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 neumatosis. 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 neumatosis and lactic acidosis that mimicked mesenteric ischemia.

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


Source of support: Nil

Conflict of interest: None declared

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 weight loss. At the time of presentation, 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.

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

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

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

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