Ampullary Somatostatinoma: Diagnostic Challenge and Associations

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Context

Duodenal somatostatinoma is exceedingly rare neuroendocrine tumor (NET) originating from the delta cells of the pancreas or enterochromaffin cells of the duodenum. Pancreatic tumors usually produce clinical symptoms due to excess somatostatin production manifesting as a triad of diabetes mellitus, cholelithiasis, and steatorrhea, referred to as inhibitory syndrome due to the suppression of insulin, cholecystokinin, and pancreatic exocrine enzymes. The duodenal somatostatinomas are often nonfunctional. Herein, we report a subtle case of somatostatinoma arising from an ampulla.

Design

64-year-old male started experiencing sudden onset shortness of breath, melena, intermittent abdominal pain, and anorexia a few months after being involved in an uneventful motor vehicle collision.

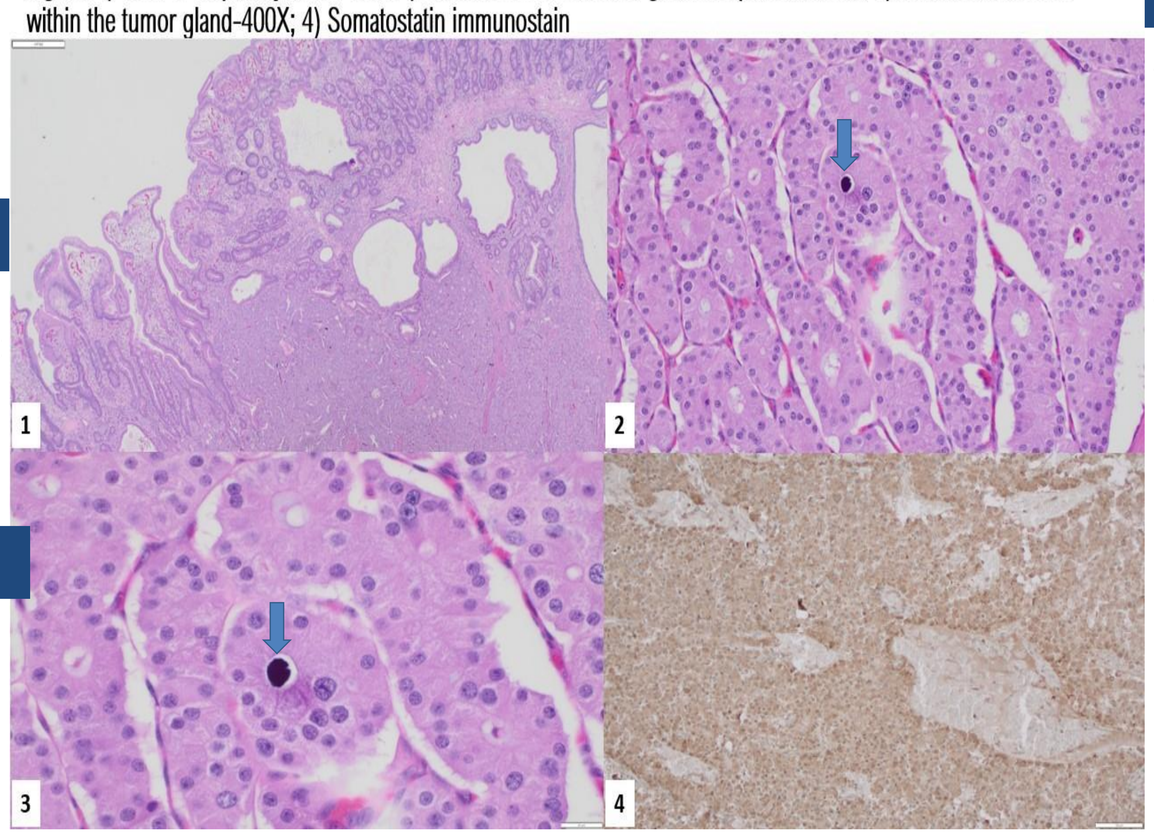
Results

Imaging displayed 2 cm ampullary nodule protruding into the duodenum. The upper endoscopy revealed non-obstructing mass, biopsy of which showed well differentiated neuroendocrine tumor.

Results

Subsequently, the Whipple resection unveiled 2.5 cm subepithelial lesion, infiltrating the sphincter of Oddi and duodenal muscularis propria. Histologically, lesion was composed of small nests of cells with stippled chromatin arranged in a glandular pattern with focal psammoma bodies within the glandular lumina. Immunohistochemically tumor cells were positive for synaptophysin, chromogranin, CAM-5.2, and somatostatin. Mitotic count was <2 mitosis/ 2 mm2 with Ki-67 proliferation index of 5%, confirming the diagnosis of somatostatinoma (NET, WHO Grade 2) (**See Figure**). Three peripancreatic lymph nodes were positive for metastatic carcinoma. The psammoma bodies are concentric lamellated calcified structures which are most frequently associated with duodenal somatostatinomas in 49.4% of cases (**See blue arrow sign below**).

Figure: 1) H&E of ampullary mass, 40X; 2) Small nests of cells in a glandular pattern, 200X; 3) Psammoma bodies within the tumor gland-400X; 4) Somatostatin immunostain



Discussion

According to The World Health Organization criteria, most duodenal somatostatinomas are malignant, and hence prompt diagnosis is crucial, but can be challenging particularly in non-functional ones as in our case. The diagnosis of somatostatinomas is confirmed only by histological and immunohistochemical studies. Seeing psammoma bodies on histology are diagnostic clues. Somatostatin analogs such as Octreotide and Lanreotide are used as first-line agents after confirmation. Ampullary tumors may require genetic evaluation since their syndromic associations (NF1, MEN1, VHL).