

Acute onset of non-Hodgkin's lymphoma with serious gastrointestinal haemorrhage: diagnosis and surgical treatment (case report)

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Abstract. The authors report a rare case of acute onset of gastrointestinal non-Hodgkin's lymphoma with acute gastrointestinal haemorrhage. The patient, a man aged 49 years, was admitted to the surgical department for the evaluation of an increased anemia and weakness. Physical examination disclosed mild epigastric tenderness in response to palpation but no palpable mass. Few hours after hospital admission, he underwent a serious gastrointestinal haemorrhage with shock. Laparotomy revealed that the malignant lesions were in the upper and in the middle of the stomach and multiple white lesions were found in the small bowel. A standard radical gastrectomy with D2 lymphadenectomy and bowel resection (about 120 cm) was performed; bowel canalization was restored by esophago-jejunal end-to-side anastomosis Roux-en-Y. The main problems of surgical treatment are discussed.

Key words: Gastrointestinal haemorrhage, non-Hodgkin's lymphoma, surgery

Introduction

A high percentage of non-Hodgkin's lymphomas grows out of the primary lymphoid system: at least one-fourth arises from sites which normally contain no lymphoid tissue and they are usually defined as "extranodal primary lymphomas".

Gastrointestinal tract non-Hodgkin's lymphomas arise from Mucosa Associated Lymphoid Tissue (MALT), which probably develop within inflammatory process in response either to prolonged or abnormal antigenic stimulation, with appearance of pathological lymphoid cellular clone that gradually replaces the normal lymphoid population.

Mucosa Associated Lymphoid Tissue (MALT) lymphomas of the stomach, account for 4-8% of all gastric malignancies.

Clinical presentation and endoscopic findings in

gastrointestinal lymphoma may mimic benign and other malignant gastric disorders.

The median interval between the first symptom and diagnosis is 4-6 months but at least in 40-50% of cases, diagnosis is made at laparotomy for abdominal mass or for complications (haemorrhage, perforation, bowel obstruction).

The authors report a rare case of a patient who underwent emergency laparotomy.

Case Report

A 49-years old male complained of three weeks of epigastric pain, malaise, fatigue and anemia.

Physical examination disclosed mild epigastric tenderness in response to palpation but no palpable mass.

Laboratory tests revealed the following pathological signs:

RBC 2,910,000 (rate 4,000,000-5,500,000)

Hgb 7.8 g/dl (12-16)

Hct 26.9% (38-48)

MCH 26.8 pg (27-32)

MCHC 29.0 g/dl (32-38)

>alfa-1 globuline 3.4% (1.4-3.0%)

<gamma-globuline 8.6% (9.0-20.0%)

<total prot. 6,1 g/dl (6.4-8.2)

>fibrinogen 520 mg/dl (170-410)

At once he underwent endoscopic examination of the upper gastrointestinal tract which showed a single ulcerative lesion on the greater curve of the stomach on the body zone.

Few hours after hospital admission, he underwent a serious gastrointestinal haemorrhage with shock.

Emergency surgery was immediately performed with the suspect of gastric ulcer bleeding.

After a midline incision a standard radical gastrectomy was performed; a standard radical gastrectomy consists of a gastrectomy with D2 lymphadenectomy (resection of N1 and N2 nodes) and omentectomy; small bowel resection of about 120 cm beyond Treitz was performed.

Esophago-jejunal anastomosis end-to-side with EEA-28 stapler and entero-enteric end-to-side, hand-sutured anastomosis were made at last.

Laparotomy revealed many white and soft lesions of the small bowel while the stomach showed increased thickness with soft aspect.

After gastrotomy we revealed the presence of multiple gastric ulcerative lesions especially on the proximal and middle third, and the cavity appeared full of red blood.

There were also some increased lymph-nodes along the left gastric artery and celiac artery.

Intraoperative histopathological examination of a lymph-node and of an ulcerative lesion, showed the presence of primary gastrointestinal lymphoma; besides histopathological examination of small bowel stump of anastomosis, revealed only a flogistic infiltration by lymphocytes and plasmacells.

The postoperative course was uneventful and the patient was discharged on postoperative day 12.

The definitive histological examination indicated

a "pure high-grade B-cell non-Hodgkin's lymphoma (Ann Arbor stage mod. Musshoff IV E).

Polychemotherapy was administered in adjuvant setting 4 weeks after operation (CHOP= cyclophosphamide, doxorubicina, vincristina and prednisone).

Discussion

Primitive gastrointestinal lymphoma is a rare tumor and although its incidence is rising (1-3) for the increasing of immunodepressed condition such as in AIDS patients (4) or after transplation, it is difficult to state the role of the various therapeutic methods in treating this disease. The occurrence of a malignant tumor is a frequent complication in HIV-positive patients and is often the cause of death: non-Hodgkin's lymphoma is one of the most common neoplastic forms in AIDS patients.

Primary gastrointestinal non-Hodgkin's lymphoma is a rare tumor, representing <5% to 8% of gastrointestinal malignances.

As reported in literature, the stomach is the most commonly involved site (60%), versus 25-30% and 10-15% of the small bowel and colon, respectively; however this underlines the need for exhaustive endoscopic and radiological staging of the entire gastrointestinal tract.

Primitive gastrointestinal involvement is not common; its onset may take the form of aspecific abdominal pain, nausea and vomiting, diarrhea, bleeding, constipation, malaise and fatigue, weight loss. Today, the need for surgery for pathological diagnosis is lower and restricted to lymphomas not accessible by endoscopic or needle procedures. However in other patients (10-20%) diagnosis is made at emergency laparotomy for arising complications such as bowel obstruction (7), perforation, particularly in T-cell non-Hodgkin's lymphoma (8) and patients underwent steroid or chemotherapy, gastrointestinal haemorrhage and obstructive jaundice in duodenal lymphoma (9).

There is no current agreement on the best therapeutic approach to primary digestive tract lymphoma, because prospective studies with accurate histological typing are scarce and adequate clinical staging is even rarer. Most reports concern retrospective studies of

poorly characterized patients submitted to various treatments. Little is therefore known about the respective efficacy of the three main therapeutic approaches, surgery, chemotherapy and radiotherapy, used either alone or in combination although new therapeutic guide lines included initial surgical tumoral reduction.

The positive role of surgical debulking of the lymphoma has frequently been stressed in the literature: although most of the studies are retrospective, complete or incomplete surgical resection in association with chemotherapy has been shown to improve the 5-year survival rate significantly more than chemotherapy alone.

Surgical resection at any side can also usually prevent fatal complications, including the risk of bleeding or perforation caused by chemotherapy or radiotherapy. The incidence of such complications varies markedly between series of studies and they are caused by tumor necrosis.

During emergency surgery, as in this report, we must perform the intraoperative diagnosis and a real stage of the malignance (histological examination during surgery).

A standard radical gastrectomy with D2 lymphadenectomy (resection of N1 and N2 nodes) with bowel resection and histological examination of bowel stump before esophago-jejunal anastomosis is the treatment of choice because it seems possible that radical excision of the tumor, whenever possible, may have remarkably improved the course of the disease.

This is clearly only a hypothesis but it may pave the way for more extensive use of elective surgery in gastrointestinal non-Hodgkin's lymphoma, in view of the steadily decreasing operative risk.

The case report suggests some reflections:

- the diagnosis of lymphoma can be established on endoscopy examination with biopsies (using a large calibre biopsy forceps) in more than 90% of patients: instead in this case endoscopic examination has misestimated the current pathology
- endoscopic ultrasound has been shown to be an accurate procedure for primary staging in gastric lymphoma: extension of the disease is defined according to the Ann Arbor staging system,

as modified by Musshoff; in addition to the classic Ann Arbor classification, this modification accounts for depth of infiltration in stage I (confined versus beyond submucosa) and extension of lymph-node involvement in stage II (regional versus beyond regional)

- radical gastrectomy is usually considered an essential component of multimodal treatment and the meticulous lymphadenectomy is performed as an excellent staging tool, providing ample histopathologic data on perigastric nodal involvement and enabling discrimination between Ann Arbor mod. Musshof stages IE, IIE1 and IIE2 with absolute accuracy
- the incidence of nodal metastasis is high, even for "early gastric lymphoma" not infiltrating beyond the submucosa (T1)
- Musshoff suggests that stage IIE1 with only paratumoral lymph-node involvement has a prognosis similar to that of stage IE lymphomas, whereas stage IIE2 with more distant subdiaphragmatic lymph-node involvement has the same poor prognosis as stage IIIIE and IV tumors.
- multiple localizations belong to stage IV
- the Ann Arbor staging does not take into account either tumor size or serosal invasion; large primary tumors have a poorer prognosis such as serosal involvement is mainly associated with high stage
- no currently available methodology can exclude residual disease beyond the surgical lymphadenectomy margin once nodal involvement is confirmed. This fact, together with the excellent results reported from several institutions, convinced us to administer adjuvant chemotherapy for a stage IIE patient, even after a potentially curative resection (10, 11)
- surgical resection can also usually prevent complications (bleeding, perforation) caused by chemotherapy or radiotherapy due to tumor necrosis (lymphoma grow not induces fibrosclerosis tissue grow like in adenocarcinoma) with a variability rate (1-10%)
- importance of intraoperative histopathological examination.

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