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Pyothorax-Associated Lymphoma: A Case Report and Review

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1. Abstract

Pyothorax-associated lymphoma (PAL) is a rare disease with variations in endemic prevalence. PAL arising from a posttraumatic empyema are a rare occurrence. Here, we report a case of Epstein-Barr virus (EBV)-positive PAL arising from a posttraumatic empyemae. A male patient, 83 years old. Twenty years earlier, the patient fell from a height, sustaining a fracture to the right rib, which improved after conservative treatment. In September 2018, the patient had right chest pain (ribs 9–10), and be diagnosed with "old fractures". In February 2019, the chest pain became worse, especially during sleep. Combined with the medical history, immunohistochemistry and 18F-FDG PET/CT results, the patient was diagnosed as EBV-positive PAL. In conclusion, this case described a combination of imaging and pathological examinations which described the relationship between clinical pathological diagnosis and treatment process of a single case of pyothorax-associated lymphoma, providing important diagnostic information regarding to the rare disease of PAL.

2. Introduction

Pyothorax-associated lymphoma (PAL) is a diffuse large B-cell lymphoma (DLBCL) of the non-Hodgkin's type originating from tuberculous pyothorax [1, 2]. According to the current World Health Organization histological classification published in 2015 [3], PAL is classified as diffuse large B-cell lymphoma associated with chronic inflammation. Diffuse large B-cell lymphoma with chronic inflammation is a kind of lymphoma involving long-term chronic inflammation and is closely associated with the Epstein–Barr virus (EBV). PAL is a prototype of diffuse large B-cell lymphoma with chronic inflammation. PAL was firstly reported to have pathological and immunohistochemical features of a diffuse large B-cell lymphoma (DLBCL) in 1987 [4]. Because its clinical manifestations and imaging features are not typical, the diagnosis of PAL is very difficult.

3. Case Report

We present a male patient, 83 years old, with a history of diabetes going back more than 20 years, with controllable blood sugar. Twenty years earlier, the patient fell from a height, sustaining a fracture to the right rib, which improved after conservative treatment. In September 2018, the patient had right chest pain (ribs 9–10), no fever, no shortness of breath after physical activity, and no discomfort such as dyspnea. In February 2019, the chest pain became worse, especially during sleep. He presented to a local hospital after the chest pain became worse.

CT examination showed inflammation of the right lower lobe, chronic empyema on the right with subdiaphragmatic infection, fibrosis of the right lung, atherosclerosis, thickening of the left pleura (Figure 1a) with rib destruction, and membranous calcification around the lesion (Figure 1b). Physical examination showed that his vital signs were normal, the superficial lymph nodes throughout the body did not touch the swollen area. Each segment of the left

and right calves touched a circular subcutaneous nodule of 2 cm×2 cm and 1 cm×3 cm, respectively, with multiple skin lesions on the surface, itching, no purulence, and no tenderness. Laboratory data at admission were as follows: white blood cell count 12.5 × 109/L、neutrophil count 9.3 × 109/L、neutrophilic granulocyte percent 74.5%、red blood cell count 4.08 × 1012/L、hemoglobin 108g/L、platelet count 353 × 109/L ; C-reactive protein (CRP) 79.20mg/L、proealcitonin 0.6ng/L. After 5 days of treatment with cefoperazone, sulbactam sodium, and ornidazole, the patient's chest pain did not improve. He was admitted to our hospital on April 4, 2019 for further diagnosis and treatment. At the time of admittance, he still had chest pain and discomfort on his right side. During his illness, he lost approximately 5 kg of weight.

The results of 18F-FDG PET/CT showed the following (Figure 2): a solid mass in the lower right thoracic cavity; FDG metabolism increased periodically (SUVmax 18.0) in a manner consistent with lymphoma manifestations; the right lower lobe was swollen with inflammation, and the right pleura was thickened with calcification; right pleural effusion, multiple inflammation of the right lung; mediastinum; and both hilar multiple lymphadenitis hyperplasia were enlarged.

CT-guided needle biopsy was performed. Pathological examination (Figure 3a–d) showed inflammatory exudation, necrosis, and loose fibrous connective tissue with much lymphocytic infiltration, scattered atypical lymphocytes, basophilic cytoplasm, with large and deeply stained nuclei, small nucleolus were visible in the center of the nucleus, and mitosis readily visible. Immunohistochemistry (Figure 4a–e) showed that tumor cells were negative for CD20 and CD3, positive for background mature T lymphocytes, positive for Pax-5, positive for MUM-1, Ki-67 (70%, +); AE1/AE3, CD138, Bcl-6, CD10, CD5, CD56, MPO (-); CD79α (part +), EMA (part +), CD38 (+), bcl-2 (+). In situ hybridization (CISH): EBER+ (Fig. 4f). Based on the medical history and immunohistochemical results, the diagnosis was PAL. EBV DNA is detected: 5.03E+02 copy number /ml. The express of EBV-IgA was negative. These findings were consistent with a diagnosis of PAL (clinical stage IE).

According to the medical history, imaging examination, and pathological results, mini-CHOP chemotherapy was implemented on April 11, 2019. The patient stopped taking analgesics on the day of chemotherapy. The pain disappeared one week later, and the EBV-DNA fell to the normal range. In the later stage, the family members of the patient declined further patient treatment and reexamination. The patient's condition worsened in July 2019 and he died on September 25, 2019.

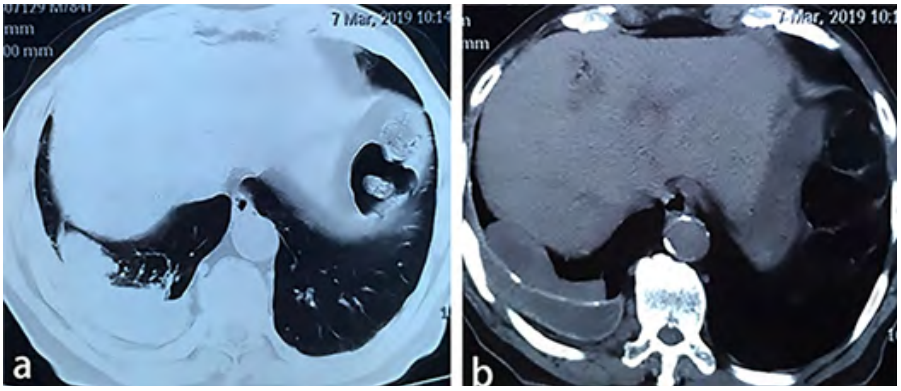

Figure 1: The results of CT examination showed (a) inflammation of the right lower lobe, chronic empyema on the right with subdiaphragmatic infection, fibrosis of the right lung, atherosclerosis, thickening of the left pleura, with (b) rib destruction, and membranous calcification around the lesion.

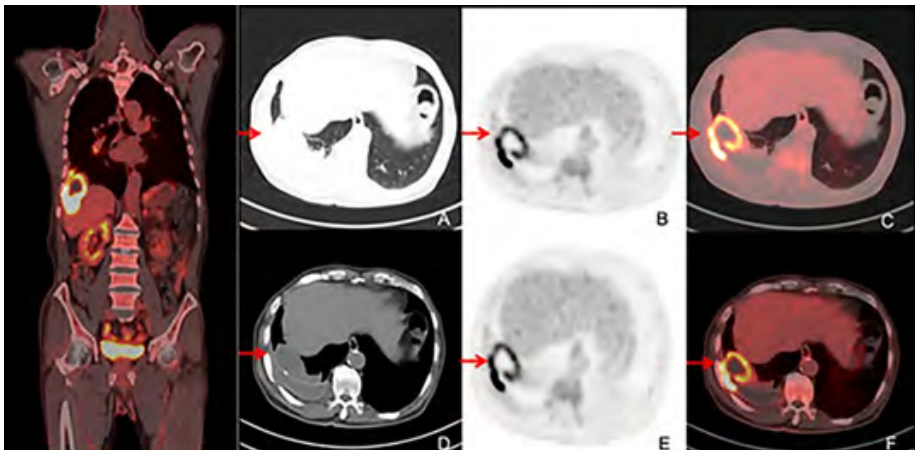

Figure 2: Examination of 18F-FDG PET/CT.

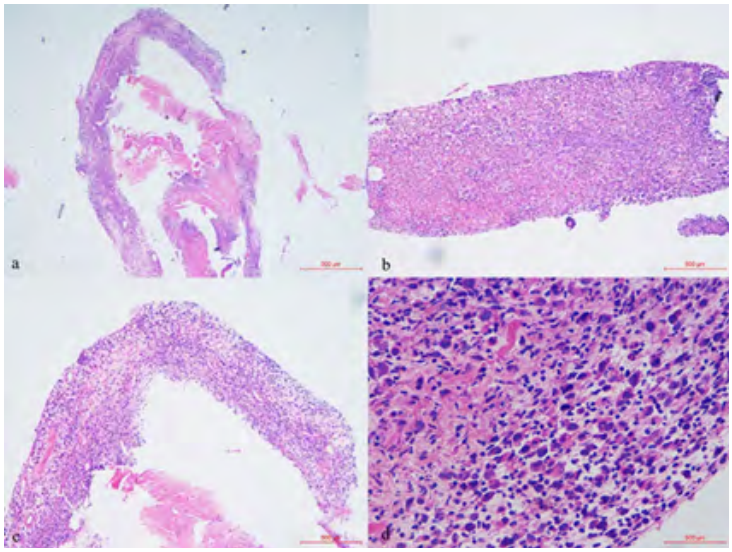

Figure 3: Histopathological findings of the tumor showing diffuse large B-cell, non-Hodgkin's lymphoma (a, HE×40, b, HE×100, c, HE ×100, d, HE×400).

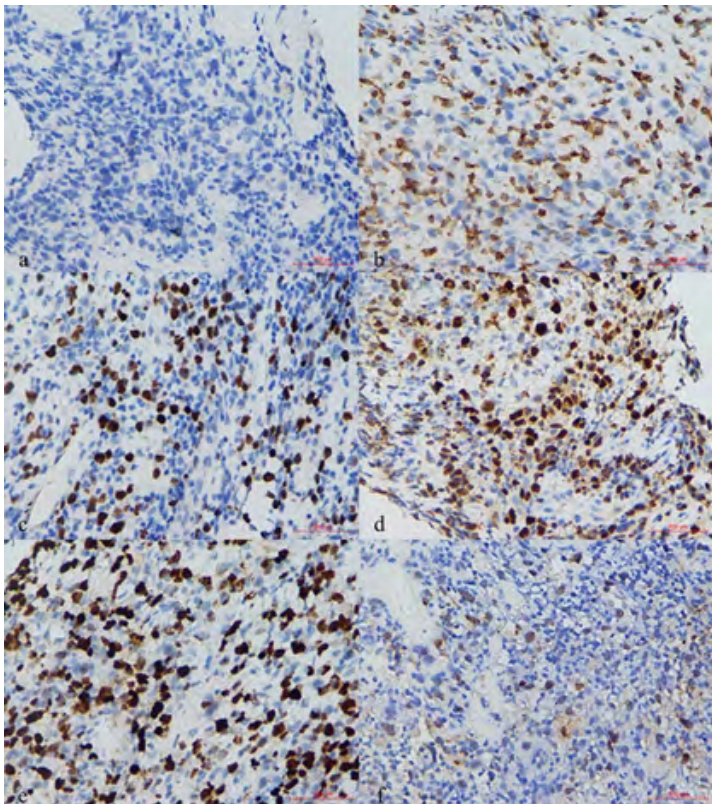

Figure 4: (a) Tumor cells show negative immunoreactivity for CD20. (b) Tumor cells show negative immunoreactivity for CD3. (c–f) PAX-5, MUM-1, KI-67, EBER were positive (a–e: IHC×400. f: CISH×400).

4. Discussion

PAL is a type of non-Hodgkin's lymphoma of mainly the B-cell phenotype. It develops in the pleural cavity in patients with longstanding histories of pyothorax. This disease was originally reported by Aozasa in 1987 and the term "pyothorax-associated lymphoma (PAL)" was proposed [4]. A nationwide study in Japan collected 37 cases of pleural lymphoma. The EBV genome was detected in lymphoma cells in all PAL by polymerase chain reaction, in situ hybridization, and immunohistochemistry [5]. As early as 1993, Yamabe et al. [6] discovered that PAL is closely associated with EBV infection. Sorting out the reported literatures indicated that the EBV genome is detectable in the lymphoma cells of most PAL cases [7, 8].

The endemic prevalence of PAL is different in whole world. Due to the use of artificial pneumothorax for the treatment of tuberculosis. PAL cases are most commonly reported in Japan. Nakatsuka et

al. [1] reviewed clinical and pathological findings in 106 patients with PAL in the age range of 46–82 years (median, 64 years) collected through a nationwide survey in Japan. A male/female ratio was 12.3:1. All patients had history of pyothorax caused by artificial pneumothorax. In an situ hybridization study, 70% of patients were positive for Epstein-Barr virus (EBV).

Most cases of PAL develop from a pyothorax caused by an artificial pneumothorax created during the treatment of pulmonary tuberculosis or tuberculous pleuritis, and it is attributed to chronic inflammation of the pleura. However, from the cases we have come into contact with, PAL has been shown not to be restricted to pulmonary tuberculosis or similar conditions; it can develop from any type of empyema. Taniguchi et al. [9] reported the first case of PAL arising from a posttraumatic empyema. This case indicates that posttraumatic empyema could be a risk factor for the development of malignant lymphoma, and caution should be exercised to prevent chronic empyema when treating patients with chest injuries. Our case is also PAL arising from posttraumatic empyema. This patient was EBV-positive, as in most cases of PAL.

Another interesting finding is that the patient was CD20-negative. Consistent with our case, in the first PAL patient with empyema after trauma, CD3 and CD20 of the primary tumor were negative. There have also been other case reports of CD20-negative PAL. Kenji Fukuno et al. [10] proposed that PAL probably originates from B cells, CD20 is then lost during B cell differentiation into plasma cells. Therefore some CD20-negative pyothorax-associated B cell lymphomas are expected.

PAL is mostly associated with underlying diseases. The imaging features of primary malignant lymphomas originating from the chest wall are the tumor spreading along the pleura, sometimes involving the ribs but mostly maintaining rib structure. However, lymphomas with similar features are also recognized in other chronic inflammatory conditions. PAL can be difficult to diagnose using imaging alone. This disease is occasionally misdiagnosed as lung cancer or a form of tuberculous abscess. Rare, empyema-associated malignant tumors and malignant tumors accompanied by empyema that should be considered in the distinguishing diagnosis of pyothorax-associated lymphoma [11]. Wan and Lan et al. [12] also pointed out that PAL needs to be distinguished from primary exudative lymphoma. Research have shown that specific imaging features of pyothorax-associated lymphoma, such as symmetric growth pattern of a mass at the margin of chronic empyema appear to be distinct from other empyema-associated malignant lesions [11]. In this case, CT examination revealed chronic empyema on the right side of the chest with subphrenic infection. The lesion protruded inward into the right lower lobe, adjacent to the diaphragm and liver compression, local thickening of the pleura not consistent with nodular calcification. However, it is still not enough to confirm the diagnosis of PAL directly by imaging examination.

5. Conclusion

Pathological morphology is vital important to make a final diagnosis and avoid misdiagnosis or miss diagnosis. This case is described by a combination of imaging and pathological examinations that reports the clinical pathological diagnosis and treatment process of a single case of pyothorax-associated lymphoma, and it provides some diagnostic information regarding the rare disease of PAL.

6. Funding

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