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Case Report

Neuroendocrine tumor of distal bile duct

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Introduction

Neuroendocrine tumors (NET) are rare, albeit, well known in gastrointestinal tract (GIT).[1] However, in GIT, biliary tree (BT) is an unusual site for NET.[2] Further, in BT, distal common bile duct (CBD) is a very rare site indeed, as we report the 8th case in literature.[2] Ours is the first case where tangential portal vein excision was employed for its radical resection.

Case Report

A 45-year-old female patient presented with 2-month history of painless progressive jaundice, pruritus, and clay-colored stools with associated weight loss. On examination, she had the following: performance status ECOG-0; no pallor, icterus +; no generalized lymphadenopathy; palpable distended gall-bladder. Relevant laboratory results were as follows: serum bilirubin −7.9 mg% (direct – 6.9 mg%); SGOT – 41 IU/L; SGPT – 54 IU/L; ALP – 188 IU/L; CEA −0.2 ng/ml and CA 19.9 was

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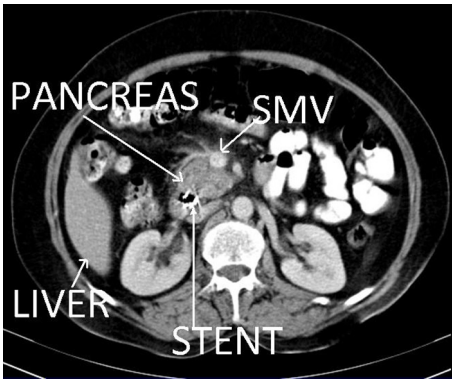

Fig. 1 – Axial CT image showing metallic stent in CBD. (SMV – superior mesenteric vein.)

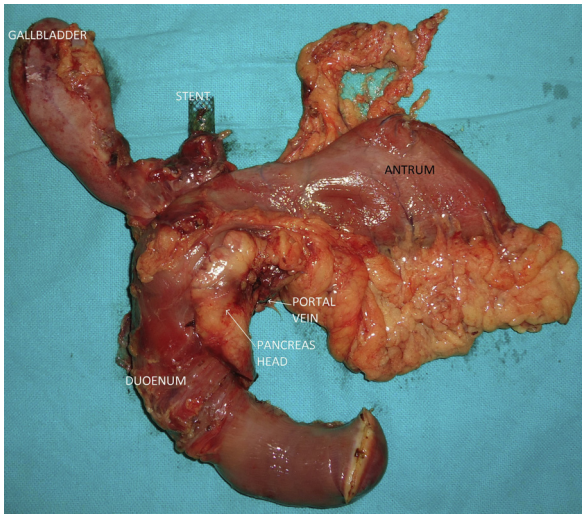

Fig. 2 – Resected pancreatoduodenectomy specimen with metal stent protruding from cut end of common bile duct (CBD).

marginally raised at 39.9 U/ml. Ultrasonography showed dilated intraheptic biliary radicals with dilated CBD with abrupt cutoff in its terminal part along with gallbladder calculi. Contrast-enhanced computerized tomogram (CECT) showed the same. Endoscopic ultrasonography (EUS) showed a 3 cm × 2 cm hypoechoic mass in terminal CBD. The patient was inappropriately stented with a metal stent at a different center (Fig. 1). Brush cytology was negative for malignant cells. Whipple's pancreatoduodenectomy with tangential portal vein (PV) excision and primary repair was done, with tumor being operable but densely adherent to PV (Fig. 2). Patient had an uneventful recovery. Cut specimen revealed 3.2 cm × 2.7 cm × 1.8 cm tumor in distal CBD. Histopathology showed tumor cells, originating from distal bile duct, arranged in trabeculae, ribbons, nests, and islands separated by fibrous tissue. Few mitoses were seen along with infiltration of duodenum and pancreas with perineural invasion and dense surrounding desmoplasia (Fig. 3). One, out of 15 nodes dissected, was found metastatic. Immunohistochemistry (IHC) stained positive for neuron-specific enolase (NSE) and synaptophysin but negative for chromogranin (Figs. 4 and 5). Ki67 was <1%. The pathologic

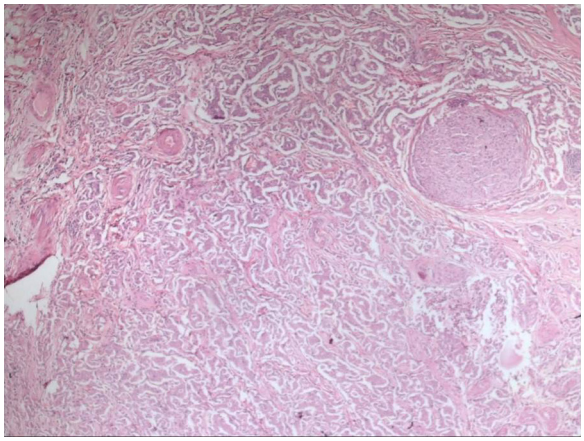

Fig. 3 – Tumor cells arranged in cords and trabeculae infiltrating the stroma. H&E stain (100×).

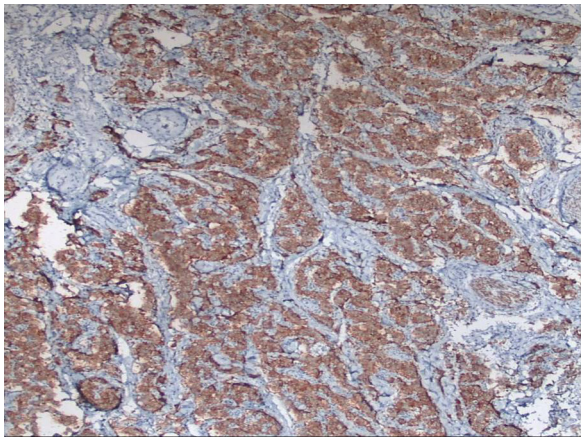

Fig. 4 – Immunohistochemistry with Neuron-specific enolase (NSE) positivity in tumor cells (100×).

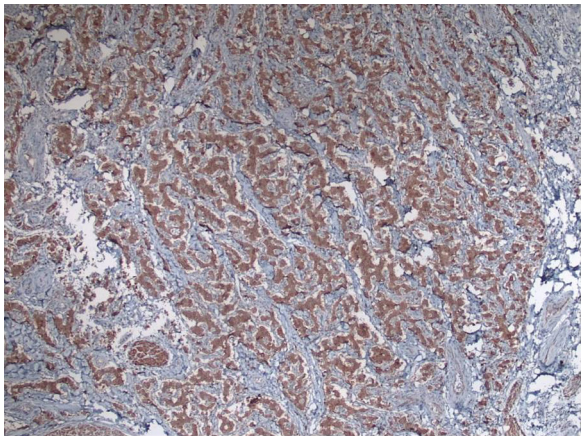

Fig. 5 – Immunohistochemistry showing synaptophysin positivity in tumor cells (100×).