

Case Report

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A rare case of primary pulmonary diffuse large B cell lymphoma with CD5 positive expression

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Abstract: Primary pulmonary diffuse large B-cell lymphoma (PPDLBCL) is extremely rare. Its clinical symptoms and signs are nonspecific, and imaging features also have not yet been well-defined. Further description is important for the diagnosis and treatment of PPDLBCL. Herein, we reported a case of a patient who suffered from bilateral chest pain and dyspnea. Computed tomography (CT) of chest demonstrated bilateral lung mass, consolidations and reverse halo sign, while consolidations and reverse halo sign are uncommon according to previous reports. Tissue samples were taken by CT guided needle biopsy. The histological samples showed PPDLBCL. This case was special in view of positive expression of CD5. After the case was treated by cyclophosphamide pirarubicin vindesine dexamethasone (CHOP) chemotherapy for six courses, her clinical symptoms were partially alleviated, while CT showed progression disease. This case report highlights

different imaging features and characteristics of molecular biology, and reviews study progress of PPDLBCL.

Keywords: Primary pulmonary diffuse large B cell lymphoma; CD5; Multidetector Computed Tomography

1 Introduction

Primary pulmonary lymphoma (PPL) is rare. The majority of PPLs are mucosa-associated lymphoid tissue lymphoma. Primary pulmonary diffuse large B-cell lymphoma (PPDLBCL) is particularly rare and occurs only in 10% cases of primary pulmonary NHL [1]. To our knowledge, PPDLBCL has been reported only in case reports or small sample studies [2-4]. Thus, its clinical characteristics, treatment and prognosis have not been clearly delineated. We present a case of PPDLBCL which had multiple imaging manifestations, and this case was special because of its positive expression of CD5. In addition, we briefly review the literature related to PPDLBCL.

2 Case report

A 71-year-old female was taken to our hospital after experiencing bilateral chest pain and dyspnea for 20 days. She had no other complaints, such as fever, cough and bloody sputum. The patient had a history of arthritis pauperum for 20 years, and denied traditional Chinese medicines for treatment. She had no personal history of smoking and family history of cancer. On clinical examination, no palpable lymph nodes and hepatosplenomegaly were found. There was no obvious rale in bilateral lungs. Computed tomography (CT) of the chest demonstrated a high density shadow in the right middle lower lobe and left lower lobe, and air bronchogram was obvious (Figure 1 C). Bilateral hilar masses exits (Figure 1 E) and low density area was visible (green arrow). Mediastinal lymph nodes enlarged.

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The crescent shaped liquid density was in right pleura (Figure 1 D). CT of abdomen showed no abnormalities. Bone marrow biopsy showed no infiltration. CT-guided transthoracic core needle biopsy was performed (Figure 2). Pathological section demonstrated diffuse large B cell infiltration in small fibrous tissue. Immunohistochemical staining showed Mum-1(-), Ki-67 index 70%, Cyclin D1(-), CD43(+), CD5(+), Bcl-2(-), CD3(-), CD79a(+), CD23(-), CD10(-), Bcl-6(-), Syn(-), CD20(++), CD56(-) (Figure 3). Based on these findings, PPDLBCL was diagnosed. Due to personal reasons, the patient underwent cyclophosphamide pirarubicin vindesine dexamethasone (CHOP) chemotherapy without rituximab, which was planned to be repeated every 21 days for 6 cycles. Following the administration of 6 cycles of CHOP chemotherapy, chest pain and dyspnea were alleviated. Nevertheless, CT demonstrated bilateral masses and consolidation partially diminished (Figure 4 E F), while a new mass emerged in the right upper lobe (Figure 4 A D blue arrow). Unfortunately, at 9 months of follow-up, the patient died.

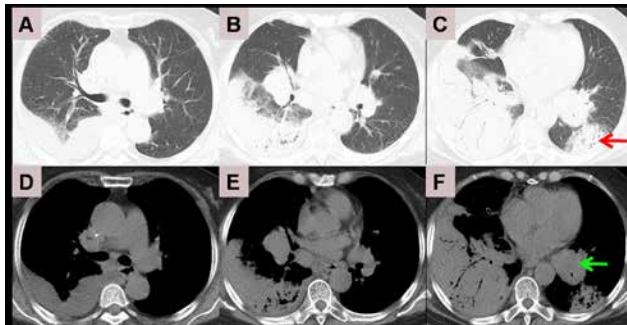


Figure 1: Initial chest computed tomography. Lung window: A B C; mediastinal window: D E F. Red arrow indicates reversed halo sign, and green arrow indicates low density area in left hilar mass.



Figure 2: Computed tomography guided transthoracic core needle. Puncture needle is in the consolidation area of right lower lobe.

Ethical approval: The research related to human use has been complied with all the relevant national regulations, institutional policies and in accordance the tenets of the Helsinki Declaration, and has been approved by the authors' institutional review board or equivalent committee.

Informed consent: Informed consent has been obtained from all individuals included in this study.

3 Discussion

PPDLBCL is extremely rare in primary lung malignant lymphomas. Some case reports and small sample studies delineate the clinical characteristics. Its respiratory symptoms are nonspecific [5]. The key features of CT are single or multiple solid pulmonary nodules or masses, cavitation and mediastinal lymph node enlargement (3, 6 and 7). Consolidation is seldom [5]. In addition to enlarged mediastinal lymph nodes, necrosis in right hilar mass is also

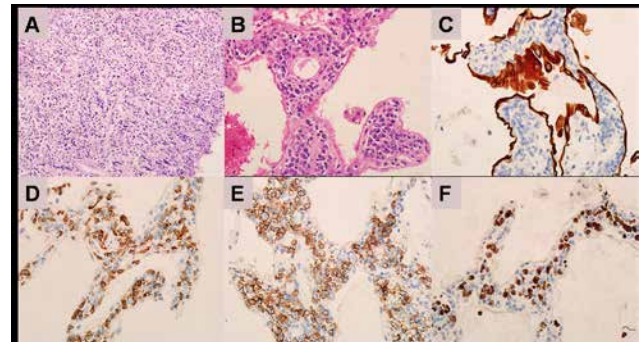


Figure 3: Hematoxylin and eosin (H&E) staining and immunohistochemical staining. A: Alveolar septal broadens, and tumor cells infiltrate (HEx100); B: The tumor cells are large and rich in cytoplasm, the karyotype is slightly irregular, and nucleolus is visible (HEx400); C: immunohistochemical staining for cytokeratin (CK) (x400); D: immunohistochemical staining for CD-5 (x400); immunohistochemical staining for CD20 (x400); immunohistochemical staining for Ki-67 (x400).

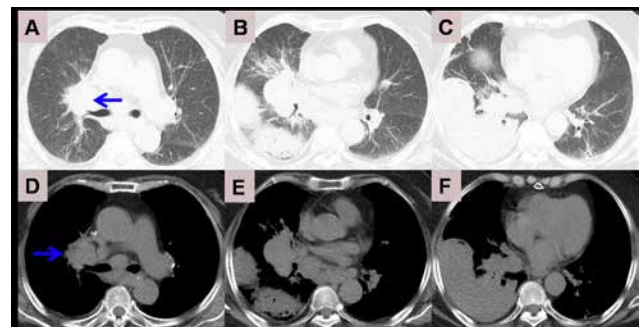


Figure 4: Chest computed tomography after 6 cycles of chemotherapy. A B C; mediastinal window: D E F. Blue arrow indicates newly emerging mass.

visible in this case (Figure 1 F green arrow). Consolidation was obvious. Pathological specimen was from consolidation area (Figure 2), so it indicated that PPDLBCL could cause consolidation. Notably, Marchiori *et al* reported the reversed halo sign (RHS) can be useful for differentiating invasive pulmonary aspergillosis from pulmonary lymphoma [8]. Nevertheless, we also found RHS in this case (Figure 1 F red arrow). In a word, imaging of PPDLBCL is complex and diverse.

Now that the imaging of this case was different from common cases' imaging, we further observed its biological characteristics. Immunohistochemical staining demonstrated positive expression of CD5. CD5+ DLBCL represents 5% to 10% of DLBCLs [9], and has some differences from other typical DLBCLs. CD5 molecule can promote B-cell survival. The precise mechanisms with which expression of CD5 alters the behavior of B cells in DLBCL are unclear [10]. In general, these patients have more aggressive clinical courses and poor diagnosis. At present, we have not found the report of imaging characteristics of CD5+ PPDLBCL.

The CHOP chemotherapy regimen has been the mainstay of therapy [11]. The addition of the anti-CD20 monoclonal antibody rituximab to this chemotherapy dramatically improved the outcomes, resulting in a 16% absolute improvement in 10-year overall survival in elderly patients ≥ 60 years of age [12]. Additional trials further demonstrate the benefit of rituximab and establishment of R-CHOP as the standard of care [13]. Rituximab based regimens can improve the outcomes of patients with CD5+ DLBCL, but this improvement in outcome is inferior to that seen in patients with CD5- DLBCL. Due to personal reasons, CHOP chemotherapy was performed without rituximab in this case. The result showed that simple CHOP chemotherapy is not ideal. Further studies are warranted to investigate a more effective therapy.

Conflict of interests: No authors report any conflict of interest.

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