P132

Synchronic Ampullary Neuroendocrine Tumor and Extra-gastrointestinal Stromal Tumor in Association with Neurofibromatosis type 1; A Case report

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Introduction : The coexistence of synchronic gastrointestinal stromal tumor (GIST) and neuroendocrine tumor (NET) in a patient with neurofibromatosis type 1 (NF1) is extremely rare, and furthermore report of combined extra-gastrointestinal stromal tumor (E-GIST) and NET in NF-1 patient was not described yet, in the literature. Here, we report two discrete synchronous neoplasms in a patient with NF-1.

Methods : A 80-year-old lady presented with general weakness and dizziness. She had a medical history of perforated acute cholecystitis and peritonitis managed with percutaneous cholecystostomy, 2 years ago. Physical examination revealed icterus skin color and multiple neurofibromas all over the body. Before her visiting our hospital, abdomen CT scan was done at local clinic and CT showed dilated both intra- and extrahepatic bile ducts and hypervascular mass around 2nd portion of duodenum. Laboratory test revealed no specific abnormal findings. Endoscopy was performed and it showed an ampullary mass lesion with ulceration which was biopsied but other mass around duodenal 2nd portion wasn't discovered. Histopathologic result was reported neuroendocrine tumor of ampulla of Vater with histologic grade 1.

Results : The patient underwent open laparotomy, cholecystectomy and transduodenal ampullectomy with retroperitoneal mass excision placed at the retroperitoneum, behind posterior wall of duodenum in view of CT findings. On microscopic section and immunohistochemistry, ampullary mass reported NET, grade 1 as same as previous endoscopic biopsy and retroperitoneal mass, behind the duodenal 2nd portion was turned out GIST, very low risk.

Conclusions : Herein, we report a extramly rare case of synchronous E-GIST and NET with a history of neurofibromatosis type 1.

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