A very advanced case of a T cell peritoneal lymphomatosis

Ann. Ital. Chir., 2012 83: 71-73

Marco Pericoli Ridolfini*, Paola Caprino*, Stefano Berardi*, Fabio Rotondi*, Giacomo Cusumano*, Luigi Sofo*, Fabio Pacelli*, Giovanni Battista Doglietto**

**Unit of Surgical Oncology, Centro di Ricerca e Cura "Giovanni Paolo II", Campobasso*
***Department of Surgical Science. Unit of Digestive Surgery, Catholic University of Rome, Italy*

A very advanced case of a T cell peritoneal lymphomatosis

Small-bowel lymphoma is not a common desease, accounting for 15-20% of primary extranodal gastrointestinal lymphomas. Peritoneal lymphomatosis is considered a rare and aggressive presentation.
We describe the case of a 55 years-old man affected by T-cell intestinal lymphoma, presenting with diffuse abdominal involvement, bowel dysfunction, severe ascites and pleural effusion, who underwent surgery. Clinical course led dramatically to death. Preoperative cytology and radiologic investigations did not yield diagnosis and were unable to differentiate between peritoneal carcinosis and lymphomatosis. It is suggested that, in such advanced cases, with rapidly deteriorating clinical conditions and huge systemic involvement, surgery is not indicated. On the contrary, maximum effort has to be spent to obtain a preoperative diagnosis.

KEY WORDS: Carcinomatosis, Peritoneal lymphomatosis, Small-bowel lymphoma, T-cell lymphoma.

Case Report

A 55 years-old man was admitted to the Oncologic Surgery Unit of our Department complaining diffuse abdominal pain, weight loss and bowel dysfunction started nearly a month before. At the admission he presented also with general asthenia and dyspnoea. He had no previous important medical history. On clinical examination signs of bilateral pleural effusion were found. The abdomen was painful, distended for severe ascites. Peristalsis was present but torpid.

Laboratory data showed a platelets' amount of 537000/mmc. White Blood Cell count was in normal range (6530/mmc) with an altered formula: neutrofile granulocytes 80,2%, lymphocytes 11,3%. Coagulation tests were normal; Fibrinogen was 511 mg/dl. CPK and LDH levels were normal. Albumin level and total proteins levels were 3.0 g/dl and 5.5 g/dl respectively. A value of 664,4U/ml (normal range <35 U/ml) of the CA 125 was found, while Alpha-Phetoprotein, CEA and CA19-9 levels were normal.
Patient was started i.v. fluid infusion, diuretic therapy for ascites and a radiologic work-up.
Chest X-ray confirmed bilateral pleural effusion with signs of parenchymal hypoventilation. Chest computed tomography confirmed bilateral pleural effusion and presence of strongly enlarged lymph nodes at all mediastinal levels. Abdominal CT scans showed hepatic nodules and bowel distension. A dilated jejunal loop for the length of 14 cm, under the right portion of the mesocolon was noted. The intestinal segment presented an

Pervenuto in Redazione Febbraio 2011. Accettato per la pubblicazione Maggio 2011.
Correspondence to: Marco Pericoli Ridolfini, Largo Agostino Gemelli, 1. 86100 Campobasso. Italy (e-mail: marco.pericoliridolofini@rm.unicatt.it)

air-fluid level and a concentric wall thickening causing distal critic stenosis (Fig. 1). A large amount of pluricompartmental ascites along with irregular diaphragmatic and peritoneal inspissations were noted. There were also confluent lumps disseminated to the parietal, phrenic and pelvic peritoneum. All the lymph-nodal stations were pathologically enlarged. A lymphatic disease originating from the jejunum and diffusing to the thoracic and abdominal levels was suspected.

An ultrasound-guided paracentesis was firstly performed, draining two liters of hemorrhagic ascites. Cytology was unable to give diagnosis: it only revealed the presence of erythrocytes and many histiod elements with nuclear dimorphism. Immunochemistry of the centrifugalized cells deposit was C20-negative. Colonoscopy was performed without evidence colonic mucosa disease. Appearance of bowel occlusion, necessity to make diagnosis and fast deteriorating clinical conditions dictated surgery. At laparotomy about 3 liters of hemorrhagic ascites were drained and 10 cm sized neoplasia in one of the first jejunal loops was found, representing the stenosing primary tumour. Omentum was completely occupied by secondary nodules (Fig. 2). One nodule withdrawn for frozen-section analysis was consistent with proliferative lymphatic disease. The intestinal pathologic segment was resected and a jejunal side to side anastomosis was performed. The severity of the clinical conditions was not improved by the surgery and the following postoperative course was complicated by respiratory insufficiency for huge pleural effusion. The patient was admitted to the intensive care unit where he underwent repeated thoracentesis. Clinical conditions did not recover and he died ten days after the operation for respiratory and renal failure. Histopathology diagnosed a peripheral extra-nodal enteropathic T-cell lymphoma constant in middle-size lymphocytes, with vescicular nucleus and a proliferative index of 55-60%. At the immunohystochemistry lymphocytes' cytoplasm was positive for CD3 and BCL-2, negative for CD4, CD8, CD20, PAX-5 and BCL-6.

Discussion

Lymphomas of the gastrointestinal tract are the most common type of primary extranodal lymphomas, accounting for 5 to up 10% of all non-Hodgkin's lymphomas, and those localized to the small bowel represent about 15-20% [1]. Peritoneal and omental lymphomatosis mimicking carcinomatosis is a rare manifestation of a very aggressive histological subtypes of high-grade lymphoma. CT or MR imaging alone cannot be satisfactory in differentiating between the two conditions [2] as well as the uncommon form of peritoneal tuberculosis or other pathological entities with cellular spread into the peritoneal cavity [3]. In the presented case, as well as observed by other authors [2-5], CT images suggested the possibility of a lymphoma: in this condition, surgery is an acceptable strategy only for early or resectable stages. Usually the resection is followed by systemic chemotherapy and in this settings, the B-cell lymphomas retrospectively show better prognosis than T-cell types. The prognosis of patients with T-cell lymphoma are unsatisfactory compared to B-cell lymphoma, maybe

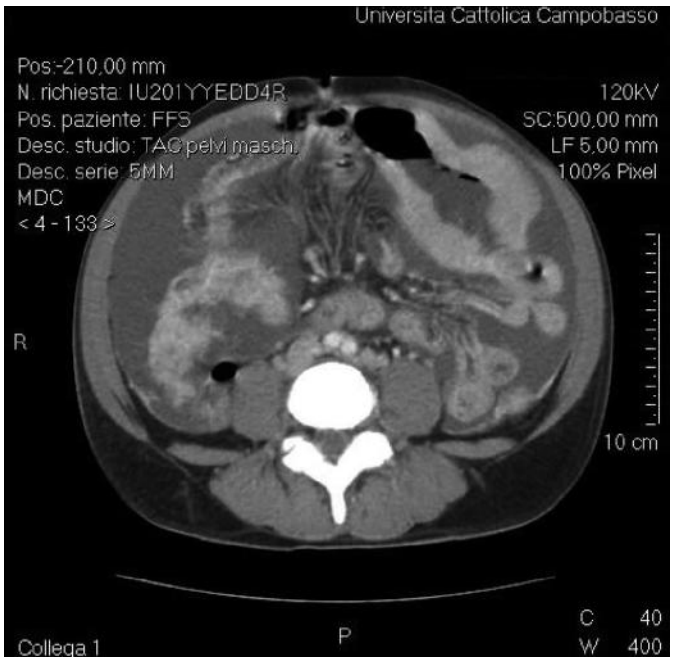

Fig. 1: CT scan showing the ileal loop, site of the primary T-cell lymphoma.

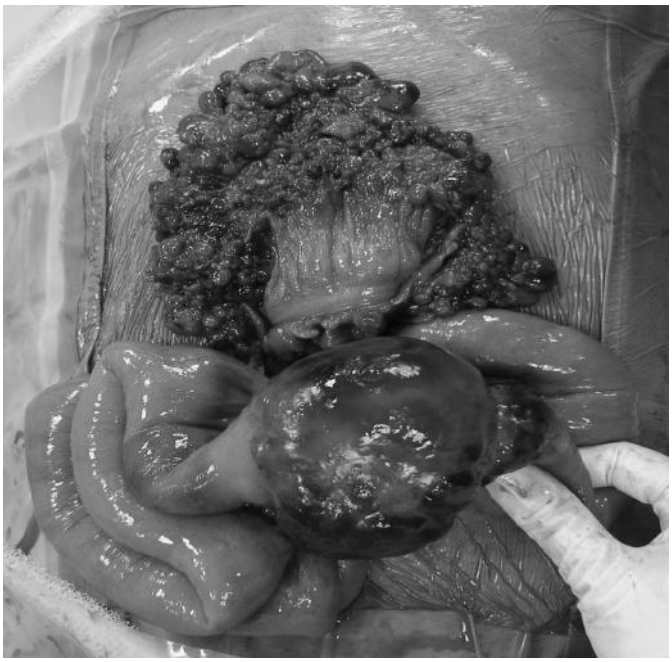

Fig. 2: Surgical field: primary tumour and omental lymphomatosis.

due either to more advanced stages at the diagnosis or to specific other prognostic factors under identification. [7,8]. Data present in literature suggest surgery for T-cell Lymphoma with peritoneal diffusion is often performed in emergency setting, maybe bearing higher morbidity. By the contrast more advanced case are best treated without surgery; biopsy or fluid sample are strongly required to make diagnosis. These methods did not prove to be conclusive in the present case, moreover the worsening symptomatology motivated a surgical treatment resulting in a fast and fatal evolution.

The experience suggests that the severity of the systemic conditions of this kind of patients enhance enormously the risk also of a laparotomy. Clinical presentation and laboratory data do not reveal such a scarce ability to recovery. Palliative bowel diversions or small resection in occluded patients affected by peritoneal carcinomatosis from adenocarcinomas, usually temporary solve critical conditions but the peritoneal lymphomatosis is a more invalidating disease. The treatment of the peritoneal seeding is based over the assumption of a still locally advanced tumour, at least in the early stage, compared with metastatic neoplasia (liver, lungs, bones etc.) sign of diffused disease. This consideration should not include conceptually the peritoneal lymphomatosis that must be retained a priori a systemic disease. The wrong assumption of resemblance between the two form of peritoneal involvement could lead to overestimate the palliative chances of any surgical attempt in the peritoneal lymphomatosis. Therefore in the clinical or radiological suspect of a peritoneal lymphomatosis we strongly suggest the importance of repeated cytological examinations attempting to achieve the diagnosis by the most soft available method in order to avoid the mistake of an unnecessary and potentially fatal surgical treatment.

Riassunto

Il linfoma del piccolo intestino non è una patologia frequente, rappresentando il 15-20% di tutti i linfomi primitivi extralinfatici gastrointestinali. La linfomatosi peritoneale è considerata inoltre una rara e aggressiva manifestazione clinica.

Noi riportiamo il caso di un uomo di 55 anni affetto da linfoma intestinale a cellule T, che lamentava dolori addominali diffusi, disturbi digestivi generalizzati, ascite severa e versamento pleurico che è stato sottoposto successivamente ad intervento chirurgico. Il decorso clinico ha però avuto esito infausto. L'esame citologico e le indagini radiologiche preoperatorie non sono state sufficienti a fornire una diagnosi né a distinguere la linfomatosi dalla carcinosi peritoneale.

Questo suggerisce che in questi casi avanzati, con rapido decadimento delle condizioni generali e diffuso coinvolgimento sistemico, la chirurgia non è il trattamento indicato. Al contrario, bisogna cercare di ottenere una diagnosi preoperatoria il più precocemente possibile.

References

1. Zinzani PL, Magagnou M, Pagliani G, Bendandi M, Gherlinzoni F, Merla E, Salvucci M, Tura S: *Primary intestinal limphoma: Clinical and therapeutic features of 32 patients.* Haematologica, 1997; 82:405-408.

2. Horger M, Muller-Schimpfle M, Yrkin I, Wehrmann M, Claussen CD: *Extensive peritoneal and omental lymphomatosis with raised CA 125 mimicking carcinomatosis: CT and intraoperative findings.* BJR 2004; 77:71-73.

3. Hamrick-Turner JE, Chieci MV, Abbitt PL, Ros P: *Neoplastic and inflammatory processes of the peritoneum, omentum and mesentery: Diagnosis with CT.* Radiographics, 1992; 12:1051-68.

4. Lynch MA, Cho KC, Jeffrey RB Jr, Alterman DD, Federle MP: *CT of peritoneal lymphomatosis.* Am J Roentgenol, 1988; 151:713-15.

5. Kim Y, Cho O, Song S Lee H, Rhim H, Koh B: *Peritoneal lymphomatosis: CT findings.* Abdom Imaging, 1998; 23:87-90.

6. Vakar-Lopez F, Jang M: *Peripheral T-cell lymphoma presenting as ascites; A case report and review of the literature.* Diagn Cytopathol, 1999; 20:382-84.

7. Chen CQ, Yin L, Peng CH, Zhao R, Chen GM, Zhou H, Li HW: *Primary non-Hodgkin lymphoma of small bowel: A clinical analysis of 34 cases.* Zhonghua Wei Chang Way Ke Za Zhi, 2007; 10:249-52.

8. Daum S, Ullrich R, Heise W, Dederke B, Foss HD, Stein H, Thiel E, Zeitz M, Riecken EO: *Intestinal non-Hodgkin's lymphoma: A multicenter prospective clinical study from the German Study Group on Intestinal non-Hodgkin's Lymphoma.* J Clin Oncol, 2003 15; 21:2740-46.