MULTIPLE BURKITT'S SMALL BOWEL LYMPHOMA COMPlicated BY ACUTE INTESTINAL OBSTRUCTION (A CLINICAL CASE REPORT AND REVIEW OF PUBLICATIONS)

Small bowel lymphoma is a comparatively rare disease. Usually no typical nor explicit symptoms are determined during its course and the first manifestation can be noticed when a complication occurs. We report a case of a 40 year old man, operated on for acute obstruction of small bowel. Intraoperatively 17 tumor formations were found along the small bowel, d 3 to 5 cm, some of which had caused almost tight obstruction of the small bowel lumen. In addition, small bowel invagination was determined in 3 places. A subtotal resection of the small bowel was performed followed by a smooth postoperative period and then chemotherapy after the patient was discharged from the surgery.

Keywords: Burkitt's lymphoma, small intestine, intussusception, obstruction.

UDC: 616-006.441

Introduction

Small intestine tumors consist 2-6% of all neoplasms in gastrointestinal tract (Howdle et al., 2003; North et al., 2000). The most cases of benign tumors are: leiomioma, adenoma, lipoma, haemangioma, neurogenic tumors, polypi, etc. The malignant tumors are presented by their four forms: adenocarcinoma, sarcoma, lymphoma and carcinoid. Adenocarcinoma is dermied in approximately 50% of the cases with location mostly in the proximal section of the intestine. The sarcoma type that prevails is leiomiosarcoma, which occurs most often in ileum. Some rare cases are angiosarcoma and lyposarcoma. The small intestine lymphoma is represented mainly by non-Hodgkin’s lymphoma (NHL). Small intestine carcinomas consist less than 1-2% of all malignant tumors of gastrointestinal tract and approximately 20% of small intestine malignomas (Mutsuo et al., 1994; Serour et al., 1992). Metastatic malignomas of small intestine are rarely found in patients with melanoma, and breast or pulmonary carcinoma.

Case report

A man at 40 is hospitalized in the Gastroenterologic Clinic with pain in the epigastric area and presence of melena. Complaints have started 2-3 days after intake of non-steroid inflammatory medications against humeral joint pain. Regardless discontinued intake the epigastric pain intensifies and in moments leads to collapse-like condition. The patient was previously operated on for acute appendicitis. During the clinical examination only a faint palpatory painfulness in the epigastrum is register ed. Biochemical blood test and X-ray scanning of thorax and abdomen show no departure from the norm. The urgent fybrogastroscopy performed determines erosive gastritis with no bleeding at the moment of performance. Medication treatment with H2 blockers is started. On the third day of hospitalization, at midnight, the patient gets severe colic-like pain with nausea and heavy perspiration. Blood pressure decreases and puls rate accelerates up to 110-120 beats per minute. Passage disorders occur, abdomen gradually swells up, breathing becomes more frequent. X-ray scanning and echography reveal small intestine obstruction. The followed treatment with water/salt and glucose solutions, spasmoletic medications and purgative enema lead to reverse development of the clinical manifestation and the patient is
prepared for X-ray contrast examination of small intestine tract. Two days later, at midnight, the clinical manifestation reoccurs with greater intensity. The repeated X-ray and echographic examinations confirm the initial diagnosis of acute intestine obstruction. A decision is made for urgent operative intervention. Intraoperatively the small intestines are determined to be bloated with presence of liquid and wind. Their examination by lig. Treitzi in the distal direction 17 tumor formations are found arising from the small intestine wall in sizes between 2-3 and 5-6 cm, irregular form and solid texture that narrow the lumen. In some of them the process involves the mesenterium as well (Figure 1).

**Figure 1.**

![Image](image1)

**Figure 2.**

![Image](image2)

The first formation is registered at the 12-th cm, the next one - at the 15-th cm, and the third (with explicit stenosis) - at the 18-20-th cm. In distal direction 14 similar formations
are found. There are 3 invaginations registered (2 in the jejunal section and 1 - in the ileum) at the respective tumors. The attempts of manual desinvagination of the invaginations of the two distal invaginations are strongly complicated. The last formation is at about 60 cm away from valvula Bauchini in the form of thickened and with very solid texture 10 cm small intestine segment with almost total obstruction of intestinal lumen (Figure 2). The adhesive accretions in the ileocecal area are related to the previous appendectomy. No exaggerated para-aortal lymph nodes are found.

The rest of the organs have no pathological aberrations. Approximately 1 liter of clear transudate is evacuated from the peritoneal cavity. A subtotal resection of the small intestine is performed from the 10th cm of lig. Treitz up to the 50th cm of v. Bauchini. The continuity of the intestinal tract is reconstructed by an one-layer side to side anastomosis. Debridement of adhesions in the area of the cecum is performed. A gastroduodenal tube is placed in close proximity of the anastomosis. The postoperative period goes comparatively smooth. On the 3-4th day the patient's passage is restored, on the 4-5th day he starts feeding. Histological examination: The received material shows small intestine consisting of mucosa, submucosa and muscularis mucosae. All the layers of the specimen are diffusely infiltrated with a neoplastic lymphoid infiltrate. The infiltrate consists of intermediate to large-sized cells, with round to slightly irregular nuclei. Most nuclei display a clumped chromatin pattern and have one or several peripherally localized nucleoli. The cells have a scarce amount of cytoplasm. It is note manne mitotic figures as well as many apoptotic cells. Immunophenotype of the malignant cells: CD20+, CD10+, Bc12-, CD5-, cyclinD1-,CD23-, Bc16. CAT shows no aberrations in the peritoneal cavity. The controlling MRI discovers two tumor formations in the mediastinum: one with d 5 cm and the second - d 3 cm. The patient is hospitalized in an oncologic clinic and undergoes a treatment with chemotherapy where in three months the outcome is exitus letalis.

Discussion

The World Health Organization's Revised European American Lymphoma (REAL) Classification (Armitage et al., 1998; Harris et al., 1994; Pittaluga et al., 1998) modified in 1995 by members of the European and American Hematopathology societies (Harris et al., 1999; Pileri et al., 1998) adopted 3 main types of lymphoid malignancies regarding the morphology and the cell origin, namely: B-cell neoplasms, T-cell/natural killer (NK)-cell neoplasms, and Hodgkin's lymphoma.

Updated REAL/WHO Classification

B-cell neoplasms

1. Precursor B-cell neoplasm: precursor B-acute lymphoblastic leukemia/lymphoblastic lymphoma (B-ALL, LBL).
2. Peripheral B-cell neoplasms.
   2. B-cell prolymphocytic leukemia.
   3. Lymphoplasmacytic lymphoma/immunocytoma.
   4. Mantle cell lymphoma
   5. Follicular lymphoma.
8. Splenic marginal zone lymphoma (+/- villous lymphocytes).

**T-cell and putative NK-cell neoplasms**

2. Peripheral T-cell and NK-cell neoplasms.
   2. T-cell granular lymphocytic leukemia.
   4. Peripheral T-cell lymphoma, not otherwise characterized.
   5. Hepatosplenic gamma/delta T-cell lymphoma.
10. Adult T-cell lymphoma/leukemia (HTLV 1+).
11. Anaplastic large cell lymphoma, primary systemic type.

**Hodgkin’s lymphoma (Hodgkin’s disease)**

   2. Lymphocyte-rich classical Hodgkin’s lymphoma.

The non-Hodgkin's lymphoma occurs more and more often. Since 1970 its morbidity rate has almost doubled. Most often the location of the primary lymphoma is in the gastrointestinal tract where the small intestine is next to the stomach in the cases. The primary gastrointestinal lymphoma occurs as a primary tumor (i.e. Western type tumor) and usually is a B-cell lymphoma originating from the B-cells in the mucous lymphoid tissue. The criterion when a diagnosis “primary NHL” is determined is the presence of lymphoma only of the small intestine with regional lymphonodopathy without affection of liver, spleen or marrow.

Small intestine lymphoma presents approximately 30% of the small intestine malignancies and may occur synchronically or metachronically. Some lymphomas occur along with a chronic disease - Sprue-like malabsorption syndromes, congenial immune deficiency conditions, HIV infection, after organ transplantations with immunosuppressive treatment, hypogamma-globulinemia with lymphoid hyperplasia, etc. Ileum is the most often affected followed by jejunum and duodenum.

**Burkitt’s lymphoma/diffuse small noncleaved cell lymphoma**

In 1950 the Irish surgeon Denis Burkitt found out a high frequency rate of a clinical syndrome including malignancy of maxillae and other organs in children from Uganda which, in his honor, was called Burkitt's Lymphoma (Burkitt, 1958). It is encountered most often in Eastern Africa and the Middle East in hot and humid areas and is spread by
mosquitoes which spread malaria and other viruses and is an endemic disease. In 1960 O’Conor made the pathological description of the tumor (Mutsuo, Eto, Tsunodo et al., 1994) and soon after that an identical tumor was reported in children from the USA and Europe (Dorman, 1965; O’Conor, 1961). In 1964 the connection between the African Burkitt's lymphoma and the Epstein-Barr virus was determined. Cytologically identical tumor has been sporadically found also in patients of a non-African origin most often located in the gastrointestinal tract, ovaries and kidneys. The difference between the non-African and the African type is the less frequent osseous tumors and the more frequent affection of nasopharynx and terminal ileum (Levine et al., 1982). Burkitt's lymphoma usually is encountered with in young people and is the most frequent type of the juvenile NHL (Gisselbrecht et al., 1998). Most often it is located in ileum and ileocecal angell and consists only 5% of the small bowel lymphomas.

Lymph tissue may be found in small bowel in the deep layer of mucosis or submucosis. The process started there may spread lengthwise or crosswise affecting the mucous (tumorous or multinodular type) or may spread all over the intestinal wall (infiltrative type). Its development into serosis affecting the intestinal meshes and mesenterium constitutes the extralumen form. It is possible that the local lymph nodes are included as well. Histologically, the tumour consists of lymphoblast-like cells with high mitotic rate interspersed with macrophage containing cellular debris (starry-sky). This extremely aggressive extranodal B-cell lymphoma is probably the most studied type of the NHL group. It is characterized by translocation and disorder in the c-MYC gene on 8q24 chromosome. Schematic diagram of chromosomes 8, 2, 14, and 22 which are involved in the translocations (usually 8;14, rarely 8;22 or 2;8) which occur in Burkitt's Lymphoma. The break points for these translocations are identified by the banding regions (q24, p13, q32 and q11). These break points on chromosomes 2, 14, and 22 correspond to chromosomal regions to which have been mapped the kappa, heavy chain and lambda constant region genes respectively. In some patients with bigger B-cells a partial concurrence with the diffusive big-cell B-lymphoma is reported. These so-called “Burkitt's-like lymphomas” reveal c-MYC deregulation and greater proliferative potential compared to the “classic” Burkitt's lymphoma (O’Conor and Davies, 1960). In some forms of Burkitt's Lymphoma a connection exists between the Epstein-Barr virus and gene relocation.

The clinical symptoms are various and non-typical. These include: abdominal pain of various character and intensity, vomit, inexplicable weight loss (approximately 39% of all the patients), torpidity, apathy, permanent fatigue, night perspiration, subfertility, sporadical gastrointestinal bleeding (Anderson and Chabner, 1982). The combination of the above mentioned symptoms is reported in 10-40% of the cases. To the above the following symptoms can be added: the symptoms of acute gastrointestinal hemorrhage typical for erosion of a large vessel, the symptoms of acute peritonitis in cases of small bowel perforation, the symptoms of acute intestinal obstruction in cases of small bowel lumen obstruction.

The preoperative diagnostics of Burkitt's lymphoma is a real challenge to the clinical practicing. The compulsory elements of the examination include: contrast X-ray examination in cases of inexplicable abdominal pain, uncertainty about invagination, difficult passage, etc. and CAT.

X-ray examination can indicate four types of small bowel lymphoma: multinodular, infiltrative, tumorous and mesenteric. It must be kept in mind that CAT has limited possibilities in diagnostics of small lymphomas <0.5 cm or diffusive infiltration of the intestinal segment. The manifestation is presented by a broad range of signs: multiple intraluminal polypoid formations, broad-base ulcers, aneurismal dilatations, narrowed lumen with mycosis destruction, large excavations. In some cases lymphography may be performed as well.

The general treatment includes:

In cases of intestinal wall affection: surgical resection may be enough if 12 or more lymph
nodes are removed and they are negative. It may be supplemented by chemotherapy and irradiation.

In cases of affection of local lymph nodes: surgical resection. Chemotherapy is optional.

In cases of impossibility for resection or spreadout disease: combined chemotherapy is optional, irradiation is used to inhibit the rysk of reoccurrence. In some cases marrow transplantation is performed (Levine et al., 1982).

References


