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Lung: Case Report

Localized Synchronous Pulmonary Langerhans Cell Sarcoma and Langerhans Cell Histiocytosis

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A synchronous presentation of pulmonary lesions of Langerhans cell sarcoma (LCS) and Langerhans cell histiocytosis (LCH) is extremely rare. We report the case of a 57-year-old man with 3 pulmonary nodules, 1 in the left lung and 2 in the right lung. In a 2-stage operation, he first underwent a left segmentectomy, and the nodule was diagnosed as LCS. Thereafter, 1 of the 2 right pulmonary nodules disappeared. The residual nodule was resected by right segmentectomy and was diagnosed as LCH. This is a report of complete resection of localized, synchronous pulmonary lesions of LCS and LCH.

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A synchronous presentation of pulmonary lesions of Langerhans cell sarcoma (LCS) and Langerhans cell histiocytosis (LCH) is extremely rare. We report the case of a 57-year-old man in whom LCS and LCH manifested simultaneously as pulmonary nodules in the left and right lungs, respectively, and were completely resected.

A 57-year-old man with a 37-pack-year active smoking history was referred to our hospital with an abnormal opacity on his chest radiograph. Chest computed tomography (CT) revealed a 23-mm nodule in the left upper lobe, a 20-mm nodule in the right lower lobe, and a 6-mm cavitating nodule in the right middle lobe (Figure 1a). ^{18}F-Fluorodeoxyglucose positron emission tomography/CT (FDG PET/CT) revealed a significant uptake in the left upper lobe nodule (maximum standardized uptake value [SUVmax], 4.8), right lower lobe nodule (SUVmax, 2.9), and left hilar lymph nodes (SUVmax, 2.6; Figures 1b, 1c). Bronchoscopic findings of the left upper lobe nodule were suggestive of adenocarcinoma. A 2-stage operation was planned first for the left pulmonary nodule (cT1 cN1 M0, c-stage IIB, TNM classification, 8th edition) followed by the right pulmonary nodules (cT1b N0 M0, c-stage IA2 and cT1a N0 M0, c-stage IA1). His preoperative pulmonary function test showed an obstructive ventilatory defect (forced expiratory volume in 1 second, 3200 mL; forced expiratory volume in 1 second/forced vital capacity ratio, 67.25%; vital capacity, 132.5% predicted; and diffusing capacity of lung for carbon monoxide, 76.2%).

The intraoperative findings confirmed that the tumor was infiltrating the left lower lobe across the fissure. The hilar lymph nodes that showed a high uptake on FDG PET/CT were enlarged and had infiltrated into the pulmonary artery from A5 to A8. As we could not separate these lymph nodes from the pulmonary artery, we performed a left lingulectomy and S8 segmentectomy. The tumor size was 38 × 23 × 12 mm. The postoperative pathologic diagnosis was LCS with lymph node metastasis; histopathologic examination revealed proliferation of cells with acidophilic granular cytoplasm and rumpled tissue nuclei and kidney bean-shaped nuclei (Figure 2a). The tumor showed dense cellularity. On immunohistochemical analysis, the tumor cells were positive for S100, CD1a (Figures 2b, 2c), and CD163, and the MIB-1 labeling index was 30% to 40% (Figure 3b). Mitotic activity was 15 to 22 mitotic figures per 10 high-power fields; however, the region of the lymph nodes infiltrating into the pulmonary artery showed a mass of Langerhans cells (Figure 2d).

Two months after the initial operation, follow-up chest CT revealed that the right lower lobe nodule had not changed in size, whereas the right middle lobe nodule had disappeared. Bone scintigraphy and bone marrow aspiration did not reveal LCS. The patient underwent thoracoscopic S6 segmentectomy of the right lower lobe. The tumor size was 21 × 17 × 13 mm. The postoperative pathologic diagnosis was LCH (Figure 3a).

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Ann Thorac Surg Short Reports
2023;1:231-234

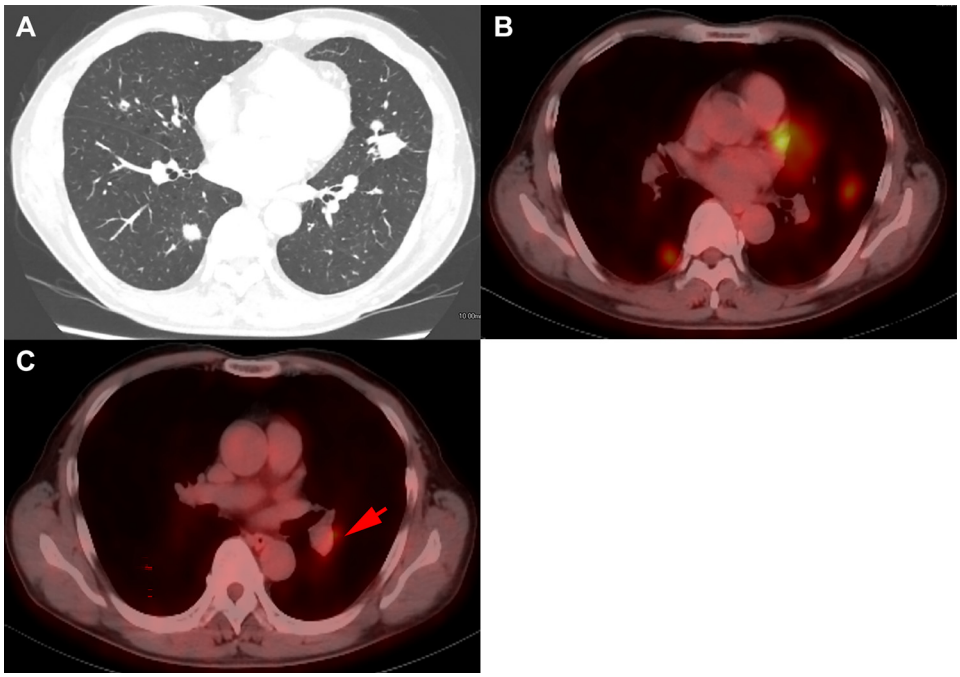

FIGURE 1 Preoperative imaging findings. (A) Chest computed tomography showing a nodule in the left lower lobe and right middle and lower lobes. (B, C) Fluorodeoxyglucose positron emission tomography/computed tomography showing uptake in the pulmonary nodules of the left upper lobe, right lower lobe, and left hilar lymph nodes (arrowhead).

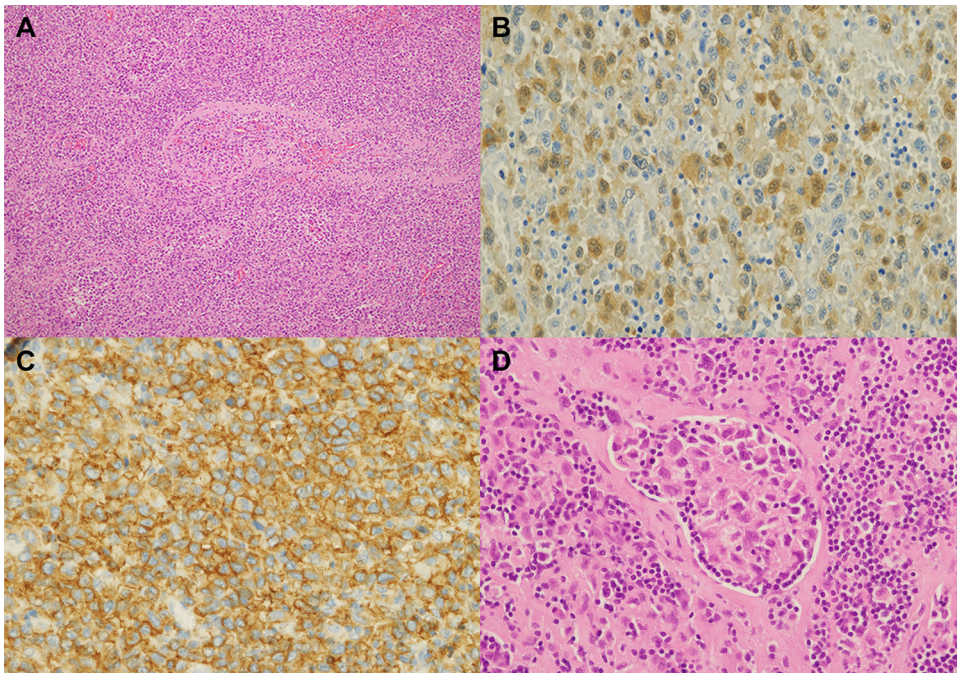

FIGURE 2 (A) Hematoxylin and eosin staining of the left upper lobe nodule. It shows dense cellularity. (B, C) Immunohistochemical analysis of the nodule. The tumor stains positive for (B) S100 and (C) CD1a. (D) Hematoxylin and eosin staining of the hilar lymph node. It shows a mass of Langerhans cells.

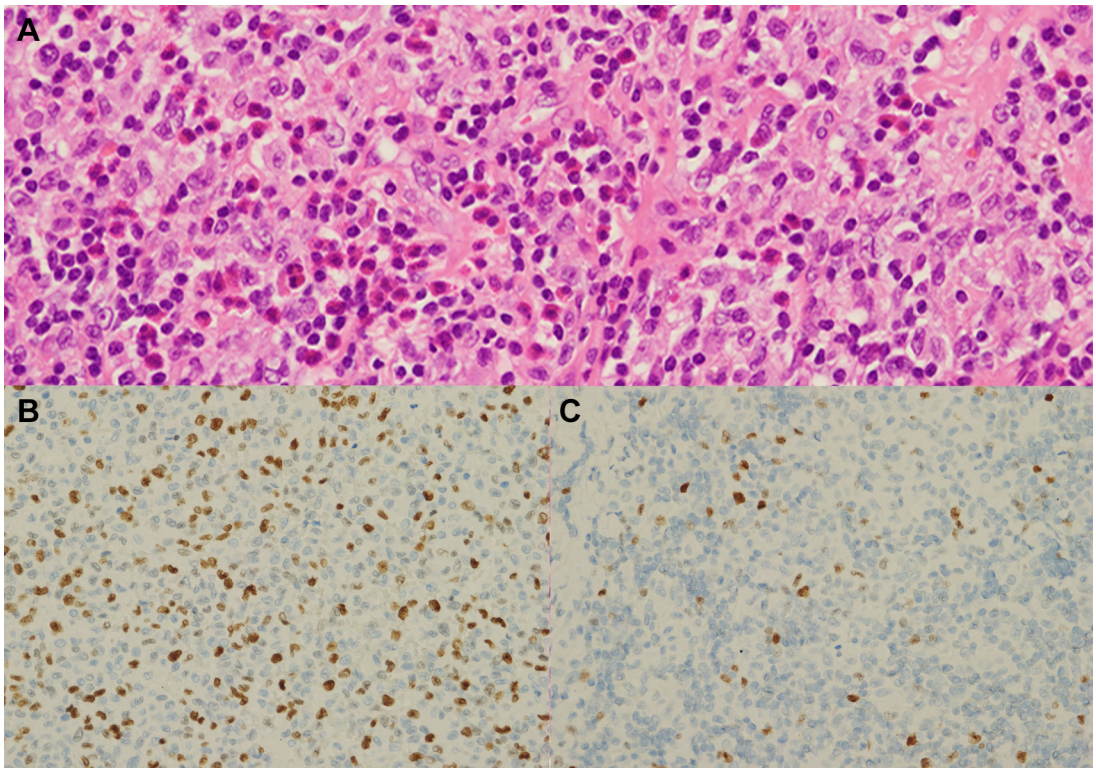

FIGURE 3 (A) Hematoxylin and eosin staining of the right lower lobe nodule. It shows minimal cellular atypia. (B, C) Comparison of MIB-1 in the pulmonary nodules. (B) MIB-1 labeling index of the left upper lobe nodule is 30% to 40% and (C) that of the right lower lobe nodule is 12%.

Mitotic activity was low (<1 mitotic figure per 10 high-power fields). On immunohistochemical analysis, the tumor cells were positive for S100, CD1a, and CD163, and the MIB-1 labeling index was 12% (Figure 3c).

COMMENT

Langerhans cells are antigen-presenting dendritic cells. Tumors derived from these cells are rare and are divided into 2 subgroups, LCH and LCS, according to the World Health Organization classification.[1] LCH is a benign clonal proliferation of Langerhans cells, whereas LCS is neoplastic histiocytosis, in which the cells clearly exhibit malignant characteristics. LCH and LCS in particular are multisystem diseases. Lesions are found in the bones, lungs, pituitary gland, thyroid gland, lymph nodes, and skin.[1]

Chest CT in LCH generally reveals multiple cysts and micronodules of size <10 mm predominantly in the middle and upper lobes.[2] A localized pulmonary nodule of size >20 mm, as in our case, is rare. The pulmonary lesions of LCS are larger than those of LCH and are usually multiple and bilateral.[1,3] It is reported that FDG PET/CT shows an uptake in the pulmonary lesions of both LCH and LCS.[2,4] It is difficult to distinguish between them by preoperative imaging findings. Therefore, biopsies involving surgical procedure or bronchoscopy are usually required for an accurate diagnosis to be made. The primary lesion of LCS is often in the organs other than the lungs, such as the skin and lymph nodes. We need to consider the biopsy specimens from these lesions in these cases.

Treatment strategies for LCH and LCS have not yet been established. The pulmonary nodules of LCH are related to smoking, and early smoking cessation is important in LCH treatment.[2] In our case, the right middle lobe nodule disappeared after smoking cessation. The relationship between LCS and smoking is not clear. Patients with multisystem diseases of LCS received chemotherapy.[5] In our case, no chemotherapy was planned because the lesions were completely resected.

Five cases of LCS with pulmonary lesions at diagnosis have been reported.[1,3,5,6] In only 2 cases, including ours, isolated pulmonary lesions were seen.[3] In both these cases, the patients underwent pulmonary resection for treatment. Lee and coworkers[3] reported that lobectomy was performed for a 4-cm lung mass that tended to grow; however, residual multiple, small, bilateral lung

Ann Thorac Surg Short Reports
2023;1:231-234

nodules were seen. In addition, LCS and LCH with pulmonary lesions were diagnosed in the same patient in only these 2 cases.

This is a case of complete resection of localized, synchronous pulmonary lesions of LCS and LCH. Considering that LCH and LCS are systemic diseases, close follow-up for systemic manifestations is needed regardless of treatment.

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DISCLOSURES

The authors have no conflicts of interest to disclose.

PATIENT CONSENT

Informed consent was obtained from the patient and the patient's family.

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