SHORT COMMUNICATION

Carcinoid appendicitis in a 14-year-old girl - case report with an overview of literature

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ABSTRACT

Carcinoid is a neuroendocrine tumor, secretes into the bloodstream biologically active substances (e.g., serotonin, histamine). These are rare tumors, localized in different areas of the body, including the gastrointestinal tract; mainly in the appendix. Here are found accidentally during histopathological examination of the appendix removed because of acute inflammation. The paper presents a case of carcinoid of the appendix in a 14-year-old girl. The aim of the study is to present diagnostic difficulties in the course of carcinoid pediatric patient and draw attention to the unusual clinical picture.

Keywords: carcinoid, appendix, pediatrics
1. INTRODUCTION

Carcinoid is built of cells belonging to the endocrine system APUD producing serotonin and other active substances. Most commonly occurs in the gastrointestinal tract. The average age of patients with carcinoid tumor of the appendix is 36 years. The average frequency: 1-2 / 100 000, they are more frequent in women and the elderly [1]. Enterochromaffin cells (EC) produce mainly serotonin, these cells are the most important building material of neuroendocrine tumors of the distal small intestine and appendix. Very probably that these types of tumors (consist of EC cells) behave more aggressively than neuroendocrine tumor of the duodenum and stomach constructed from ECL cells (enterochromaffin-like) [2-4].

Among the comorbidities the most important are: MEN-1, the second primary not-endocrine cancer of the gastrointestinal tract: adenocarcinomas of the colon, sigmoid, rectum, stomach, small intestine, esophagus. It is also indicated for the compound carcinoid with the presence of: scleroderma, liver cirrhosis, autoimmune thyroiditis, autoimmune gastritis and sarcoidosis [1]. Carcinoid of appendix is the most commonly diagnosed because of appendicitis, this is the most often first symptom in this location. Neuroendocrine tumors of the appendix in the majority of the cases are found accidentally after appendectomy. Most carcinoid tumors of the appendix are detected at an early stage of development.

This promotes a high survival: among the children, more than 80% of the diagnosed tumors are less than 1 cm. Tumor's size is essential for the occurrence of metastases. For lesions of less than 1 cm metastases occur in 2%, for the lesions of 1-2 cm at 50%, lesions bigger than 2 cm - from 80 to 90% of patients. Metastases occur mainly in regional lymph nodes [2-5]. In biochemical diagnosis of neuroendocrine tumors of the small intestine and appendix is useful to determine the concentration of chromogranin A (CgA, chromogranin A) - a marker-sensitive, but not specific. Significantly increased CgA can be taken as an indication of poor prognosis [4, 6, 7]. Radisotope's diagnostic using a radioisotope-labeled analogs of somatostatin (SRI, somatostatin receptor imaging) is more sensitive than x-rays. Searching for the primary lesion SRI sensitivity is approximately 80% [8]. Study using positron markers (e.g., 68 Ga) is a preferred method for imaging, particularly in the case of lesions smaller than 1 cm [9]. Searching for the primary lesion and stage of the disease it is recommended to make SRI in correlation with multiphase CT / MRI. In the case of an incomplete operation of neuroendocrine carcinoma (NEC) of the appendix or suspicion of distant metastases should be assessed SRI (by PET / CT with analogues of somathostatin labeled 68Ga lub SPECT SSA / CT EDDA 99m / HYNIC octreotate) [10]. Rare occurrence of carcinoid of the appendix obligates to a thorough macroscopic examination and biopsy. Piece of tumor should be taken from the end of the appendix, its central portion and the base, and it's necessary to specify the size of the tumor.

Macroscopic's description has to include:

- the length of the appendix sent to the examination, with description of the tumor's position relative to the resection margin.
- assessment of tumor
- the volume in three dimensions, the appearance in section,
- the state of serosal and mesenteric at the height of the tumor
- it is necessary to take numerous clippings [3].
Case report: 14-year-old girl diagnosed with recurrent pain around the bottom of the right hip. Pain occurred regardless of the time of day or intake of food. Initially, because of previous frequent urinary tract infections in an interview, doctors suspected cystitis. They abandoned the analysis of urine due to menstrual bleeding. Used empirical antibiotic therapy. Antibiotic therapy has been used for three weeks and did not produce the expected results, the pain intensified. With suspected appendicitis girl was referred to the department of pediatric surgery in order to perform appendectomy. During surgery, intraoperatively was found: large infiltration around appendix, gangrenous appendicitis, diffuse peritonitis. Surgery went without complications. Histopathological examination showed neuroendocrine tumor G1, the largest dimension of invasion; 0.8 cm, tumor limited to appendix, no exceeding the serous membrane.

The patient was referred to a Haematoncology Clinic in order to extend diagnosis, exclude distant foci. Girl was admitted to the clinic in good general condition. Laboratory tests and imaging didn't show any abnormality. In physical examination was only found scar after excising the appendix and the drain. Low clinical and histological stage determined decision of outpatient observation. Currently girl remains under the outpatient control in Haematoncology Clinic of Children's Hospital.

Discussion: The cases where the tumor's diameter is greater than 1 cm and less than 2 cm, and tumor infiltrates the tissues of the mesentery (extension into mesoappendix), according to a report College of American Pathologists, have to be regarded as an uncertain prognosis. Tumors of potentially mild course have a particle size of less than 1 cm and not infiltrate the mesentery. Tumors treatment of the appendix which are smaller than 2 cm, with no risk factors is to cut appendix. In case of advanced forms when tumors are bigger than 2 cm and infiltrate serous should be considered hemikolektomii [2,11].

Hemikolektomii is a procedure of cut-out right or left side of the colon.

Long-acting somatostatin analogues are the first choice of treatment for hormonally active neuroendocrine tumors of the small intestine. Carcinoid is poorly sensitive to chemotherapeutics. The most commonly used 5-fluorouracil with streptozocin somatostatin analogues - Octreotyd or somatulin fight with the symptoms of the disease, slow its progression. Carcinoid is a tumor with own sensitivity to ionizing radiation. Radiation therapy is reserved for advanced cases of bone metastases [2].

Proposed treatment sequence:

- surgical treatment,
- SSA in hormonally active
- radioisotope therapy
- targeted therapies (everolimus),
- symptomatic treatment.

Monitoring the course of disease and treatment should be individual for each patient. You should take into consideration the clinical picture, the clinical and histological stage of the disease and the treatment applied. For highly-differentiated tumors with a maximum diameter of the tumor less than 1 cm and status resection (R0) further monitoring is not required. Patients with cancer in stadiumG1 should be monitored every 6-12 months, the G2 stage - every 3-6 months, and G3 - every 3 months [2].
2. CONCLUSIONS

Because of nonspecific symptoms of carcinoid, tumor is a difficult condition for clinical diagnosis. Carcinoid should be taken into a consideration in diagnosis of abdominal pain for children. Confirmation of Carcinoid obligates to extended diagnosis in order to exclude metastases.

References


