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Neuroendocrine tumor of the ampulla of Vater with distant lymph node metastasis: a case report

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Purpose:

Neuroendocrine tumors of the ampulla of Vater (NETAV) are extremely rare. Because of the rarity of NETAV, its clinical features and treatment outcomes are not well understood. A proper diagnosis and the effective therapeutic plan therefore remain clinically challenging. We report a rare case of a NETAV with metastasis to distant lymph nodes

Case presentation:

A 23-year-old man was admitted to our department because of jaundice. Computed tomography revealed a periampullary tumor 20 mm in diameter. Upper gastrointestinal endoscopy revealed a bulging papilla with protruding ampullary mass. Biopsy and imaging findings resulted in a diagnosis of NETAV. The periampullary tumor was successfully managed with totally laparoscopic pancreaticoduodenectomy without any procedure-related complications. Immunohistochemically, the tumor cells were positive for synaptophysin, chromogranin A, and CD56. Ki-67 labeling index was 5% of the tumor cells. Mitotic rate was 4 mitoses per 10 high-power microscopic fields. There were two lymph node metastasis. The final diagnosis was non-functional neuroendocrine carcinoma grade 2 of the ampulla of Vater, designated T3N1M0 stage IIIb. Postoperatively, the patient underwent adjuvant chemotherapy and no recurrence for eleven months.

Conclusion:

This case demonstrates that NETAV have aggressive metastatic potential even with a small primary lesion, and radical resection with lymphadenectomy is recommended for all cases.