



The first-reported case of diffuse purulent peritonitis in a patient with retroperitoneal hodgkin disease (etiopathogenetic hypotheses)



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Introduction

We considered it would be interesting to report a case of purulent diffused peritonitis in an Hodgkin disease affected patient because of its rarity and of the particular clinical expression of the disease.

At the beginning we evaluated the hypothesis of two different diseases casually developing simultaneously. Then a retrospective analysis of clinical, surgical, bacteriological, and histological data suggested to us the presence of a cause-effect relationship between the retroperitoneal findings and the peritonitis. This consideration seems to be confirmed by the fact that a partial resolution of the first problem brought the second to complete control. We were not able to link precisely the physio-pathological issues between purulent diffused peritonitis and Hodgkin disease, but we could formulate two valid hypotheses that will be discussed in the report. Not having found any similar reports in the literature, we decided to publish this case, in order to have the attention of a wide that could feedback with its opinion.

Patient and methods

A 54 years old man admitted to our Department as an emergency having presented with abdominal pain and distension. He had a family history of lung cancer from

Abstract

We report a case of purulent diffuse peritonitis in a patient who was affected by Hodgkin lymphoma, with no evidence of other abdominal diseases. This is a 54 y. old. white male who was admitted to our department with a history of asthenia, recurrent fever, dysphagia and abdominal pain. In the plain abdominal radiology pneumoperitoneum was evident.

Duodenal perforation suspicion was confirmed by anamnesis and plain radiology which showed the presence of intra abdominal air.

Emergency exploratory laparotomy showed a purulent diffuse peritonitis, which relapsed after multiple surgical toilettes and peritoneal lavage. A postoperative abdominal CT scan and histology of a biopsy taken during the second surgical operation showed a retroperitoneal Hodgkin lymphoma, which went to remission after chemotherapy.

Considering the two simultaneous clinical manifestations (retroperitoneal Hodgkin lymphoma and peritonitis), we made two pathogenetic hypotheses: a) The retroperitoneal disease produced lymphatic stagnation and peritoneal transudation, which then was infected; b) The abnormal lymph nodes were infected and the abdominal cavity was contaminated from retroperitoneum from blood/lymphatic stream or by contiguity.

Riassunto

PRIMO CASO DESCRITTO DI PERITONITE PURULENTA IN UN PAZIENTE AFFETTO DA LINFOMA DI HODGKIN

Viene descritto un caso di peritonite purulenta diffusa in un paziente affetto da Linfoma di Hodgkin, in assenza di altre affezioni addominali. Si tratta di un paziente di sesso maschile di razza bianca ricoverato nel nostro Dipartimento con una storia clinica caratterizzata da astenia, febbre ricorrente, disfagia e dolori addominali. Con una radiografia dell'addome è stata evidenziata la presenza di pneumoperitoneo.

Il sospetto di perforazione duodenale è stato confermato dall'anamnesi e dalla radiografia diretta che ha mostrato

la presenza di aria nella cavità addominale. La laparotomia esplorativa d'urgenza ha evidenziato una peritonite purulenta diffusa recidivata dopo plurime toilette chirurgiche e lavaggi peritoneali. Una TC addominale post-operatoria e l'esame istologico di un campione biotico prelevato nel corso del secondo intervento chirurgico ha consentito di evidenziare la presenza di un Linfoma di Hodgkin retroperitoneale giunto a remissione dopo chemioterapia. In considerazione delle due manifestazioni cliniche contemporanee (Linfoma di Hodgkin retroperitoneale e peritonite), possiamo formulare due ipotesi patogenetiche: a) l'affezione retroperitoneale ha provocato ristagno nel sistema linfatico e trasudazione peritoneale che successivamente si è infettata; b) i linfonodi degenerati si sono infettati e la cavità addominale è stata contaminata dal retroperitoneo attraverso il flusso linfo-ematico o per contiguità.

which his father had died aged 70, and breast cancer from which his mother had died aged 65. The patient clinical background included dyspepsia in the previous months; selfprescribed assumption of NSAD for arthrosic pain. Tonsillectomized at the age of 14; and inguinal hernia repair at the age of 30. The patient had been treated in the Medicine Department for dysphagia and dyspepsia and then referred to the surgical team. At that moment, examination showed a distended and tympanis abdomen. A duodenal perforation being suspected, the patient underwent plain abdominal radiology which showed the presence of extraluminal air under the diaphragm. After routine blood tests, the patient underwent laparotomy with the preoperative diagnosis of gastric or

duodenal perforation. Surgical exploration showed of abundant intraperitoneal pus (~ 1 litre) and gas. After a complete toilette, an accurate exploration of gastrointestinal tract was made, and methylene bleu was injected through gastric and rectal tubes without evidence of gastrointestinal perforation.

Besides, other abdominal findings were: 1) Macroscopically normal liver and spleen. 2) Slightly congested appendix that was removed (Fig. 1-2); 3) An adherence of the omentum running through the left internal inguinal opening which was released and resected. 4) No evidence of disease in peritoneal lymph nodes. 5) A large retroperitoneal infrarenal mass, 6 cm in diameter, hard in consistence, not pulsating. It was thought to be a thrombosed abdominal aortic aneurysm. It was not technically possible to do an intra-operative ultrasonography, and it was not possible to explore the retroperitoneum because of the general conditions of the patient.

Three days after the operation the patient underwent abdominal CT scan of with contrast enema, which showed no evidence of aneurysm, while there were large paraaortic and infrarenal lymph nodes (diam. 4 cm). A systemic disease involving the lymph nodes was suspected, and the hypothesis of a relation with the septic peritonitis was made (Fig. 3-4).

A FNAB was attempted twice without any help for diagnosis and a lymph nodal biopsy was requested (1). A blood culture and culture of the pus taken during laparotomy was positive for *Bacterioides Fragilis*. Postoperatively and despite the daily washed abdominal drainages, neutrophile leucocytosis was maintained (25-30000 cells/mm²) with fever relapsed.

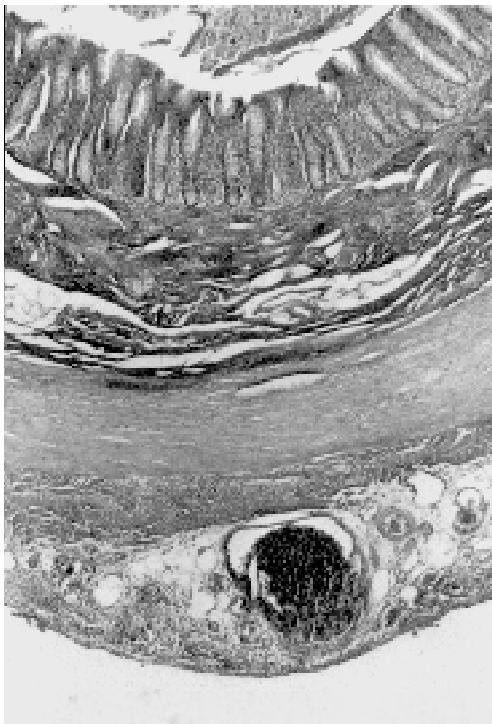


Fig. 1: Heavy congested appendix and inflammatory serosal infiltrate. E.-Van Gieson - 50X.

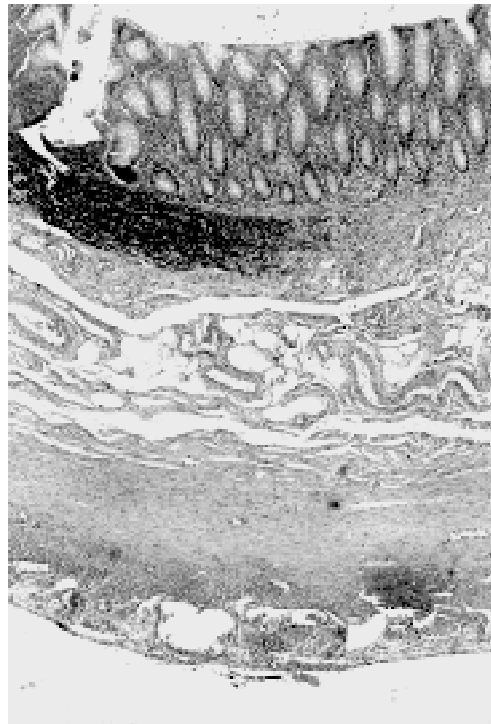


Fig. 2: Appendix wall with slowing mild inflammatory infiltration of muscular wall and thick inflammatory infiltration of the serosal layer. E. E. - 50X.



Fig. 3-4: CT scan. Enlarged paraaortic lymph nodes (max diam > 4 cm).

A new abscess was drained under the right hemidiaphragm, but it was not possible to reach the pus collected in the pelvis (Fig. 5). In day 15 day from the first operation, the patient underwent surgery again. A complete toilette of the pelvis and right hemidiaphragmatic region was performed (Fig. 6), the retroperitoneum was explored and a sliced biopsy of para-aortic lymph node was taken.

The fever, leucocytosis and enlargement of lymph-nodes were persistent after the second operation too (Fig. 7). The histological examination confirmed our first diagnostic suspicion: a mixed cells Hodgkin lymphoma (2, 3) (CD30+, CDIS-, CD79+ CD3-). The patient general condition was worsening despite continuous lavage through the drains and the absence of new abscesses, confirmed by means of daily ultrasonography.

Relying on the correlation between the two diseases we decided to remove the abdominal drains and to transfer the patient to the Oncology Department, where he could start chemotherapy with the ABVD protocol.

After the first shut, leucocytosis reduced and fever settled as the patient general condition rapidly improved.

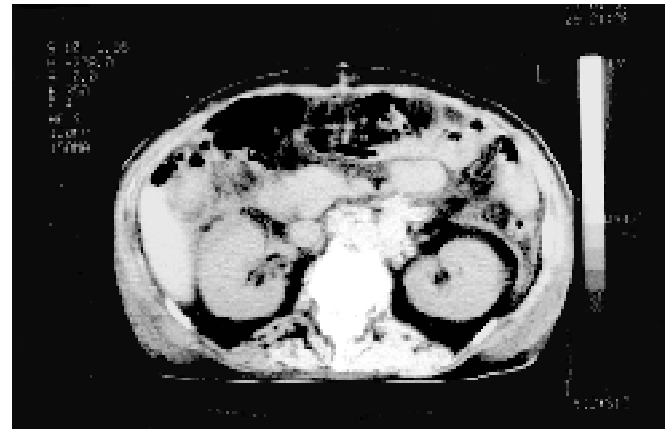


Fig. 6: Abdominal U.S. Drainage of an abscess in right subdiaphragmatic region.

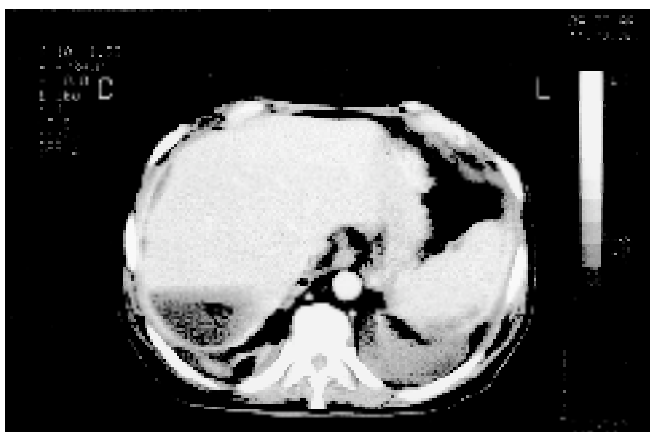


Fig. 5: Abdominal CT scan. Abscess in right subdiaphragmatic region.

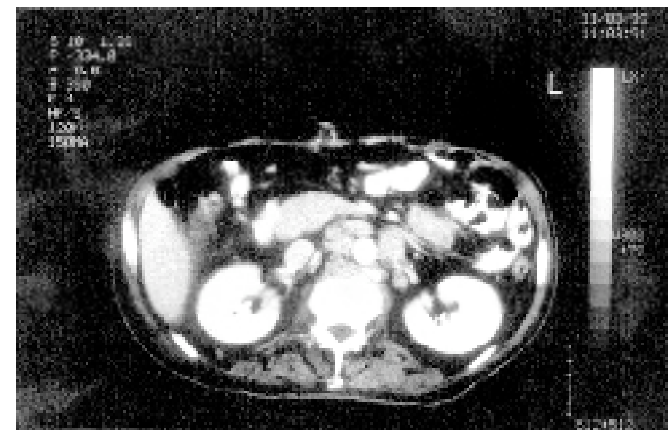


Fig. 7: CT scan. Enlarged paraaortic lymph nodes (max diam > 4 cm).

Eight months after the second operation the patient underwent CT scan which showed reduction in size of retroperitoneal lymph-nodes (from 4 to 2 cm), and complete absence of abdominal abnormalities (Fig. 8).



Fig. 8: Abdominal CT. Paraaortic lymph nodes with max diameter of 1 cm.

Endoscopic and ultrasonographic examinations confirmed the complete absence of disease in the digestive tract. Fourteen months later the retroperitoneal lymph-nodes were unremarkable. At 2 years of follow-up a new stationation of the patient was made, and the complete absence of disease was demonstrated.

Discussion

We were not able to demonstrate the etiopathogenetic mechanism that brought to the formation of an acute purulent peritonitis from retroperitoneal Hodgkin disease, but we are certain of the relationship between the two entities. This consideration is confirmed by the fact that the treatment of the lymphoma brought to the resolution the peritonitis. On the bases of the fact that it was not possible to find any similar case in literature, we hazarded some plausible hypotheses:

- 1) The neoplastic obstruction of the lymph-nodes and of the lymphatic vessels brought to lymphatic stagnation. The normal pressure of the lymph is 10 mmHg, but in case of duct obstruction it can reach 50 mmHg. This can cause the opening of lymphatic-venous shunts (4).

The lymphedema, which is consequent to the stagnation, often appears in the inferior limbs or on the external genitals, but it can be present in internal organs too, especially in abdominal organs. In lymphomas, effusions are often observed in the pleura, because of the infiltration of the pleural sheet (essudative effusion) or due to obstruction to venous drainage by compression from enlarged lymph-nodes (trasudative effusion) and in the peritoneum secondary to compression on the portal circulation (trasudative ascitis) or because of neoplastic infiltration (essudative ascitis). In those patients, a high incidence of infection, associated with tuberculous, viral or mycotic microorganisms has been observed.

This could depend on the alteration of immunity mechanisms, with the impairment of cellular immunity and the conservation of humoral immunity, that may bring the patient to anergy.

Therefore, an inflammatory or tumoral obstacle on the lymphatic ways determines a dilatation of the upstream vessels, and can cause inverted flow of the lymph, which contains waste materials, CO_2 and a poor concentration of oxygen. This is a very fertile ground for anaerobic germs. Para-aortic lymph nodes receive lymphatic vessels coming from digestive tract, liver, pancreas, and spleen. The lymphatic drainage of the abdominal digestive system has a meeting point behind the pancreas, on the confluence of the branches of the portal vein (6).

Retro pancreatic lymphoma, results in a severe clinical scenario where rapid and intense worsening of general conditions, anaemia, asthenia, meteorism and relapsing fever occur the longer lasting cases often complicate with serous effusions, sometimes with itterum and frequently with enteric haemorrhages and perforations with peritonitis. Fever occurs in a very characteristic way. Periods of two or more weeks of higher temperature are followed by periods of complete absence of fever (Pel-Ebstein fever).

The periods of regression of the fever are characterized by spontaneous regressions of the lymphoma, followed by cyclic relapses. We think that, in the case

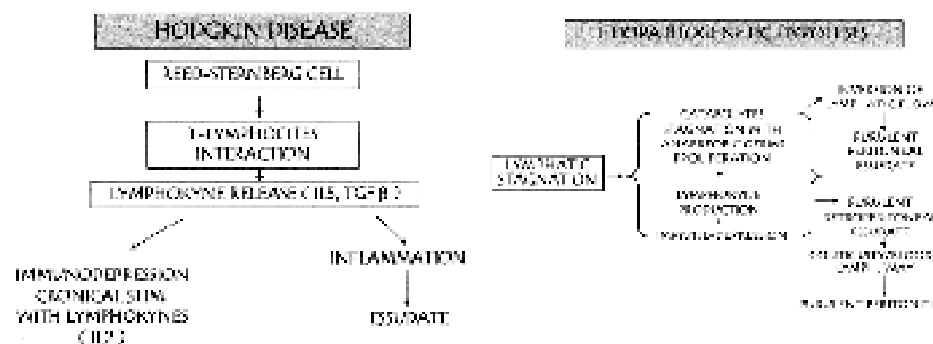


Fig. 9-10: Etiopathogenetic hypotheses.

we report, lymphatic stagnation, secondary to Hodgkin disease, brought catabolites stagnation in anaerobiosis conditions of and inversion of the lymphatic flow in the mesenteric lymphatic drainage. The consequence was the development of a peritoneal essudate or a trasudative ascitis, that subsequently was infected due to the characteristic immune depression brought by the disease. This process was worsened by the cytokines (IL5, TGF β), produced and released by T lymphocytes that interact with Reed-Sternberg cells (7). This could cause the state of immune depression and an essudate-producing generalized inflammation, with infection of the essudation material by anaerobic bacteria (8).

- 2) The purulent peritonitis could be secondary to peritoneal contamination for contiguity: the inflammatory process, located in the retroperitoneum, could have affected the peritoneum through the serosal barrier, in correspondence of para-aortic lymph-nodes.
- 3) The infectious process could be transmitted from retroperitoneum trough blood or lymphatic drainage. Therefore, the etiopathogenetic hypotheses could be scheduled as in Fig. 9, 10.

Conclusions

A close cause-effect relation of between Hodgkin disease and diffuse purulent peritonitis can be perceived in the case reported.

The complete recovery of the patient after the diagno-

sis and the beginning of the treatment for the lymphoma seems to confirm it.

Therefore the knowledge of this case could be useful to stimulate the Authors to research for possible pathogenetic relations between the two entities.

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