Gangliocytic Paraganglioma Masquerading as a Duodenal Carcinoid Tumor

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Gangliocytic paraganglioma (GP) is a rare benign tumor usually seen in the second portion of the duodenum, especially in the proximity of the ampulla of Vater, in the duodenal papillary region and adjacent areas. The clinical manifestations range asymptomatic incidental finding on endoscopy to upper gastrointestinal bleeding and/or abdominal pain. GP shows unique immunohistologic findings featuring a carcinoid or paraganglioma-like appearance, ganglion cells and spindle-shaped Schwann cells. We experienced a case in which a 24-year-old man visited emergency room presenting with episodes of melena on this day in the absence of abdominal pain. The patient had several months of intermittent vague upper abdominal discomfort before. Emergency endoscopy found a 2.0×1.8 cm-sized polypoid mass with surface ulceration, close to the ampulla of Vater, substantially occupying intraluminal space of the duodenum. The histologic finding was that the tumor had some atypical cell clusters in submucosa, probably carcinoid tumor. Pylorus-preserving pancreaticoduodenectomy (PPPD) was performed. The immunohistochemical studies revealed positive cellular reaction for synaptophysin, neuron-specific enolase, CD 56, S-100 protein, negative for cytokeratin, CD 34, CD 31, CD 117 and vimentin. Consequently, the confirmed diagnosis was duodenal gangliocytic paraganglioma. The patient was discharged with recovered health and is on follow-up without any symptoms. We report a case of duodenal gangliocytic paraganglioma masquerading as duodenal carcinoid tumor.

Key Words: Gangliocytic paraganglioma; Duodenum; Carcinoid tumor; Neoplasm