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Intravascular Large B-cell lymphoma with Abdominal Pain

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Abstract A 76-year-old male patient presented at a local medical facility with complaints of upper abdominal pain. He was subsequently referred to our hospital for emergency care due to an abdominal aneurysm, as well as leukopenia and thrombocytopenia. Upon arrival, the patient exhibited moderate fever. A physical examination revealed a flat abdomen with tenderness and recoil pain in the upper abdomen. Consequently, a comprehensive blood test revealed bicytopenia, an inflammatory reaction, abnormal glucose tolerance, and hyperlactate dehydrogenase (LDH)emia. A whole body computed tomographic scan revealed an enlarged pancreatic head and abdominal aorta with mural thrombus without significant lymph node enlargement. The patient was administered steroids under the diagnosis of hemophagocytic syndrome, resulting in the resolution of fever, a decline in LDH and CRP levels. On the 19th day, a skin biopsy revealed a diagnosis of intravascular large B-cell lymphoma (IVLBCL). Consequently, the patient underwent repeated chemotherapy. However, the patient developed multiple organ failure and died on day 74.

Keywords: *intravascular large B-cell lymphoma, random skin biopsy, abdominal pain*

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1. Introduction

Intravascular large B-cell lymphoma (IVLBCL) is a rare clinical entity characterized by malignant B-lymphocyte growth in the lumina of small-sized blood vessels, mostly capillaries and post-capillary venules, and usually, there is no lymphadenopathy and mass formation. [1,2] The incidence is estimated to be around one in a million. [1] The median age at diagnosis is 70 years old and no sex prevalence. [1] The hallmark of the condition are the presence of B-symptoms (sweating, weight loss, or fever), anemia, and an increase of serum lactate dehydrogenase. A wide variety of skin lesions can be seen in patients with IVLBCL, and neurological symptoms are present in 35% of patients. [1] The symptoms associated with this condition are diverse and can manifest in a variety of organ-specific ways. [1] Due to the lack of lymphadenopathy and mass formation, and its unspecific symptoms, IVLBCL is always challenging to diagnose. Therefore, many cases have been diagnosed by autopsy. [2] Sampling from normal-appearing skin, called random skin biopsy (RSB), has been reported useful for diagnosing IVLBCL. [1,2]

We herein report a case of IVLBCL with abdominal pain, diagnosed by the RSB.

2. Case Report

A 76-year-old male patient presented with a history of hypertension, diabetes, ureteral stones, cataracts, and back pain. He sought medical attention at a local medical facility due to upper abdominal pain and was subsequently referred to our hospital for emergency care. His case was further evaluated using computed tomography (CT) imaging, which revealed an abdominal aneurysm, and a complete blood count revealed low platelet levels. Prior to his visit, the patient had experienced a headache and undergone magnetic resonance imaging (MRI) that did not reveal any abnormalities, resulting in his discharge from the initial facility. The following day, the patient reported nasal bleeding and bloody mucus, and he presented to a different hospital. However, he was discharged without undergoing any medical examinations. The patient's medication regimen included esaxerenone, valsartan, and empagliflozin/linagliptin. The vital signs at the time of presentation were as follows: The Glasgow Coma Scale score was E4V5M6; systolic blood pressure, 133/72 mmHg; heart rate, 71 beats per minute; respiratory rate, 16 breaths per minute; percutaneous saturated oxygen under room air, 95%.; and body temperature, 38.0°C. A physical examination revealed a flat abdomen with tenderness and

recoil pain in the upper abdomen. Additionally, there were transient scattered small erythematous patches on the abdomen and both lower extremities. A venous blood gas analysis revealed the following results: pH, 7.381; pCO_2, 32.5 mmHg; pO_2, 61.1 mmHg; HCO_3^- 18.8 mmol/L; base excess, -4.8 mmol/L and metabolic acidosis. Furthermore, complete blood counts and biochemical tests revealed leukopenia, an inflammatory reaction, abnormal glucose tolerance, and hyperlactate dehydrogenase (LDH)emia (see Table 1 for details).

A whole body CT scan revealed an enlarged pancreatic head and abdominal aorta (maximum short diameter 3.7 mm) with mural thrombus. No significant lymph node enlargement or splenomegaly was observed. Therefore, the etiology of the abdominal pain could potentially be attributed to various factors, including an impending rupture of an abdominal aneurysm, an infected aneurysm, pancreatitis, or an infectious lesion of the gastrointestinal tract. With regard to thrombocytopenia, viral infections—including rickettsia or severe fever with thrombocytopenia syndrome (SFTS)—and coagulopathy were unremarkable, suggesting idiopathic thrombocytopenic purpura (ITP), thrombotic thrombocytopenic purpura (TTP), and malignant homological disease based on elevated LDH levels. Given the high inflammatory response, cefmetazole was administered in consideration of intestinal bacterial infection, and minocycline was also administered with fasting. On the second day, abdominal pain disappeared, and blood samples showed no elevation of amylase and no diarrhea, ruling out complications of pancreatitis or gastroenteritis. On the third day, oral intake was started with a small amount of drinking water. A bone marrow aspiration was performed to determine the etiology of the patient's low platelet and high LDH levels. On the fifth day, the patient was found to be negative for rickettsia and SFTS, and minocycline was discontinued. Bone marrow puncture results showed hemophagocytosis and atypical cells; however, the number of cells observed was insufficient to diagnose malignant disease. The patient's blood test results exhibited signs of improvement, with the exception of leukopenia, and he was able to perform daily activities independently and had a good appetite. On the 7th day of the illness, the patient experienced a worsening of his chronic back pain after falling on his buttocks, and he began to show signs of delirium at night. On the 8th day of the illness, at the weekend, the patient began to experience fever and an increased oxygen demand. The patient tested negative for both SARS-CoV-2 and influenza, which were prevalent during that period. Serum LDH levels were elevated at 4,721 IU/L, and CRP levels were elevated at 32 mg/dL. Additionally, an elevated blood ferritin level (50,022 [norma range, 25-250] ng/ml) prompted the performance of the RSB on Monday, the 10th day of the disease, based on the findings from a bone marrow aspiration that were deemed inconsequential. The patient's escalating hemophagocytic syndrome prompted the initiation of steroid administration (dexamethasone, 33 mg/kg) and subsequent taper. This intervention led to the resolution of fever, a decline in LDH and CRP levels (Figure 1), and the disappearance of oxygen demand.

On the 19th day, immunostaining results of the RSB revealed a diagnosis of IVLBCL [CD20(+), CD5(+)] (Figure 2).

Large atypical cells are present in some vessels in Hematoxylin and eosin stain (x 400, left). Immunostaining shows CD20 (+, middle), CD5 (+, right) and a diagnosis was intravascular B-cell lymphoma.

Consequently, first-line THP-COP therapy [pirarubicin, cyclophosphamide, vincristine, and prednisolone] was initiated on day 20, followed by rituximab on day 33, a second course of THP-COP on day 47, rituximab on day 48, and intrathecal administration of methotrexate, cytarabine, and prednisolone on day 66. On day 68 of illness, the patient developed multiple organ failure and died on day 74.

Table 1. Results of biochemical analysis on arrival

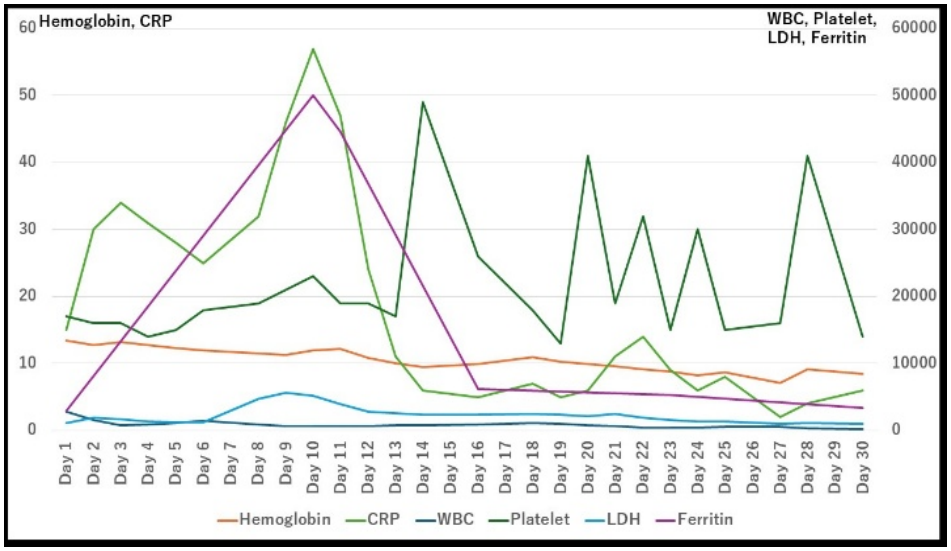
white blood cell count	2,800	/μL
hemoglobin (Hb)	13.4	/μL
platelets	17,000	/μL
total protein	6.6	g/dL
albumin	3	g/dL
total bilirubin	0.8	mg/dL
aspartate aminotransferase	55	U/L
alanine aminotransferase	20	U/L
cholinesterase	277	U/L
gamma-glutamyl transpeptidase	31	U/L
alkaline phosphatase	84	U/L
lactate dehydrogenase	1053	U/L
glucose	180	mg/dL
HbA1C	8	%
blood urea nitrogen	21.5	mg/dL
creatinine	0.81	mg/dL
creatine kinase	36	U/L
amylase	45	U/L
uric acid	10.2	mg/dL
sodium	141	mmol/L
potassium	4.5	mmol/L
chloride	101	mmol/L
C reactive protein	15.08	mg/dL

prothrombin time	10.1 (11.4)	second
activated partial thrombin time	22.1 (27.0)	second
fibrinogen	598	mg/dL
fibrinogen degradation product	7.5	μg/mL
Known at a later date		
ADAMTS-13 activity	within normal limit	
high soluble interleukin-2 receptor	3398	U/mL (normal range, 122-496)
SFTS/Rickettsia	negative	
Lactate dehydrogenase isozyme	increased 3 & 4	

Table 2. Results of comprehensive literature search

No	Author	Year	Age	Sex	LDH (IU/L)	Cause	Treatment	Chemotherapy	Outcome	Remark
1	Nakamoto	2021	70	m	920	superior mesenteric venous thrombosis, small intestine necrosis	resection	rituximab prednisone doxorubicin oncovin endoxan	6 month survival	
2	Zhang	2021	48-73	6m, 8f	no data	no data	no data	no data	3 died during 0.5 to 24.0 months followup	lower abdominal pain (2/14 cases)
3	Aasfara	2021	52	m	normal	myelo-radiculo-neuropathy	steroid	cyclophosphamide and high-dose corticosteroids	die in six weeks	
4	Atalar	2021	58	m	625	high-proliferative, neoplastic B-cell infiltration in the vascular structures at the wall and serosa of the appendix	resection	Rituximab Prednisone Doxorubicin Oncovin Endoxan	12 months survival	
5	Ronny	2017	71	f	up	portal vein thrombosis	none	none	die in 13 days	
6	Kumar	2017	67	m	471	multiple splenic infarcts	none	none	die in 9 days	
7	Crane	2014	59	m	1329	large atypical cells within vessels in the deep subcutaneous fat	none	none	die in 2 months	
8	Ishii	2008	63	f	12,200	Small intestine necrosi with superior mesenteric vein thrombosis	resection	CHP	15 months survival	
9	Kröber	2007	65	m	high	small bowel gangrene with abdominal vessels obliteration	resection	CHOP	die in 6 months	
10	Williams	2005	23	m	normal	infiltration of colonic vessels by large lymphoid cells	resection	CHOP	16 months survival	T cell lymphoma

LDH, lactate dehydrogenase; m, male; f, female; R-CHO, rituximab - cyclophophamide, hydroxydaunorubicin, oncovin, prednisolone



The patient's escalating hemophagocytic syndrome prompted the initiation of steroid administration on the 9th hospital day. This intervention led to the resolution of fever, a decline in lactate dehydrogenase and c-reactive protein levels, and the disappearance of oxygen demand.

Figure 1. Time course of laboratory data and chemotherapy

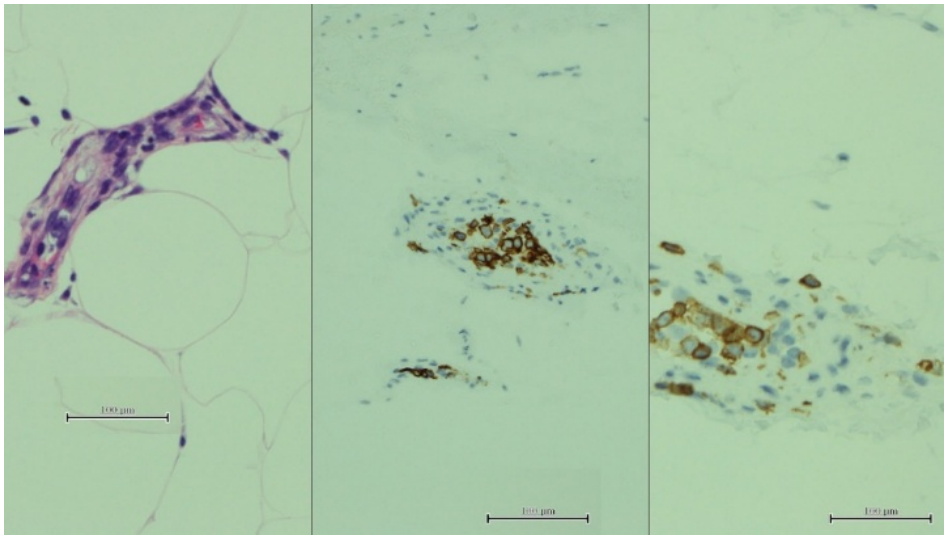


Figure 2. Immunostaining results of random skin biopsy

3. Discussion

(a) Diagnostic process

The patient presented to a medical institution with abdominal pain and underwent the RSB based on the results of bicytopenia (leukocytes and platelets), elevated LDH levels, and elevated CRP. A definitive diagnosis of IVLBCL was obtained. The final bone marrow biopsy also confirmed the diagnosis of malignant lymphoma, but the cell count was too low to determine the type of lymphoma. In this case, the RSB was useful in confirming the diagnosis.

(b) Mechanism of abdominal pain

The etiology of the abdominal pain in this case remains unclear. To address this knowledge gap, we conducted a review of previously reported cases of intravascular lymphoma (IVL) patients who also experienced abdominal pain. This review aimed to elucidate the pathogenesis of the abdominal pain in this particular case. A comprehensive literature search was conducted using the keywords "intravascular lymphoma (IVL)" and "abdominal pain," yielding a total of ten reports (Table 2). [3-12] Of these, one report was an original Chinese article that documented two cases of abdominal pain among a total of fourteen cases of IVL.[4] Notably, this report only provided an English abstract, and the underlying mechanism of abdominal pain remains to be elucidated. The report by Williams et al. was a case of T cell IVL. [12] When we analyzed the reports of nine cases, excluding the one from China, we found that eight cases were mainly due to organ ischemia symptoms caused by vascular occlusion, and only one case was presumed to be due to myelo-radiculo-neuropathy. In the present case, abdominal pain resolved spontaneously after follow-up, there were no neurological abnormalities other than peritoneal irritation symptoms, and blood gas results at the time of admission showed abnormal values for base excess, which can be caused by organ ischemia, suggesting that abdominal pain may be caused by

symptoms of organ ischemia due to temporary vascular occlusion caused by IVLBCL, which may be spontaneously relieved by infusion.

(c) Role of RSB

In this particular instance, the definitive diagnosis was rendered by the RSB rather than a bone marrow aspiration. Historically, the diagnosis of IVLBCL was frequently made using bone marrow aspiration; however, recently, the RSB has become more prevalent due to its ease of execution when compared with bone marrow aspiration. [13] Additionally, the intra-vascular malignant lymphoma in pathology results from bone marrow aspiration, and the low sensitivity of bone marrow aspiration for the diagnosis of IVLBCL is problematic, similar to us. [13] With regard to the indications for the RSB, the following criteria should be considered: First, an unexplained fever (≥ 38°C) is to be considered. Second, altered consciousness is to be noted. Third, hypoxemia (≤ 95%) is to be assessed. (<120 × 10^3/μL) to be considered. Fifth, high serum lactate dehydrogenase (LDH) is to be evaluated (>800 IU/L), and sixth, high soluble interleukin-2 receptor (sIL-2R) (>5,000 U/mL). [14] The presence of a greater number of applicable criteria indicates a higher probability of IVLBCL diagnosis. Notably, the present case in question exhibited fulfillment of all six criteria during the RSB procedure, a finding that extends to include nocturnal delirium as an altered state of consciousness. Conversely, it has been reported that the RSB should not be performed when LDH and soluble interleukin-2 receptor are normal. [15]

4. Conclusion

We present a case in which a patient presented to a medical institution with abdominal pain and underwent a bone marrow examination based on the results of two blood cell counts (leukocytes and platelets), hyper LDHemia, and elevated CRP. A definitive diagnosis of IVLBCL was made, and a discussion of the literature is provided.

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