A 50-year-old man was admitted on 2nd February 2005 with pain in the right middle and upper abdomen since 6 months and progressive jaundice since 3 months. The pain was moderate intensity, intermittent, occurring 2-3 times per day, and radiating to the back. The intensity had increased in the last 3 months and became continuous since 10 days. Jaundice was associated with high colored urine, clay colored stool and generalized pruritus. There was loss of appetite associated with unquantified weight loss. There was no history of fever, bleeding, abdominal distension or features suggestive of gastric outlet obstruction. Past history was not significant. He was a chronic smoker, smoking 2 bundles of beedi per day for the last 40 years, and consumed 250 mL country liquor, once in 3 to 4 days.

At admission, he was afebrile, conscious, oriented, and emaciated (BMI 15.33 Kg/m²). Pulse rate was 72 per minute, regular; blood pressure 112/70 mm Hg, and respiratory rate 18 per minute. He had mild pal (lor, deep icter (us, and had bilaterally enlarged axillary and inguinal lymph nodes (<2 cm size). There were generalized excoriation marks on the skin. The liver was palpable 5 cm below right costal margin, firm with a smooth surface. In addition, there was a globular, firm and nontender lump in the right hypochondrium, which moved on respiration, possibly a gall bladder (GB) lump. There was no free fluid. Bowel sounds were normal. Breath sounds were vesicular with prolonged expiration and crepitations in both infrascapular areas. Cardiovascular system and central nervous system examination was within normal limit.

Investigations

Peripheral blood film showed normochromic normocytic picture. Prothrombin time and serum amylase were normal. Arterial blood gas on 19th February: pH - 7.41, PaO₂ - 63, PaCO₂ - 28, HCO₃ - 17, SaO₂ - 93%. ELISA test for HIV was negative. Fine needle aspiration cytology from left axillary lymph node showed reactive lymphoid hyperplasia.

Radiology

Ultrasound examination showed dilated intrahepatic biliary radicles; the common bile duct (CBD) was dilated till the lower end with abrupt cut off. Liver, GB and pancreas were normal. Biphasic computerized tomography scanning was done on day 5 of hospital admission. The non-contrast study was normal. Arterial phase showed bilobar intrahepatic biliary radicles dilated with patent confluence of the hepatic ducts. There was dilatation of the suprapancreatic portion of bile duct with an abrupt narrowing at the lower end, and loss of fat planes (Fig 1). The GB was grossly distended. There was a homogenous soft tissue mass in the pre caval region (suspected to be lymph nodes). Few small lymph nodes were present around the superior mesenteric and abdominal aorta. Accessory right hepatic artery was identified. Pancreas was normal with normal vascular-
Clinico-pathologic Conference

Indian Journal of Gastroenterology 2007 Vol 26 May - June  131

ity. Pancreatic duct throughout its length showed mild dilatation. Venous phase revealed normal venous circulation. At the level of ampulla of Vater, a polypoidal structure was seen projecting into the duodenal lumen; the GB wall was not thickened (Fig 2). There was no free fluid. Rest of the abdominal organs were normal. Aorta showed atherosclerotic plaques. Impression – i) Extrahepatic biliary obstruction at suprapancreatic CBD, by lymphadenopathy or stricture. ii) Periampullary malignant growth either cholangiocarcinoma or ampullary cancer.

Course and management

The patient was initially treated with antihistaminics, ursodeoxycholic acid and multivitamins. ERCP (17-2-05) was done under antibiotic cover Papilla was bulky. After deroofing the papilla by precut papillotomy, a mass was visualized. Repeated attempts at cannulation of CBD failed. Next day he developed abdominal pain. X-ray abdomen did not show air under the diaphragm and serum amylase was normal. A day later he developed hypotension and altered sensorium with fever. Blood examination showed elevated TLC and hypokalemia. He required ionotropic support for hypotension. On February 19, percutaneous transhepatic biliary drainage (PTBD) was done; cytologic examination of bile revealed 440 pus cells. Blood culture grew *E. coli*, sensitive to amikacin, gentamycin and ceftazoxime; antibiotics were changed to amikacin and cefipime. His condition continued to deteriorate and he died on 20th February.

Treating unit’s diagnosis - Extrahepatic biliary obstruction due to carcinoma of pancreas or GB with cholangitis and septic shock.

Discussion

This 50-year-old male presented with pain abdomen, cholestatic jaundice and constitutional symptoms, had lymphadenopathy with evidence of lower CBD obstruction, palpable GB and ampullary tumor as seen on endoscopy. Terminally he probably had severe cholangitis probably following failed endoscopic intervention. The differential diagnosis that could be considered are as follows:

Causes of extrahepatic biliary obstruction (EHBO) include benign causes like CBD stone, post operative stricture, chronic pancreatitis, primary sclerosing cholangitis, hydatid cyst, liver abscess and rarely tuberculosis (TB) of bile duct. Malignant causes include carcinoma gall bladder (Ca GB), cholangio-carcinoma (CCA), carcinoma pancreas (Ca Pan), periampullary carcinoma, metastasis and uncommonly hepatocellular carcinoma (HCC). In studies from Delhi on clinical profiles of patients with EHBO, one-third had benign etiology; malignancy was seen mainly in elderly and etiologies included carcinoma in relates to GB , pancreas, periampullary and biliary tree.1,2 Besides age, the clinical features favoring malignancy were severe constitutional symptoms, ascites, GI bleeding and gastric outlet obstruction. Fluctuating jaundice, mild cholangitis and a well-preserved patient favored benign cause. Based on these clinical data, the index patient most likely had malignant cause for EHBO. There was dilation of biliary tree and pancreatic duct with a polypoid growth at ampulla on endoscopy.

**Periampullary carcinoma**

The index patient had a tumor at the ampulla. Majority of periampullary carcinomas originate from the pancreatic duct, followed by CBD, duodenum and ampulla.4 Clinically it is difficult to localize the origin of tumor as the clinical symptoms and signs are similar, but a bulky ampulla associated with abrupt cut off of CBD on radiology would suggest that the origin of tumor is either pancreatic duct or CBD. In one study, all these tumors were adenocarcinomas.5 In a recent study on surgical specimens of periampullary tumors, adenocarcinomas were seen in older patients with shorter disease duration, higher bilirubin, and infiltrative but smaller tumor.5 The index patient was an elderly man with very high bilirubin and a relatively small tumor, thereby favoring a diagnosis of adenocarcinoma. Ampullary tumors could be adenoma, gastrointestinal stromal tumors, neuroendocrine tumors, hamartoma, lipoma, paraganglioma and adenomyoma.

**Metastatic adenocarcinoma**

Malignant tumors could also be a metastatic adenocarcinoma, and rarely lymphoma. Usually a metastatic biliary obstruction results in upper CBD
obstruction at the region of porta. Common primary foci are breast, colon, lung, ovaries, endometrium and melanoma. Similar presentation could also be seen in Hodgkin’s and non Hodgkin’s lymphomas (NHL). The index patients also had peripheral lymphadenopathy, and NHL remains one possible condition. Lymphoma usually does not produce biliary obstruction. In a study of 863 patients with NHL, only 0.5% patients presented with jaundice and 0.8% had biliary obstruction.6

Tuberculosis

Tuberculous lymph node resulting in biliary obstruction could mimic malignancy. Pancreatic tuberculosis constitutes 0- 4.7% of abdominal TB, and can result in lower CBD obstruction.3

Final Diagnosis

Extrahepatic biliary obstruction due to periampullary tumor, possibly adenocarcinoma, lymphoma, orcarcinoid, with severe cholangitis. Cause of death - septic shock.

Pathology protocol

Partial autopsy was done. The peritoneal cavity contained minimal amount of exudates but with sizable amount of blood clots. Deep bile staining of all the organs was noted. Liver (weight - 1600 gms) was enlarged, and soft. The GB was grossly dilated and firmly adherent to the liver bed. The lesser omentum was pulled up towards the hilum of liver forming a mass with the surrounding soft tissue (Fig 3). The duodenal loop and head of pancreas were adherent firmly to each other and surrounding fat. Ampulla of Vater was patulous and indurated. Only the terminal one cm of CBD could be probed. A firm-to-hard ill-defined mass could be felt on the medial aspect of duodenum in the region of the head of pancreas measuring ~ 2 cm. Rest of pancreas appeared normal. Pancreatic duct was not dilated. Multiple whitish ill-defined firm nodules were identifiable at porta and around biliary tree. Suprapancreatic portion of CBD was grossly thickened and dilated measuring 15 mm in diameter. The duct abruptly became narrowed and occluded, merging with the hard nodular area within the head of pancreas. There was dilatation of the intra-hepatic biliary tree with cavitation, some filled with dark bloody necrotic material. The portal and hepatic veins, and inferior vena cava were normal. The grossly dilated GB appeared blackish, nodular with irregular thickening of the wall and loss of normal mucosal folds; histology revealed features of moderate to poorly differentiated adenocarcinoma with marked desmoplasia (Fig 4), diffusely involving the whole length of CBD extending into the ampulla through the duodenal wall (Fig 5), involving the peripancreatic fat, wall of hepatic artery and portal vein. Peripancreatic lymph nodes were free of tumor. The tumor cells were seen extending along the CBD into the proximal portion of the intrahepatic ductal system, and along the cystic duct involving the GB transmurally (Fig 6) with infiltration to the liver bed. Sections of the liver including the dilated ducts showed fibrosis, ulceration and necrotic debris within the lumen. There was extensive cholestasis with bile lake formation. Duodenal mucosa at periampullary region showed two distinct areas of deep mucosal erosions (Fig 3). Dissecting into the site of the ero-
sions revealed a 15 mm long dark grey colored stent embedded within the duodenal wall extending into the adjoining pancreatic parenchyma. There was no inflammatory cell response on histology.

Stomach showed marked mucosal congestion along distal body mucosa with diffuse mucosal granularity; at histology, the congested areas showed intra-mucosal hemorrhages and chronic gastritis with intestinal metaplasia, but no *H. pylori*. Small and large intestine showed patches mucosal congestion with fresh intramucosal hemorrhages. The lungs (weight 920 gm) were sub-crepitant with congested tracheo-bronchial tree; microscopy showed interstitial fibrosis, numerous type 2 pneumocytes with abundant pigment, some of which was hemosiderin pigment. Hilar and carinal lymph nodes were enlarged, and showed reactive hyperplasia. Heart (weight – 280 gm) showed mild reduction in epicardial fat with left ventricular hypertrophy. Aorta had grade III atherosclerosis. Spleen and kidneys were within normal limits.

**Final autopsy diagnosis**

1. Diffuse poorly differentiated carcinoma of extrahepatic biliary system and GB with extension to hilum and proximal intrahepatic biliary system.
2. Patchy confluent hepatic necrosis (patchy hepatic outflow tract obstruction) and intrahepatic tumor metastasis.
3. Interstitial pulmonary fibrosis, left ventricular hypertrophy, Grade III atherosclerosis.
4. Chronic gastritis with intestinal metaplasia and fresh mucosal hemorrhages in gastrointestinal tract.

**Comments**

The CT and endoscopy findings would favor a tumor in ampullary region. The clinical features were not exclusive either for Ca GB or CBD. There was no stone or wall thickening of GB which are the frequent associated features. In Ca CBD, the upper segment is the common site though the lower segment could also be involved. At autopsy, there was diffuse involvement of the whole length of CBD, which is rare. There was gross thickening of CBD wall; this could be the result of tumor infiltration from outside rather than from inside the biliary tract. It is possible that the lower stricturous portion of CBD was the primary site, progressing to involve the proximal CBD and GB. There is record of only one such case in the literature where Ca CBD was seen in continuity with GB. Besides, the disproportionately thickened CBD wall could suggest a pre-existing sclerosing cholangitis, which would be more susceptible to cancer development. Needle aspiration or mucosal biopsy during ERCP could be rewarding in such a setting.

ERCP resulted in cholangitis, and possibly bleeding as there was significant drop in the hemoglobin level. Following septicemia, the patient could have developed disseminated intravascular coagulation as indicated by the low platelet count and a prolonged PTTK.

**References**

3. Desai CS, Lala M, Joshi A, Abraham P, Desai D, Deshpande...

The case was discussed in the staff clinicopathological forum of PGIMER, Chandigarh. The clinical protocol was presented by A Duseja, radiology by N Kalra and pathology by K Vaiphei. The session was chaired by V Sakhuja. Others who took part in the discussion – R Kochhar, V Singh, A Rajwanshi, GR Verma, P Malhotra, G Choudhuri (Lucknow), TDS Yadav and S Jain. The case was compiled by K Vaiphei.

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Image

Preoperative demonstration of broncho-esophageal fistula by multidetector computed tomography

A 25-year-old lady presented with dysphagia and cough exacerbated by drinking. She had intermittent low-grade fever and weight loss since 4 years. Barium swallow was normal. Fiberoptic bronchoscopy revealed profuse secretions from the right lower lobe bronchus.

Dynamic CT scan (Brilliance 40-slide, Phillips), with the patient in the supine and prone positions while swallowing oral contrast, revealed dilated bronchi in the lower lobe opacified with contrast. A fistulous tract was seen from the right posterolateral wall of the esophagus towards the right lower lobe (Fig 1), better seen in multiplanar reconstructed images (Fig 2). The patient subsequently underwent surgery. The secretions through the intercostals drainage were positive for Gram-negative bacilli.

Broncho-esophageal fistula in adults is most often due to cancer or tuberculosis.1 The fistula site and size as well as lung involvement need to be demonstrated preoperatively. 2 As compared with esophagography and conventional CT, the faster speed, high spatial resolution, greater anatomic coverage area, and multiplanar and 3-dimensional imaging capability make multidetector computed tomography a useful diagnostic tool for tracheo/broncho/esophageal fistula and associated lung changes. 2

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References

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Fig 1: CT axial image shows contrast-containing fistula (black arrowhead) between right lateral wall of esophagus and right lower lobe bronchus. Esophagus is shifted towards right side due to fibrosis. Area of consolidation is seen in right lower lobe.

Fig 2: CT with coronal oblique reformation of airways shows contrast-containing fistula (arrows) between right lateral wall of esophagus and right lower lobe bronchus. Contrast-filled dilated segmental bronchioles are also seen.