B cases.\(^2,3\)

IPMN-B can be graded based on cholangiography and the management protocol is decided accordingly. Our case was cholangiographic type IIB (intrahepatic polypoid or cystic neoplasia with involvement of extrahepatic bile duct); in this situation, an aggressive resection is advised.\(^4\)

The survival of patients with IPNB reduces with progression from low-grade dysplasia to invasive carcinoma. The recurrence rate at 5 years in benign IPNBs has been reported to be nearly 20%; this rises to 60% in malignant cases. Most recurrences are locoregional.\(^5\)

Curative resection is the major treatment and bodes well for long-term survival, especially in patients with early-stage IPNB.

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