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Atypical course of Anaplastic Large Cell Lymphoma with positive anaplastic lymphoma kinase (ALK+ALCL)

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A 29-year-old man was admitted to hospital due to severe weakness, ascites, loss of appetite, bile vomiting, stool retention, insomnia, excessive sweating that had been increasing for three weeks. Five months earlier, he had already been hospitalized for suspected lymphoma manifested by fever and occasional cough, with no symptoms of infection, however the patient turned down further diagnosis and left the hospital at his own request. Currently, his condition rapidly deteriorated. On admission the patient was in a poor clinical condition with symptomatic ileus. Laboratory tests revealed mild anemia (13.0g/dl), leukocytosis (13.40×10³/µL), hypoproteinemia (5.2g/dl), hypercreatininemia (1.79mg/dl), elevated C-reactive protein (77mg/l) and prokalcitonine (1.53 ng/ml) serum levels. Computed tomography showed extensive infiltration in the abdominal fat tissue, ascites and lymphadenopathy, thickening of the small intestine walls, and hydrothorax. Due to the worsening clinical condition urgent laparotomy was performed. Disseminated tumorous infiltration of the peritoneal cavity, involving mesenterium, transverse mesocolon, small intestine, and retroperitoneal space were recognized (Figure 1 A,B). Dissection of the retroperitoneal space and decompression of the intestine was impossible due to the disease dissemination. Despite of intensive treatment, the patient died one hour after surgery due to the septic shock. In autopsy, disseminated lymphoma infiltration in whole thoracic and abdominal cavity was noted (Figure 1 C,D,E). Histopathological findings showed Anaplastic Large Cell Lymphoma with positive anaplastic lymphoma kinase (ALK+ ALCL); Ki67 was 100%. Immunohistochemistry revealed CD2+, CD4-, CD3-, EMA+, CK-, CD30+, MUM-1+, Alk-1+, CD23-, CD10-, CD20-, bcl-2-, cyklina D1-, bcl-6-, c-myc+ (Figure 1 F,G,H).

ALK+ ALCL was firstly described by Stein et al. in 1982 and defined as a peripheral T-cell lymphoma (PTCL) usually consisting of large neoplastic cells with abundant cytoplasm and pleomorphic, often horseshoe-shaped, nuclei, with a translocation involving the ALK gene, and expression of ALK protein and CD30 [1,2]. ALCL is a chemo-sensitive disease with
overall survival of 70–90\% \[3\]. It represents approximately 3 \% of large cell non-Hodgkin lymphomas \[2\]. ALK-positive ALCL most commonly occurs in the second-third decades (Male/Female ratio, 6.5). The majority (50–70\%) of patients present with stage III–IV disease with peripheral (mediastinal and/or abdominal) lymphadenopathy, systemic B symptoms (54-75\%) and extranodal involvement (60\%), such as skin (8-21\%), soft tissues (17-21\%), lung (6–13\%), liver (3–17\%), and spleen (8–21\%), as well as bone marrow (0–16\%) \[3\]. Intestinal location of ALCL is very rare. Only a few case reports have been found worldwide \[4\].

In presented case, ALCL’s course was fatal due to massive dissemination and ileus caused by intra-abdominal location. Systemic therapy was not possible due to the clinical condition. Despite the relatively good prognosis of ALK+ ALCL (5-year failure-free survival is 60\% and 5-years overall survival is 70\%) in this case, reckless discontinuation of diagnosis resulted in lethal complications \[5\]. Described case shows that early diagnosis, before the development of fatal complications, is crucial for prognosis.
References:


A. Intraoperative picture: Disseminated tumorous infiltration of the peritoneal cavity, involving all dilated intestines.

B. Intraoperative picture: Infiltrate tissues from mesenterium were taken for histopathology.
C. Autopsy: lymphoma infiltration in mediastinum.

D. Autopsy: lymphoma infiltration in abdominal cavity (tumor in mesenterium).
E. Autopsy: lymphoma infiltration in abdominal cavity (tumor in peripancreatic region).

F. Histopathological findings: Infiltrates of atypical lymphoid cells in the mesenterium (hematoxylin and eosin staining, magnification x 400).
G. Histopathological findings: Immunohistochemical staining in lymphoma cells: Alk-1+ (magnification x 200).

H. Histopathological findings: Immunohistochemical staining in lymphoma cells: MUM-1+ (magnification x 200).