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CASE REPORT

Lactic Acidosis, Multiorgan Failure and Pneumatosis: A Rare Presentation of Lymphoma

Phillip A Letourneau, Shinil K Shah, Amanda Tchakarov, Allen H Tanner, Robert A Hetz, Pushan P Jani, Peter A Walker, Eric J Martinez, Saleem A Khan

ABSTRACT

A 43-year-old man with a past medical history of HIV/AIDS presented with a cough, fever and night sweats. He was febrile, with normal heart rate and blood pressure. Initial laboratory testing included a mild leukocytosis. Chest X-ray demonstrated a right perihilar opacity and bilateral pleural effusions. He was admitted for treatment of pneumocystis pneumonia *vs* tuberculosis. He developed respiratory failure and was transferred to the ICU. He subsequently developed abdominal pain and severe lactic acidosis. CT scan demonstrated pneumatosis. Considering the clinical concern for mesenteric ischemia he was taken to the operating room, where it was noted that he had hepatomegaly and viable bowel. The mesentery demonstrated diffuse lymphadenopathy. Mesenteric lymph nodes were biopsied. He failed to improve clinically and his leukocytosis continued to increase to as high as 102,000, with a blood smear revealing atypical appearing lymphocytes. The patient continued to decline and ultimately died. Autopsy results demonstrated diffuse non-Hodgkin's lymphoma. Only 30 cases of non-Hodgkin's lymphoma have been reported in the literature as causing liver failure. This is the only case that included pneumatosis and lactic acidosis that mimicked mesenteric ischemia.

Keywords: Multiorgan failure, Pneumatosis intestinalis, Lymphoma, Lactic acidosis.

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RESUMEN

Un paciente de 43 años con una historia clínica de SIDA presentó síntomas de tos, fiebre, y sudoracion nocturna. El paciente tenía fiebre, con pulso y presión normales. Las pruebas de laboratorio iniciales indicaron una leve leucocitosis. Una radiografia toracica mostró opacidad peri-hiliar derecha y efusiones pleurales bilaterales. Se ingresó al paciente con diagnóstico de neumonia neumocística *vs* tuberculosis. El paciente desarrollo un fallo respiratorio y fue transferido a la sala de cuidado intensivo. Posteriormente, presentó dolor abdominal con una severa acidosis láctica. Una tomografia CT abdominal evidenció neumatosis intestinal. Sospechando una isquemia mesenterica se le llevó a cirugía, donde se notó hepatomegalia y un intestino de apariencia viable. El mesenterio presentaba una adenopatía linfatica difusa; y se tomaron muestras para biopsias. El cuadro clínico no mejoró y la leucocitosis aumentó hasta 102,000. Las laminas frotis mostraron linfocitos atípicos. El estado general siguió empeorando hasta que el paciente falleció. La autopsia mostró un linfoma difuso de tipo no Hodgkin. En la literatura medica solamente se han reportado 30 casos de linfoma de tipo no Hodgkin que hayan causado fallo hepático. Este es el único caso que incluía neumatosis y acidosis láctica sugerentes de una isquemia mesentérica.

Palabras claves: El fracaso multiorgánico, Neumatosis intestinal, Linfoma, Acidosis láctica.

BACKGROUND

Surgeons are frequently consulted for assistance in managing patients with signs and symptoms of mesenteric ischemia. These clinical features include abdominal pain, nausea and vomiting, bloody stools, metabolic acidosis, and in severe cases, sepsis and multiple organ failure. Considering these nonspecific findings, mesenteric ischemia can mimic many disease processes from abdominal aortic aneurysm to testicular torsion and can cause a diagnostic and management dilemma for clinicians. For surgeons, this dilemma is demonstrated by the question of whether an abdominal exploration will reveal the correct cause of the symptomatology and whether or not the patient will benefit from operative intervention.

Pneumatosis intestinalis is associated with many maladies and may range in clinical severity from benign conditions to acutely life-threatening ones. Benign conditions associated with pneumatosis intestinalis include pulmonary diseases, such as cystic fibrosis, chemotherapeutic agents and other diarrhea causing drugs. On the other hand, mesenteric ischemia, bowel obstruction, and toxic megacolon include possible life-threatening conditions associated with the finding of pneumatosis.[1]

CASE REPORT

A 43-year-old African-American man with a medical history of HIV infection and AIDS presented to the internal medicine service of an academic medical center with complaints of cough, fever and night sweats for 2 weeks. He had a history of IV drug abuse, but was drug-free at the time of presentation. The patient was closely followed in a community health clinic and had a recent CD4 cell count of 99 before presentation to the emergency department. He also had completed chemoradiation therapy for anal squamous cell carcinoma. Since his diagnosis of anal cancer, the patient had reported weight loss, with recent accelerated weight loss. At the time of admission, his temperature was 102°F with normal heart rate and blood pressure. Physical exam revealed decreased bibasilar lung sounds and mild

wheezing. There were no masses palpated on abdominal exam and no discernable lymphadenopathy. Initial laboratory testing showed a leukocyte count of 15 thousand and normal hemoglobin and platelets. A chest radiograph was obtained and demonstrated a right perihilar opacity, and bilateral pleural effusions. It was also noted that he had lymphadenopathy in the hilar and retroperitoneal regions. He was admitted for workup of possible tuberculosis *vs* PCP pneumonia, or possible extension of metastatic disease. Broad spectrum antibiotic coverage was initiated, including antifungals.

However, approximately 48 hours after admission, he subsequently developed hypotension and respiratory failure and was transferred to the ICU. After developing abdominal pain and distention along with severe lactic acidosis, the emergency general surgery team was consulted. The patient progressed to multiple organ failure with acute liver failure. A computed tomography (CT) scan was performed and demonstrated pneumatosis coli (Fig. 1.) Considering his unstable clinical state and concern for possible mesenteric ischemia, the decision was then made to explore the patient surgically.

The patient was brought to the operating room for an exploratory laparotomy. All bowel was found to be viable, though a open cholecystostomy tube was placed for an inflamed gallbladder. The liver was noted to be massive, extending to the iliac crest (Fig. 2). Extensive mesenteric lymphadenopathy was also noted and a biopsy was taken. The surgery team at this time suspected lymphoma as the leading diagnosis. A wound VAC was placed. Upon return to the ICU the patient required initiation of vasopressors to maintain blood pressure. It was noted that his peripheral blood smear contained atypical appearing lymphocytes. Postoperatively, his white blood cell count continuously increased to as high as 102, and his multiple organ failure progressed and required continuous veno-veno hemodialysis. On day 3 following surgery, after a discussion with the family concerning his condition, the decision to provided comfort care measures only was made, and the patient died soon after.

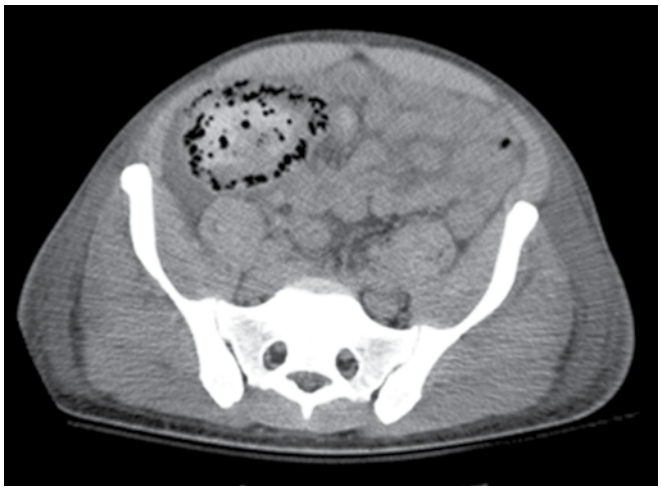

Fig. 1: Abdominal CT demonstrating pneumatosis intestinalis located in the cecum

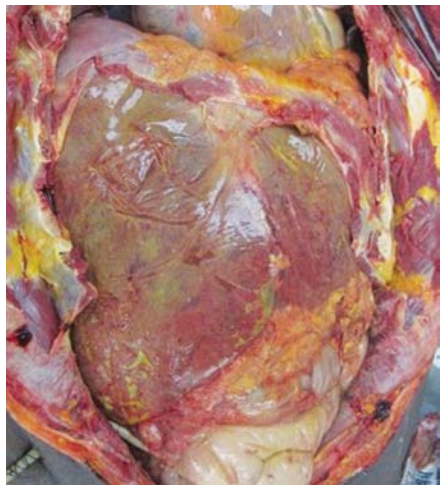

Fig. 2: Postmortem photograph demonstrating massive hepatomegaly

Autopsy results demonstrated a diffuse neoplastic process. His biopsies revealed anaplastic large cell lymphoma. All sampled lymph nodes showed large numbers of monomorphic large cells with round nuclei which stained positive for CD30 and ALK-1, consistent with a monomorphic variant of ALK positive anaplastic large cell lymphoma. It was also noted that he had findings consistent with pneumonia. The metastatic process extended to the liver with centrilobar necrosis and infiltration of neoplastic cells (Fig. 3). The kidneys and spleen were also involved.

CONCLUSION

Non-Hodgkin lymphomas (NHL) represent the most common hematologic cancers and approximately 4% of all cancers and rank seventh in total cancer incidence. Aggressive NHLs occur in 50% of all NHL cases. Anaplastic large cell lymphoma (ALCL) is a T-cell lymphoma (3% of aggressive NHLs) and is morphologically characterized by 'hallmark cells', large, pleomorphic, atypical lymphoid cells with horseshoe or multilobulated nuclei. ALCL typically present with symptoms such as fevers, weight loss, night sweats, and peripheral nodal enlargement or adenitis. Extranodal areas of involvement may be the skin, bone, soft tissues, lungs and the liver. Cases that present with a highly aggressive leukemic phase (usually small-cell variants) are rare, but can result in multiple organ involvement and poor prognosis. Current treatment for ALCL is a chemotherapeutic regimen containing doxorubicin in combination with other agents. ALK+ patients have better failure-free and overall survival compared to those that are ALK– (70-80% compared

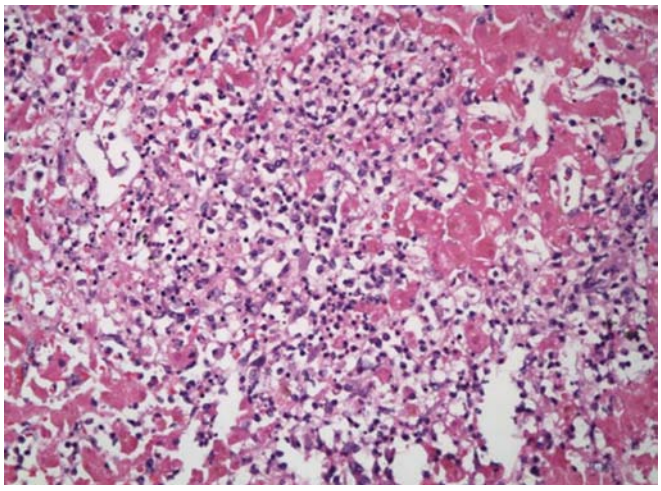

Fig. 3: Histologic examination of the liver demonstrating neoplastic cells and surrounding inflammation

to 33-49%). Patients with highly aggressive leukemic ALCL typically present with symptoms that suggest an inflammatory, rather than neoplastic condition and may be misdiagnosed by providers. In previous cases described in the literature, infections processes were initially entertained and malignancy was considered only after a failure to respond to antibiotic therapy.[2,3]

Similar cases of lymphoma presenting with acute liver failure have been previously reported in the literature in only 30 patients with a mortality rate of 83%. The previously reported cases shared common features, including hepatomegaly, lactic acidosis and rapid deterioration.[4] This case followed a similar clinical course. However, this is the first case that the authors are aware of that report hepatic failure with associated pneumatosis as presenting symptoms of lymphoma.

REFERENCES

1. Ho LM, Paulson EK, Thompson WM. Pneumatosis intestinalis in the adult: benign to life-threatening causes. Am J Roentgenol 2007;188(6):1604-1613.
2. Grewal JS, Smith LB, Winegarden JD 3rd, et al. Highly aggressive ALK-positive anaplastic large cell lymphoma with a leukemic phase and multiorgan involvement: a report of three cases and a review of the literature. Ann Hematol 2007;86(7):499-508.
3. Onciu M, Behm FG, Raimondi SC, et al. ALK-positive anaplastic large cell lymphoma with leukemic peripheral blood involvement is a clinicopathologic entity with an unfavorable prognosis. Report of three cases and review of the literature. Am J Clin Pathol 2003;120(4):617-625.
4. Lettieri CJ, Berg BW. Clinical features of non-Hodgkins lymphoma presenting with acute liver failure: a report of five cases and review of published experience. Am J Gastroenterol 2003;98(7):1641-1646.

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