Peritoneal Lymphomatosis Masquerading as Pyoperitoneum in a Teenage Boy

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ABSTRACT

A 16-year-old boy presented with 1 month of fever, abdominal pain, and distension. The ascitic tap drained pus-like fluid, and ultrasonography showed diffuse thickening of the omentum and mesentery with echogenic ascites. A diagnosis of pyoperitoneum due to peritoneal tuberculosis with secondary infection was suspected, and antitubercular therapy was started elsewhere, but there was no improvement. Computed tomography of the abdomen revealed enhancing soft-tissue thickening in the retroperitoneum, extending into the mesentery and encasing the superior and inferior mesenteric vessels. The ascitic fluid appearance deceptively resembled pus, but further analysis revealed atypical lymphocytes. Omental and bone marrow biopsies confirmed Burkitt lymphoma. Awareness of this rare presentation is imperative for making a correct diagnosis.

INTRODUCTION

Peritoneal lymphomatosis (PL) is a rare and aggressive tumor. Early diagnosis and prompt chemotherapy is the key to good outcome.1 The most common etiology in adults is diffuse large B-cell lymphoma and in children it is Burkitt lymphoma (BL).2 Good outcome is seen in 25% of patients with timely chemotherapy.2 This report aims to enable clinicians to recognize this entity and understand the close differential diagnoses.

CASE REPORT

A 16-year-old boy presented with 1 month of fever, anorexia, and abdominal pain, followed by distension for 1 week. Ultrasonography at another center showed ascites, omental thickening, and bilateral pleural effusion. The possibility of tuberculous peritonitis was considered. A percutaneous catheter was inserted sonographically, in the right paracolic gutter, after a diagnostic paracentesis showed purulent fluid. Ascitic fluid analysis showed 17,000/mm3 cells predominated with lymphocytes and degenerated cells with normal adenosine deaminase levels. Acid-fast bacilli staining and the nucleic acid amplification test for Mycobacterium tuberculosis were negative. The Mantoux test was nonreactive. The patient was started on antitubercular therapy (rifampicin, isoniazid, pyrazinamide, ethambutol) and antibiotics because of suspected secondary infection. Because there was no improvement, he was referred to us.

The patient had tachycardia and tachypnea, but no pallor or peripheral lymphadenopathy. The abdomen was distended, with a feeling of generalized firmness and ascites. No hepatosplenomegaly or distinct abdominal mass was palpable because of generalized distension. Initial possibilities included peritoneal tuberculosis with secondary infection and bowel perforation with pyoperitoneum.

Investigations showed leukocytosis 14.9 × 109/L and normal hemoglobin, platelets, peripheral smear, and liver functions. Sonography detected a large peritoneal collection, omental thickening, and multiple hypoechoic lesions in the liver. Chest radiograph and echocardiogram revealed bilateral pleural and minimal pericardial effusion, respectively. Abdominal computed tomography
demonstrated ascites and thickened and nodular omentum with enhancing soft-tissue thickening in the retroperitoneum, extending into the mesentery and encasing the superior and inferior mesenteric vessels. Both the kidneys showed 2–3 hypodense nodules of size 2 cm (largest), and the liver showed hypodense lesions encasing the portal vein branches. There was no lymphadenopathy (Figure 1).

Another peritoneal catheter was inserted sonographically which drained the white-colored fluid, with a total count of $2.56 \times 10^9$ cells/L with 90% atypical lymphocytes, lactate dehydrogenase (LDH) of 2,864 U/L, serum-ascites albumin gradient of 0.9, total protein of 3.7 g/dL, albumin of 2.7 g/dL, and glucose of 24 mg/dL, suggesting an exudative nature. The pleural fluid was drained to relieve tachypnea, which also showed atypical lymphocytes. Ultrasonography-guided biopsies from the thickened omentum showed atypical lymphoid cells with enlarged nuclei, coarse chromatin, and scant cytoplasm. Immunohistochemistry showed positive leukocyte common antigens, CD20, CD10, and Bcl6, suggestive of BL (Figure 2). The Ki 67 proliferation index was 90%, indicating a high tumor burden. The pus from the peritoneal cavity was composed only of tumor cells. The bone marrow examination revealed lymphomatous infiltrates. Fluorodeoxyglucose positron emission tomography was planned, but the patient became too sick to be shifted.

He had high uric acid 14 mg/dL, phosphorous 5.2 mg/dL, and LDH 1,870 U/L. LDH escalated over 24 hours to 7,870 U/L, signifying rapid proliferation. Chemotherapy (cyclophosphamide, vincristine, prednisolone, doxorubicin) was planned. Cyclophosphamide was started after intravenous hydration and rasburicase. He developed hyperkalemia (6.9 mmol/L) that caused refractory arrhythmia and death.

**DISCUSSION**

Most cases of PL have been reported in adults; however, the youngest case was a 4-year-old.3,4 The differential diagnoses of peritoneal thickening include tuberculosis, pseudomyxoma peritonei, peritoneal carcinomatosis, lymphomatosis, mesothelioma, mesenteric sarcoma, and desmoid tumors. In children, the
possibilities are tuberculosis and lymphomatosis. Sonography in PL shows thickened lamellar omentum, hypoechoic thickened mesentery that encases vessels, and nonseptate, echogenic ascites. Computed tomography shows omental caking, mesenteric soft-tissue nodularity along the vessels, lymphadenopathy, hepatosplenomegaly, hypoechogenic lesions in solid organs, and thickened bowel wall. In this patient, clinical presentation, ascites, omental caking, mesenteric thickening, and the lesions in the liver favored tuberculosis. However, soft-tissue thickening in the retroperitoneum extending into the mesentery and encasing mesenteric vessels, absence of necrotic lymph nodes, elevated LDH, high uric acid, and the ascitic fluid analysis pointed to a different diagnosis. Fluorodeoxyglucose positron emission tomography scan may reveal thickening with hypermetabolic activity throughout peritoneal cavity and can guide sampling.

Ascutic fluid is white with elevated LDH and protein, so atypical cells may be missed because exfoliated mesothelial cells may predominate the cytological picture. The treatment of PL is similar to that of BL of other sites with varying combinations of cyclophosphamide, doxorubicin, vincristine, prednisolone, methotrexate, and etoposide with or without rituximab based on staging. Although BL is chemosensitive, a large tumor burden in PL can lead to tumor lysis syndrome (TLS) and death. TLS leads to hyperkalemia, hypocalcemia, hyperphosphatemia, and hyperuricemia, causing cardiac arrhythmias and renal failure. Aggressive hydration and rasburicase can reduce the risk of TLS. A good response to chemotherapy and disease-free survival was reported in 25% cases of PL.

Early diagnosis and institution of therapy could have halted rapid tumor growth and reduced the risk of TLS in this case. However, precious time was lost because the presentation deceptively resembled pyoperitoneum.

DISCLOSURES

Author contributions: A. Ravindranath collected the data and wrote the manuscript. A. Srivastava wrote the manuscript and is the article guarantor. J. Seetharaman collected the data. R. Pandey evaluated and reported the histopathological material. MS Sarma, U. Poddar, and SK Yachha edited the manuscript.

Financial disclosure: None to report.

Informed consent could not be obtained from the family of the deceased. All identifying information has been removed from this case report to protect patient privacy.

Received October 24, 2018; Accepted March 27, 2019

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