Thyroid Involvement in Pulmonary Langerhans Cell Histiocytosis

Michihiro Uchiyama¹, Reiko Watanabe², Ichiro Ito² and Takashi Ikeda¹

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A 21-year-old male discovered that his thyroid gland was becoming enlarged in October 2008. Two months later, chest radiography during a routine physical examination revealed an abnormal shadow and he was referred to our hospital for a complete medical examination. He had experienced asthma-like wheezing about once a month for the past 2 years but he was not receiving any medication. He had smoked several cigarettes per day since the age of 20 years old. Physical examination revealed no abnormalities except goiter. Blood analyses showed a white blood cell count of 4,710/μL comprising 47% neutrophils, 42% lymphocytes, 3% eosinophils, and 8% monocytes. Biochemical analysis showed slightly increased levels of serum C-reactive protein (1.30 mg/dL; normal range, <0.20 mg/dL). Thyroid function was decreased (free T4: 0.66 ng/dL; normal range, 0.90-1.70 ng/dL), but antithyroid antibodies were negative. Bone marrow aspirates appeared morphologically normal. Chest radiography revealed reticulonodular infiltration and computed

¹Division of Hematology and Stem Cell Transplantation, Shizuoka Cancer Center, Shizuoka and ²Division of Pathology, Shizuoka Cancer Center, Shizuoka
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Correspondence to Dr. Michihiro Uchiyama, mi.uchiyama@scchr.jp
tomography (CT) of the chest demonstrated multiple small partly cavitated nodules, throughout both lung fields (Picture 1). Contrast-enhanced CT also demonstrated diffusely enlarged thyroid gland with spotted low-density areas. Positron emission tomography CT scans revealed increased uptake in the thyroid gland and in both lung fields (Picture 1). Magnetic resonance imaging showed no significant abnormality in other sites such as bone and brain. Pulmonary function abnormalities included reduced diffusing capacity of the lungs for carbon monoxide and a low vital capacity. The histology of the thyroid gland biopsy revealed many aggregates of mononuclear histiocytoid cells with scattered neutrophils or eosinophils, around which there were infiltrating small lymphocytes, fibrosis and atrophic or dilated thyroid follicles (Picture 2A, Hematoxylin and Eosin staining, ×40). These histiocytoid cells had some deep nuclear folds forming nuclear shapes like the kidney of a fetus; the cells were immunohistochemically stained positively with anti-CD1a antibody (Picture 2B, CD1a, ×40). They were also positive for CD68 and S-100 protein, and negative for epithelial membrane antigen, CD3, CD5 and CD20. These results suggested langerhans cell histiocytosis (LCH). Findings of a lung biopsy specimen were also consistent with a diagnosis of LCH.

LCH in adults is a rare disorder that usually presents as a unifocal disease clinically involving multiorgan systems such as bone, hypothalamus, lung, skin and mucosa (1, 2). The thyroid gland is rarely involved in LCH particularly in adults (1, 2). To the best of our knowledge, this patient is the first Japanese adult to present with thyroid involvement in pulmonary LCH. LCH involving the thyroid gland has a wide differential diagnosis, extending from benign inflammatory conditions to several types of neoplasms. Therefore, a high index of suspicion is essential for the correct diagnosis.

References