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CASE REPORT

ENDOSCOPIC MUCOSAL RESECTION OF A NEUROENDOCRINE DUODENAL TUMOR

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Abstract. Duodenal neuroendocrine tumors (NETs) are rare neoplasms. In the US they have increased by 300% - 500% in the last 35 years. At the same time, their prognosis has improved. Today, most neuroendocrine tumors (NETs) of the duodenum are detected "incidentally" and therefore recognized at an early stage. Duodenal NETs which are well differentiated, not larger than 10 mm and limited to the mucosa/ submucosa can be endoscopically resected. We report a case of a 70 years old male patient admitted for iron deficiency anaemia. The colonoscopy showed a 3 mm polyp on the descending colon. The polypectomy was performed without incident. A duodenal mass was revealed at the upper endoscopy and the endosonography and endoscopic biopsy confirmed the diagnostic –duodenal neuroendocrine tumor. Because the tumor was limited to the submucosa and no lymphadenopathy or distant metastasis were seen on abdominal computed tomography (CT) endoscopic mucosal resection was performed without any incident.
Keywords: neuroendocrine tumors, endoscopic mucosal resection, duodenum

Introduction

Chronic malignant lymphoproliferation are associated with autoimmune thrombocytopenia (ITP) 1-5% in chronic lymphocytic leukemia (CLL) [1, 2], 0,76% in Non-Hodgkin Lymphoma (NHL) [3], 0,29% in Hodgkin Lymphoma (HL) [4].

CLL is the most often lymphoproliferative disease associated with autoimmune disorders . ITP complicate the course of limphoproliferative diseases, but also may precede the diagnosis (in CLL flow citometry of older pacients with ITP releved CD19/C5+ lymphocytes)[1,2].

Heterogenous histopatologycal forms of NHL are more frequent associated with ITP like: diffuse large B-cell lymphoma (DCLL) , small lymphocytic lymphoma, marginal zone lymphoma, waldenstrom disease/ myeloma, follicular lymphoma, mantle cell lymphoma; T cell lymphomas and lymphomas after autologous stem cell transplantation [3].

In Hodgkin's disease ITP take place most frequent in active phases or in periods of complete remission. The most important characteristics of the pacients are: old age , advance disease and non - nodular sclerosing histology[1, 4].

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Case Report

A 70 year-old male patient with medical history of heart failure, binodal disease with DDDR pacemaker, high blood pressure, chronic kidney disease, renal cysts, diabetes, dyslipidemia was admitted for iron deficiency anaemia.

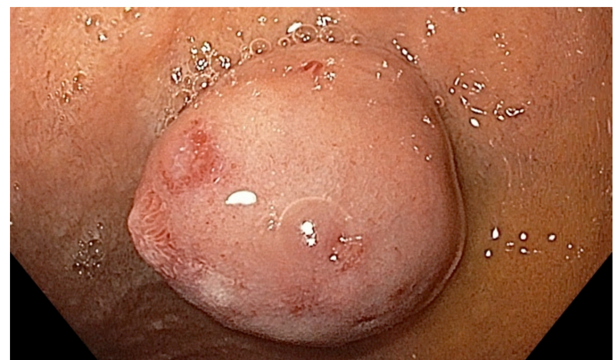

Fig.1. Upper GI endoscopy – duodenal mass

The colonoscopy showed a 3 mm polyp on the descending colon. The polypectomy was performed without incident.

The upper GI endoscopy revealed atrophic gastritis, hyperplastic gastric polyps and a duodenal mass (Figure 1.).

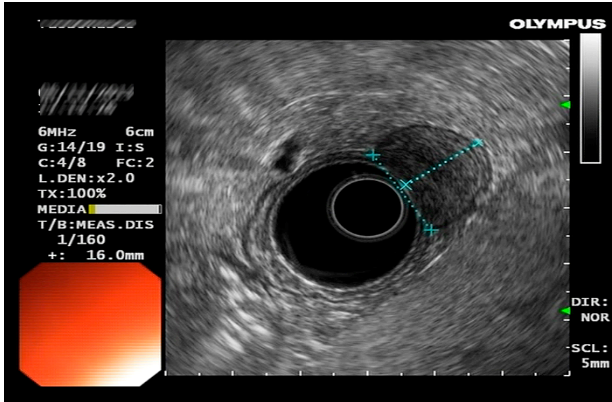

Fig.2. EUS – hypoechoic tumor

Endosonography showed duodenal mass with well-defined hypoechoic and relatively homogeneous pattern and no detectable signal at arterial Doppler examination. Also the tumor, with a diameter of 18 mm, was limited to the submucoasa.(Figure 2., 3.).

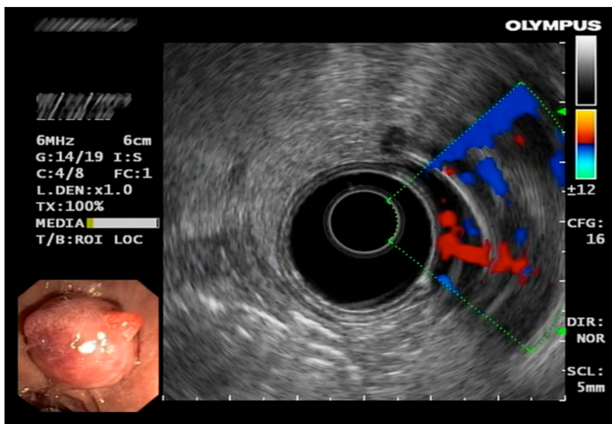

Fig.3. EUS – Doppler examination

Endoscopic biopsies were taken

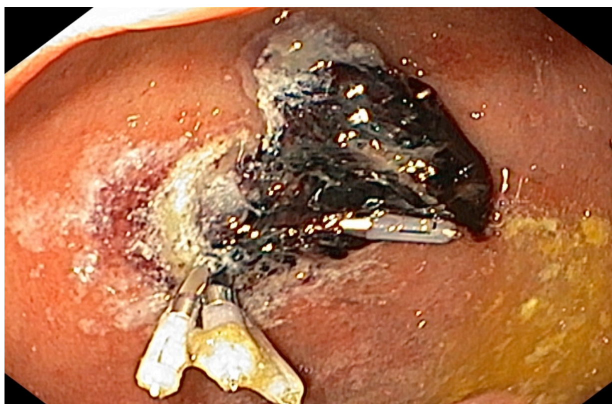

Fig.4. Hemoclipping after immediate bleeding

The histopathological and immunohistochemical diagnosis were decisive. Well-differentiated (G1) neuroendocrine tumor was the diagnosis supported by the trabecular structure, no mitosis, presence of chromogranin A, synaptophysin, somatostatin and Ki67 index less than 2 % .

Abdominal computed tomography (CT) revealed no lymphadenopathy or distant metastasis.

Endoscopic resection of the tumor was performed in one piece by endoscopic mucosal resection (EMR). Immediate bleeding after resection developed, but this was controlled by hemoclipping. No other serious complications such as late bleeding or perforation were noted (Figure 4., 5.).

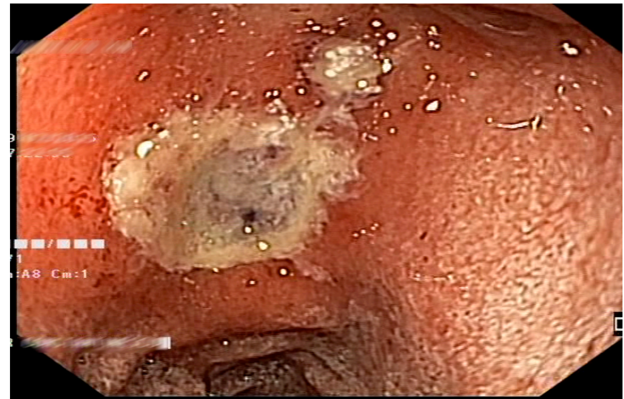

Fig.5. Endoscopic view of the resection site

Discussions

NETs originate from neuroendocrine cells throughout the body and occur most frequently (60% to 80%) in the gastrointestinal tract [9].

Duodenal NETs are categorized according to the World Health Organization classification into well-differentiated NET, well-differentiated neuroendocrine carcinoma (NEC; defined by the presence of metastases or infiltration of the muscularis propria or angioinvasion), and poorly differentiated NEC. A tumor node metastasis (TNM) classification based on tumor size, depth of invasion, and presence of lymph node metastases and/or distant metastases has been proposed and combined with a three-tiered grading system. Both G1 and G2 NETs are considered well-differentiated NETs, whereas poorly differentiated NET is graded as G3. The risk factors for metastatic disease are angioinvasion, high mitotic count, high Ki-67 index, infiltration of the muscularis propria, size > 2 cm, or metastatic spread to lymph nodes (10 -12). Well-differentiated (G1), nonfunctional duodenal NETs that are limited to the mucosa/submucosa, up to 10 mm in size, and grow nonangioinvasively, can be removed by endoscopic mucosal resection (EMR) or submucosal endoscopic dissection (ESD). These NETs carry a low risk for lymphatic or distant metastasis. [13] The recent increased use of EUS to assess duodenal NET invasion and the presence of possible lymph node metastases is particularly important in establishing NETs in this category [14]. EUS allows accurate TNM staging of duodenal NETs [15]. EUS also accurately determines the layer of origin of the lesion and the internal echo

pattern, which also adds to the differential diagnosis. EUS is necessary for the determination of endoscopic resectability.

Therapy is controversial for NETs of the duodenum that are non-functional, well-differentiated (G1), limited to the mucosa/submucosa, 10 - 20 mm in size, grow non-angio-invasively and have not metastasized. Both endoscopic therapies and surgery are considered in this situation. On the other hand, there is wide consensus that in operable patients, non-functional, duodenal NETs > 20 mm as well as all sporadic gastrinomas are to be subjected to surgical therapy [16].

Conclusions

Most well-differentiated, nonfunctional duodenal NETs that are limited to the mucosa/submucosa can be treated effectively with endoscopic resection. After it is completely removed by endoscopic mucosal resection, follow-up endoscopic examinations, abdominal ultrasound or CT and serum chromogranin A levels are recommended at 6, 24 and 36 months.

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