Langerhans cell histiocytosis (LCH) is a monoclonal disease of histiocytes, may involve several organ systems but rarely primarily involves the thyroid gland. This report presents an extremely rare case of LCH of the thyroid in a 3-year-old boy who presented with a neck mass for several weeks. LCH of the thyroid should be considered in the differential diagnosis of a child with a thyroid mass. Pulmonary examination should be done in these patients.

Key words: Langerhans cell, histiocytosis, thyroid gland

Langerhans cell histiocytosis (LCH) is a rare disease involving clonal proliferation of langerhans cells, abnormal cells deriving from bone marrow and capable of migrating from skin to lymph nodes. Clinically, its manifestations range from isolated bone lesion to multisystem disease (1).

LCH is part of group of clinical syndromes called histiocytoses, which are characterized by abnormal proliferation of histiocytes (an archaic term for activated dendritic cells and macrophages). These diseases are related to other forms of abnormal proliferation of white blood cells, such as leukemias and lymphomas (1-5).

The disease has gone by several names, including Hand-Schuller-Christian disease, Abt-Letterer-Siwe disease and Histiocytosis X, until it was renamed in 1985 by Histiocyte Society (5).

However, thyroid involvement in LCH is very rare in children (1, 3, 4, 6). Few cases have been reported of thyroid gland infiltration by LCH and isolated involvement (7). This report is a rare case of LCH with thyroid and liver involvement in a child who presented with progressive anterior neck mass enlargement.

CASE REPORT

A 3-year–old boy was referred for a history of progressive neck mass of 5 month. There were no fever, nausea, vomiting, or dizziness. Physical examination revealed a 3.5 x 3.5 cm non-tender mass over the anterior middle neck. Liver was enlarged. Laboratory finding showed a leukocyte count of 9700/ mm³, with differentials of 41% neutrophils, 54% lymphocytes, 4% monocytes, and 1% eosinophils. The hemoglobin level and platelet count were normal. TSH was 1.4 micro gram /dl, T4 was 7.6 micro gram /dl, and T3 was 0.9 micro gram /dl. SGOT was 57 U/L and SGPT was 66 U/L. Serum electrolytes, creatinine, albumin, total bilirubin, protein, and osmolality levels were normal.

Computed tomography (CT) of the neck revealed large mass originate from left lobe of thyroid with mass effect over trachea. CT
of the thorax revealed large superior mediastinal mass extending from neck to mediastinum with mass effect over trachea from left side (fig. 1). Liver enlargement (span=145 mm) was detected by abdominal sonography and CT scan. Chest radiograph showed mass in superior mediastinum and lower neck more in left side and lungs were normal (fig. 2). Bone survey revealed bone density was increased, and well defined lytic lesion in skull was noted (fig. 4).

Open surgery was performed and pathology findings showed proliferation of Langerhans histiocytes with nuclear grooves in a background of scattered eosinophils (fig. 3).

Immunohistochemical staining for S100 and CD1a were positive, bone marrow aspiration, and flow cytometry revealed negative findings.
The patient subsequently underwent chemotherapy, included prednisolone, vinblastine, methotrexate, and 6-mercaptopurine. The treatment resulted resolution neck mass and liver size. Follow-up neck CT showed resolution of the neck mass, whereas abdomen CT showed liver size was normal.

DISCUSSION

Involvement of the thyroid gland by LCH is rare (8). Thyroid gland infiltration remains uncommon with about 34 cases reported in the medical literature, the vast majority presenting as multisystem disease (3, 4). In children, LCH of the thyroid is usually part of a multisystemic disease (9, 10). Due to its rarity, diagnosing LCH of the thyroid can be challenging. It can be clinically confused with the far more common benign goiters, undifferentiated carcinoma, lymphoma, lymphocytic thyroiditis, and chronic granulomatous thyroiditis. Patient with thyroid LCH are euthyroid initially, but may have biochemical signs of primary hypothyroidism or hyperthyroidism (9, 10, 11). The key to diagnosis of LCH is to identify the pathologic Langerhans cells that resemble the normal Langerhans cells of the skin, except they are not dendritic in morphology. They do demonstrate a characteristic nuclear groove and positive immunostaining with CD1a, S100 (4, 6).

Additional investigations such as thoracic CT, abdominal sonography, bone scan, and bone marrow aspiration are suggested because of the importance of distinguishing isolated thyroid LCH from multisystemic cases (11). Prolonged follow up is recommended because thyroid LCH may involve other organs or precede a multisystemic disease for several months (12).

Reviewing literature, there are cases with thyroid involvement and other sites, such as skin, lymph nodes, otitis media, and central nervous system (6, 9-12). Prognosis appears to be closely related to the presence or absence of multiorgan involvement. Isolated thyroid LCH has excellent survival rate, good prognosis, and no recurrence during follow up (13).

LCH Rarely involves the thyroid gland in children. Thyroid LCH should be suspected in a child with thyroid mass involvement.

REFERENCES


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