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A Case of Lung Involvement in Extranodal NK/T cell lymphoma

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Abstract

Background: Lung involvement is rare in Peripheral T cell lymphoma and lack of sufficient clinical study. The authors describe a rare case of lung involvement in a 41-year-old male patient with Extranodal NK/T cell lymphoma.

Methods: Hematologic investigation and left upper lobectomy of the lung were performed.

Results: The patient died 2 months after diagnosis of the disease.

Conclusions: Lung biopsy is important to confirm a diagnosis of primary pulmonary lymphoma. Lung involvement in T cell lymphoma as a prognostic factor needs further studies.

Keywords: Extranodal NK/T cell lymphoma, Lung lobectomy, Lung involvement

CASE REPORT

A 41-year-old man, presenting with cough, sputum and fever over a period of 3 months, was admitted to our hospital on June 25, 2018. He had no history of dyspnea, bleeding manifestations, abdominal pain or weight loss during the last 3 months. There was no history of exposure to toxic drugs, tuberculosis or pets. His personal history and family history was non-contributory.

Physical examination at the time of admission showed normal respiratory rate and some wet whales in the left lung. Neither hepatomegaly nor splenomegaly was found.

Laboratory investigations yield following results: hemoglobin (Hb) 147 g/L; white blood cell (WBC) 10.84 x 109/L, with 18.0% lymphocytes and 66.9% neutrophils; platelet (Plt) 261 x 109/L. Renal and liver biochemical parameters were within normal limits. His serum lactate dehydrogenase and β2-microglobulin level were not tested. Chest CT showed left lower lobe lesions, considering tumors with peripheral obstructive pneumonia, left lower invaded pleura adhesion, multiple slightly larger lymph nodes in the left hilum and mediastinum. Small nodules in the dorsal part of the right lower lobe, a few fibrous foci in the lingual part of the left upper lobe and the medial part of the right middle lobe. Abdominal ultrasonography was normal. Left upper lobectomy of the lung was diffusely infiltrated by abnormal nested or patchy lymphoid cells with large patches of necrosis were seen in lung tissues. Vascular invasion and alveolar septum infiltration were observed. Tumor cells were positive for CD3, CD7, CD2, CD4, CD56, CD163, CK7, TTF-1, GB, TIA, Vimt, EBER, Ki-67 (70%), but were negative for CD5, CK5/6, CD8, CD20, P40(Figure 1). NK/T cell lymphoma was considered. A bone marrow (BM) aspirate was denied by the patient. Total body PET-CT revealed hyper metabolism masses in the left lower lobe of the patient's lung without hyper metabolism of other suspected parts of the body.

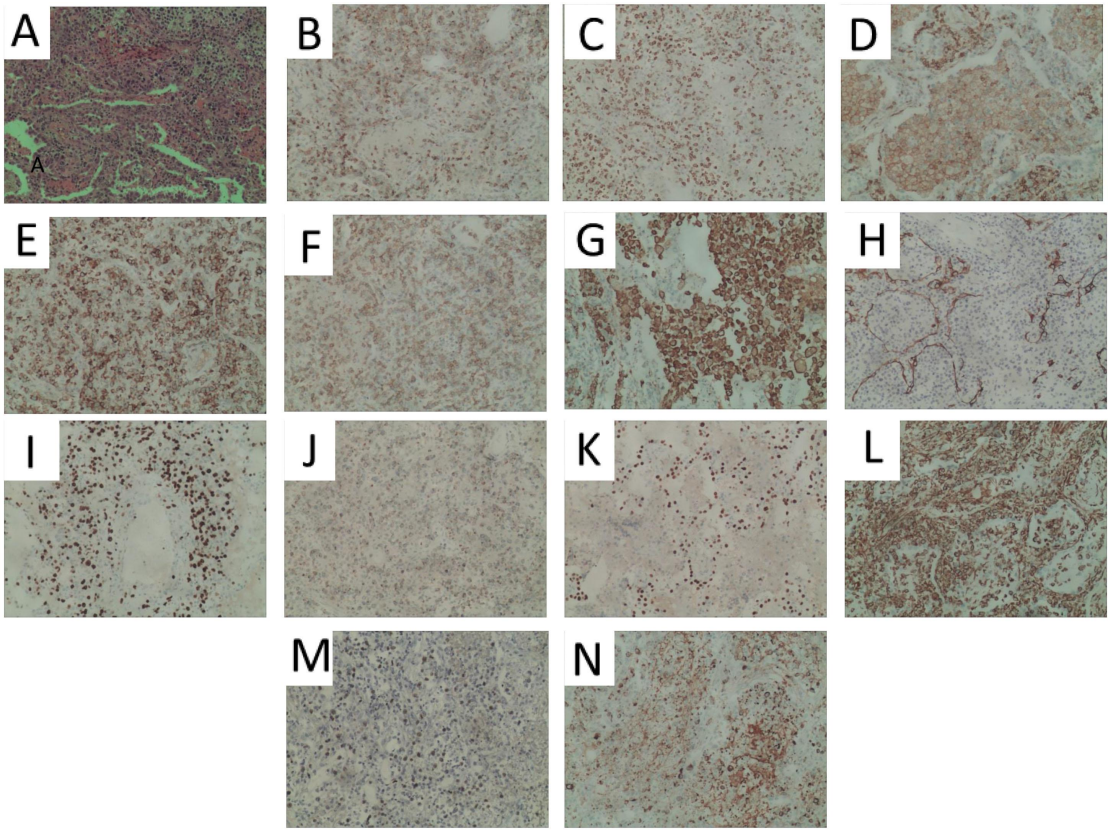

Figure 1. (A) Pulmonary biopsy revealed diffuse infiltration of uniform, medium-size lymphoid cells in the bronchial mucosa and interstitial lung tissue (H&E, x200). (B - N) Immunohistochemical staining showed the infiltrated cells were positive for CD2 (B, x200), CD3 (C, x200), CD4(D, x200) , CD 7(E, x200) CD 56(F, x200), CD 163(G, x200), CK7(H, x200) , Ki-67 (I, x200), TIA(J, x200), TTF-1(K, x200), Vimt(L, x200) , EBER (M, x200), GB (N, x200).

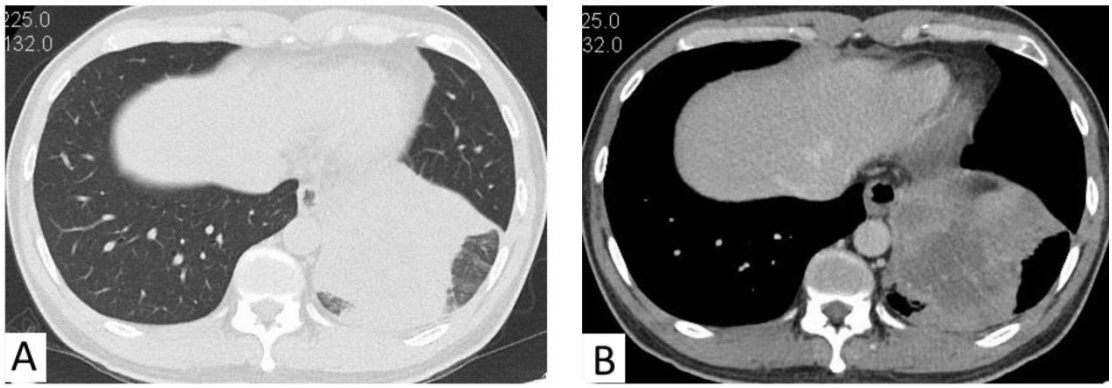

Figure 2. Chest CT revealed left lower lobe lesions, considering tumors with peripheral obstructive pneumonia, left lower invaded pleura adhesion, multiple slightly larger lymph nodes in the left hilum and mediastinum.

A diagnosis of Extranodal NK/T cell lymphoma with primary lung involvement was made. The patient refused further treatment and died 2 months after the diagnosis.

DISCUSSION

Extranodal NK/T cell lymphoma(ENKL) is an extranodal T cell lymphoma that most common in Asia and Central and South America. Nasal type Extranodal NK/T cell lymphoma is most common and accounts for 5-10 percent of all non-Hodgkin lymphoma[1-2]. But the primary lung involved Extranodal NK/T cell lymphoma is so rare that there are only some case reports around the world and the lack of definite known incidence[3]. The main manifestations of pulmonary involved symptoms are cough, expectoration,dyspnea, chest pain, etc. There is no significant difference from other diseases such as pneumonia and lung cancer[4]. The main systemic symptoms are fever, fatigue and weight loss. The pathogenesis of ENKL is poorly understood but is related in part of Epstein-Barr birus(EBV). NK/T cell associated antigens are variably expressed CD2, CD56, CD3, CD4, CD8 and EBV encoded RNA(EBER) generally negative for B cell associated antigens[3,5-6]. Asano et al. reported the first cases of T-cell non-Hodgkin's lymphoma (NHL) in the lung in 1991[7]. Chu-Chun Chien *et.al* reviewed 8 cases of Primary Pulmonary ENKL from 1998- 2015[3]. The age of onset ranged from 17 to 80, and 5 cases were less than 50 years old. The female/male ratio was 5:3. Based on the immunohistochemistry of pulmonary, our patient was definitively diagnosed as Extranodal NK/T cell lymphoma, with the positive expression of CD3, CD56, EBER and the negative expression of CD20.

The main CT findings of lung lesions of T cell leukemia or lymphoma including GGO, centrilobular nodules, and thickening of the bronchovascular bundles in the peripheral lung, multiple bilateral pulmonary nodules and patch shadow, localized Ground Glass Opacities With Multiple Pulmonary Small Cysts (''Alloy Wheel'' Appearance), crazy paving pattern and lung consolidation[8-10]. Our patient presented with pulmonary mass and multiple lymph node metastases in mediastinum, which may be misdiagnosised as lung cancer. Pulmonary lobectomy revealed NK/T cell lymphoma.

The clinical course of Primary Pulmonary ENKL is aggressive [3]. Most patients may die within months. Our patient's condition aggravated rapidly, even had no chance of HSCT, which indicated a poor prognosis.

CONCULSION

In summary, lung involvement is rare in extranodal NK/T cell lymphoma. Pulmonary biopsy is important to confirm a correct diagnosis. Lung involvement in extranodal NK/T cell

lymphoma as a prognostic factor needs further studies.

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References:

[1] Jaffe ES, Chan JK, Su IJ, et al. Report of the Workshop on Nasal and Related Extranodal Angiocentric T/Natural Killer Cell Lymphomas. Definitions, differential diagnosis, and epidemiology. Am J Surg Pathol 1996; 20: 103-111. (PMID:8540601)

[2] Chihara D, Ito H, Matsuda T, et al. Differences in incidence and trends of haematological malignancies in Japan and the United States. Br J Haematol 2014; 164: 536-545. (PMID:24245986)

[3] Chien CC, Lee HS, Lin MH, Hsieh PP. Primary extranodal natural killer/T-cell lymphoma of bronchus and lung: A case report and review of literature. Thorac Cancer 2016; 7: 140-144. (PMID:26816549)

[4] Chien CC, Lee HS, Lin MH, Hsieh PP. Primary pulmonary peripheral T-cell lymphoma: A case report and review of the literature. Thorac Cancer 2014; 5: 104-107. (PMID:26816549)

[5] Arber DA, Weiss LM, Albújar PF, Chen YY, Jaffe ES. Nasal lymphomas in Peru. High incidence of T-cell immunophenotype and Epstein-Barr virus infection. Am J Surg Pathol 1993; 17: 392-399. (PMID:8388175)

[6] Elenitoba-Johnson KS, Zarate-Osorno A, Meneses A, et al. Cytotoxic granular protein expression, Epstein-Barr virus strain type, and latent membrane protein-1 oncogene deletions in nasal T-lymphocyte/natural killer cell lymphomas from Mexico. Mod Pathol 1998; 11: 754-761. (PMID:9720504)

[7] Asano Y, Ohshima K, Yoshizawa S, Shiraishi G, Kato K. Modified CHOP in an aged patient with non-Hodgkin's lymphoma and probable extensive pulmonary involvement. Int J Hematol 1991; 54: 327-329. (PMID:1777606)

[8] Sakashita A, Ashizawa K, Minami K, et al. Localized ground glass opacities with multiple pulmonary small cysts in adult T-cell leukemia or lymphoma: an "alloy wheel" appearance. J Thorac Imaging 2009; 24: 321-324. (PMID:19935228)

[9] Yang L, Feng W, Chen C, et al. Primary pulmonary T-cell lymphoma mimicking pneumonia: A case report and literature review. Exp Ther Med 2016; 12: 365-368. (PMID: 27347063)

[10] Fraser T, Nagarur A. Pulmonary involvement of peripheral T-cell lymphoma manifesting as crazy paving pattern. Proc (Bayl Univ Med Cent) 2015; 28: 59-61. (PMID:25552801)