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NEUROENDOCRINE CARCINOMA RESPONSIBLE FOR GASTROINTESTINAL TRACT PERFORATION

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In terms of their clinical and histopathologic presentation, neuroendocrine gastrointestinal tumours constitute an extremely diversified group of malignancies and thus are difficult to diagnose. Late and often accidental diagnosis means a multistage therapeutic process. The authors present the case of a 78-year-old female patient with clinical symptoms of visceral perforation followed by diffuse peritonitis. The patient was immediately operated. Intraoperative presentation revealed annular narrowing of the intestinal lumen by a tumour located in the cecum just above Bauhin's valve. The free tenia was microperforated in the described lesion area and had been the primary cause of diffuse fibrinous and pyogenic peritonitis. In addition, choleliths were found in the gallbladder. Right hemicolectomy with regional lymphadenectomy and cholecystectomy were performed. No postoperative complications. Histopathologic examination of resected specimen returned carcinoma neuroendocrinale. The authors argue, that the uncommon clinical course and circular, closing growth of the small cecum-located tumour with coincident perforation may originally suggest non-epithelial disease background.
Key words: neuroendocrine carcinoma, cecum, perforation

Neuroendocrine carinoma is a tumor that develops as a consequence of cellular transformation of the diffuse endocrine system (DES). The above-mentioned cells localized throughout the human body can form clusters (pancreatic islets), smaller groups, or be isolated. Diffuse endocrine system cells are mostly connected with the gastrointestinal and bronchial mucous membrane (1). The term „neuroendocrine" is derived from the phenotypic and secretory similarity to neural cells. These cells produce peptide hormones and biogenic amines. Some serve as neurotransmitters in the nervous system.

In the eighties of the past century numerous scientists elaborated the thesis that these cells form the APUD (Amine Precursors Uptake and Decarboxylation) system, and tumors derived from the above-mentioned were called APUDomas (2). The basic immunohistochemical markers of these cells, and that of tumors derived from the above-mentioned are chromogranin A and neuron-specific and synaptic enolase (3). Considering gastrointestinal tract tumors the previous term carcinoid was replaced by Gastroenteropancreatic Neuroendocrine Tumors (GEP NET). Due to the secretory activity of the above-mentioned, these tumors can be divided into active- those secreting hormones, and non-active, which do not secrete hormones (4). The clinical presentation of hormonally active tumors is dominated by symptoms connected with hormonal hypersecretion, and the presence of relatively insignificant local symptoms, which often renders impossible the localization of the primary neoplastic focus.

Imaging diagnostics of hormonally active GEP NET tumors should be preceded by biochemical examinations designed to confirm the diagnosis. Considering imaging diagnostics of inactive primary NET tumors, computer tomography (CT), magnetic resonance (MR), somatostatin receptor scintigraphy (SRS), endoscopic ultrasonography (EUS), endoscopy, and venous angiography are the methods of choice.

Somatostatin receptor scintigraphy is the method of choice in case of localization diagnostics of GEP tumors, since nearly 80% of the above-mentioned demonstrate somatostatin receptor type 2 expression. Imaging is possible by means of its analogues, such as octreotide labeled by indium 111 or tectrotide labeled by technetium 99m. The above-mentioned method is especially effective, when imaging tumors, such as gastrinomas, glucagonomas, inactive pancreatic tumors, and carcinoids (5).

The only effective therapeutic method is the radical excision of the tumor. In case of hormonally active tumors short-lasting somatostatin analogues are used during the perioperative period, in order to prevent hormonal crisis. It is possible to remove metastatic hepatic lesions if the liver is the only affected organ (>90% can be excised). Considering palliative therapy the following procedures are used: selective embolization or chemoembolization of arterial vessels supplying the tumor, radioablation, and isotopic therapy. Palliative surgical treatment is possible in case of uncontrolled pharmacotherapy of hormonal syndrome symptoms (individual indications).

CASE REPORT

The Authors presented a case of a 78-year old female patient with clinical symptoms of visceral perforation followed by diffuse peritonitis. The patient (W.G. No 30131/1020) with a history of obesity, arterial hypertension, and degenerative joint disease was admitted to the II Department of General and Gastroenterological Surgery, Medical University in Białystok with symptoms of diffuse peritonitis. At the time of admission to the hospital the patient presented with dehydration symptoms. The physical examination showed a normal-sized liver, with hypoperistalsis and positive peritoneal symptoms. Biochemical examinations revealed insignificant hyperglycemia and respiratory alkalosis. The abdominal cavity X-ray examination demonstrated the presence of air under both phrenic domes. Abdominal ultrasonography only showed cholelithiasis. Due to the clinical condition, after parenteral fluid supplementation, the patient was subject to emergency surgery. Intraoperative presentation revealed an annular narrowing of the intestinal lumen by a tumour localized in the cecum, just above Bauhin's valve. The free tenia was microperforated in the described area, and was the primary cause of diffuse fibrinous and pyogenic peritonitis (fig. 1).

Additionally, the patient was diagnosed with cholelithiasis. Right hemicolectomy with regional lymphadenectomy and cholecystectomy were performed (fig. 2).

The postoperative period proved uneventful. The patient was discharged from the hospital 10 days after the procedure with a properly healing postoperative wound. The histopathological examination of the resected specimen revealed the presence of a small-cell neoplastic infiltration, imitating a malignant lymphoma. Immunohistochemical examinations showed that the tumor had the specificity of carcinoma neuroendocrinale [chromogranin A (+), cytokeratin (+)] (fig. 3).

Metastatic lesions were diagnosed in one of the 12 removed lymph nodes. The patient re-

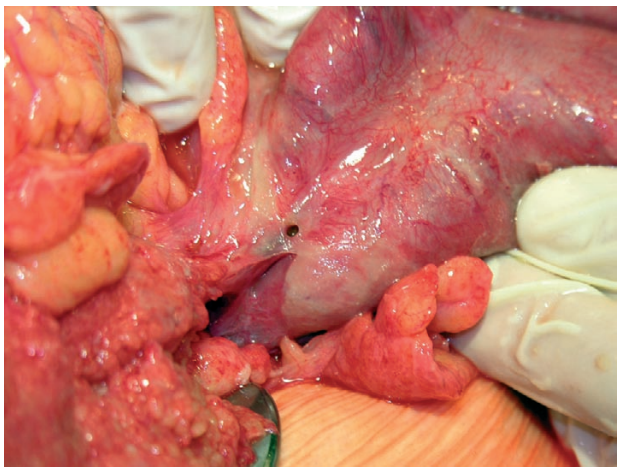

Fig. 1. Intraoperative presentation of the cecal perforation

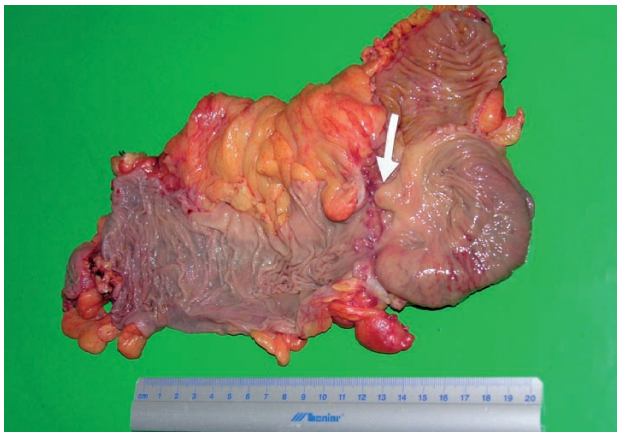

Fig. 2. The resected bowel fragment with the neoplastic lesion

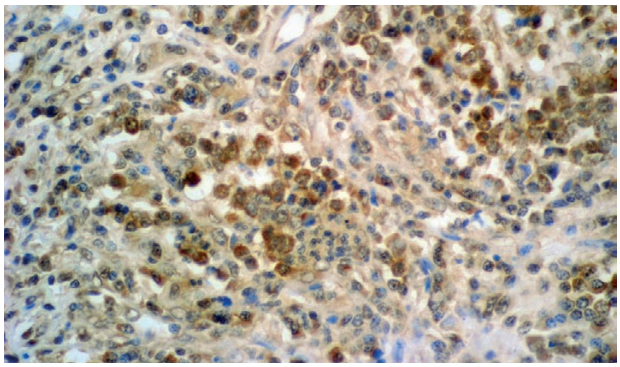

Fig. 3. Positive immunohistochemical staining for chromogranin A in neuroendocrine carcinoma cells (enlargement 400x)

mained under the control of the Specialistic outpatient clinic of the Department of General and Gastroenterological Surgery. The patient was subject to complete body scintigraphy using 99m-Tc-Tectrotide, which revealed no other pathological foci (fig. 4).

characteristic and suggested acute peritonitis, due to perforation of the infected appendix. The intraoperative presentation also posed doubts rendering impossible proper diagnosis. The circular, closing growth of the small, cecum-localized tumour with simultaneous microperforation was uncharacteristic of the commonly found adenocarcinomas. The histopathological presentation revealed a small-cell tumor of homogeneous cytoarchitecture suggesting a lymphatic background of the disease. Additionally, the immunohistochemical examinations did not confirm specific reactions towards lymphomas, and thus, directed diagnostics in the proper direction. Further immunochemical examinations demonstrated the presence of chromogranin A, specific of neuroendocrine neoplasms (3, 6). Such diagnosis enabled to perform complete body scintigraphy during the postoperative period using ^{99m}Tc HYNIC-Tyr3-octreotide.

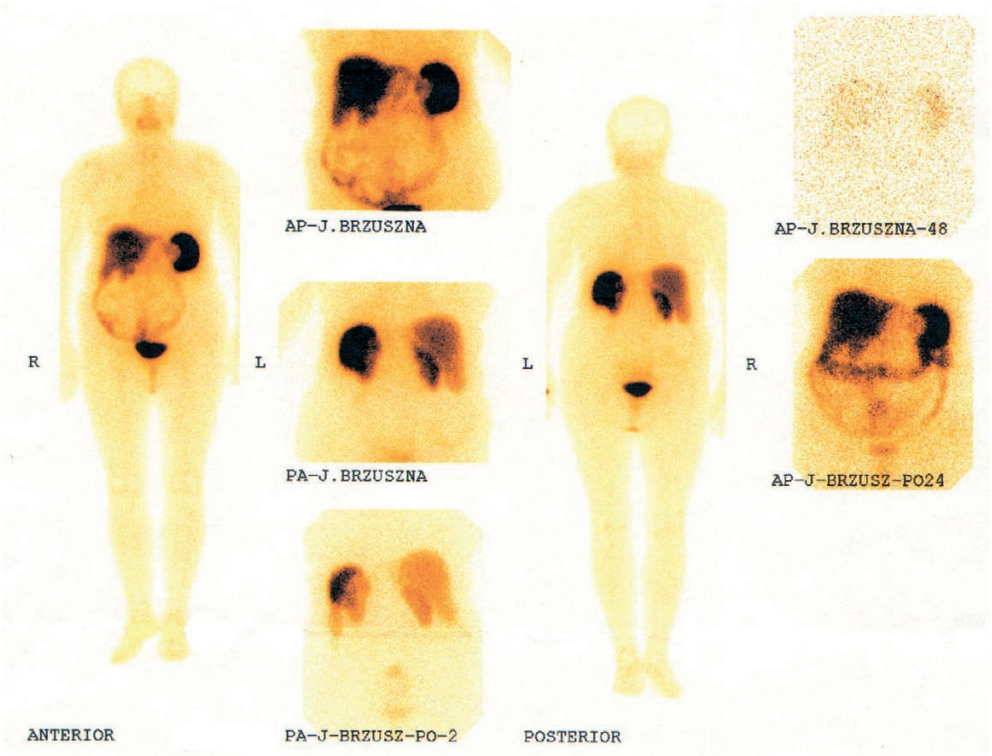

Fig. 4. Somatostatin receptor scintigraphy using ^{99m}Tc HYNIC-Tyr3-octreotide

DISCUSSION

The Authors of the study once again mentioned that the diagnosis of GEP NET was accidental. Symptoms with which the patient was admitted to the Department were non-

The presented case once again showed that medicine, and especially surgery should not be only based on statistics. The inquisitiveness of physicians and the use of all available diagnostic means enables proper diagnosis, and begin treatment.

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