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CASE REPORT

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Langerhans cell histiocytosis isolated to the thyroid gland

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Abstract Isolated Langerhans cell histiocytosis of the thyroid is an extremely rare occurrence, with only one case previously reported. A case of Langerhans cell histiocytosis isolated to the thyroid gland associated with lymphocytic thyroiditis is presented and clinical implications are discussed.

Key words Langerhans cell histiocytosis · Thyroid gland · Histiocytosis X · Differential diagnosis

Introduction

Langerhans cell histiocytosis (LCH) is the appellation now applied to a rare spectrum of disorders that includes eosinophilic granuloma, Letterer-Siwe disease and Hand-Schuller-Christian disease [7, 18]. LCH is a disorder of histiocytes of uncertain etiology and generally affects bone, lung, lymph nodes and central nervous system. Adults are infrequently affected. Although there has been involvement of the thyroid gland reported in the literature as part of systemic disease [1–6, 8–17], only a report by Coode and Shaikh [2] has described isolated thyroid disease. The patient herein reported presented with localized thyroid disease. This case report is part of an ongoing review of thyroid Langerhans cell histiocytosis.

Case report

A 32-year-old Caucasian male presented to an endocrinologist with a 3–4 week history of a thyroid mass. His thyroxine, triiodothyronine uptake and thyroid-stimulating hormone level were all within normal limits. A thyroid scan was performed, and demonstrated a 2-cm cold nodule in the right lobe of the thyroid gland. The patient declined fine needle aspiration for cytologic evaluation, and a surgical hemi-thyroidectomy was performed as an excisional biopsy and definitive therapy.

The excised thyroid tissue was red-brown, homogeneous in consistency and contained a small focal nodular mass occupying its central portion. Histologic examination of hematoxylin and eosin stained preparations demonstrated a small focal collection of histiocytes with a small number of eosinophils (Fig. 1). The lobular, indented and folded nuclei were surrounded by a microvacuolated clear to eosinophilic cytoplasm (Fig. 2). The histiocytic infiltrate had destroyed thyroid follicles, effacing the normal glandular architecture (Fig. 3). A mild lymphocytic thyroiditis was present in the remainder of the gland. Immunohistochemical stains were then performed, with S-100 protein and lysozyme reacting positively. Since the disease focus was small, additional immunohistochemical reactions and electron microscopy could not be performed. However, all findings were consistent with LCH.

The patient has since remained free of systemic LCH for the past 4 years, and has not had any other thyroid abnormalities. Thyroid function tests have remained normal to date.

Discussion

To date, isolated reports about patients with LCH thyroid involvement have included patients with concurrent systemic disease [1–6, 8–17]. Additionally, all of the reported cases seem to have an associated lymphocytic thyroiditis in addition to hypothyroidism and diabetes insipidus. The latter may imply that there is involvement of the pituitary gland, but has often not been so stated in the reports published.

The occurrence of hypothyroidism in LCH could have a number of causes: it may be related to pituitary involvement, result from destruction of thyroid follicles by histiocytic infiltrate, and/or be due to lymphocytic thyroiditis. It is unknown which factor(s) is/are actually occurring.

Diagnosis of LCH is predicated on tissue histopathology, since usual thyroid function studies may be normal and nuclear scans show only cold nodules (Fig. 4). Although the histologic findings are typical for LCH as it occurs elsewhere in the body (as in our case), the lobulated and grooved or indented nuclei can cause some con-

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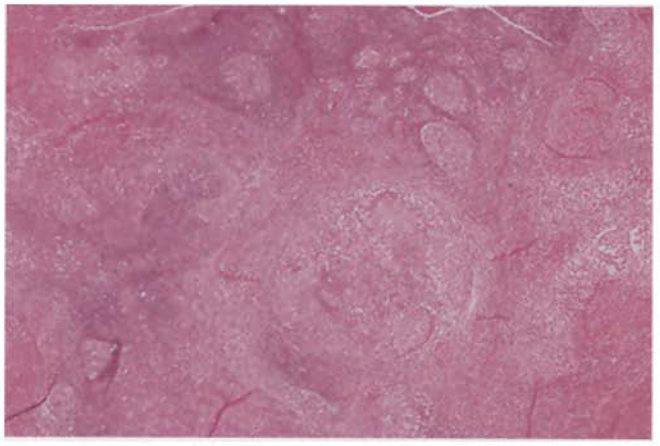

Fig. 1 Histopathology of thyroid gland demonstrating lymphocytic thyroiditis and focal Langerhans cell histiocytosis. Hematoxylin and eosin stain, × 30

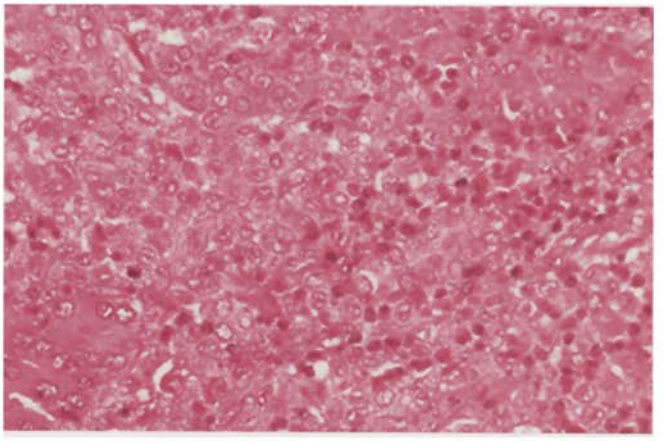

Fig. 3 Histopathology of thyroid gland specimen with typical follicular epithelial effacement, Langerhans cell histiocytes and eosinophils. Hematoxylin and eosin stain, × 400

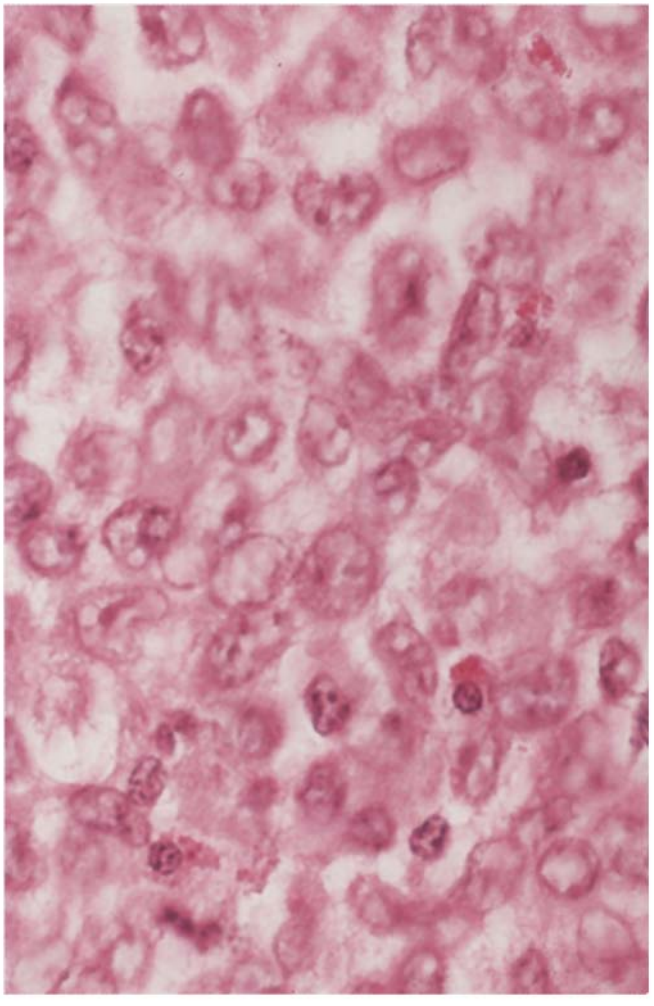

Fig. 2 Histopathology of thyroid gland specimen showing lobulated, indented and folded nuclei surrounded by pale to eosinophilic cytoplasm. A few eosinophils are seen. Hematoxylin and eosin stain, × 900

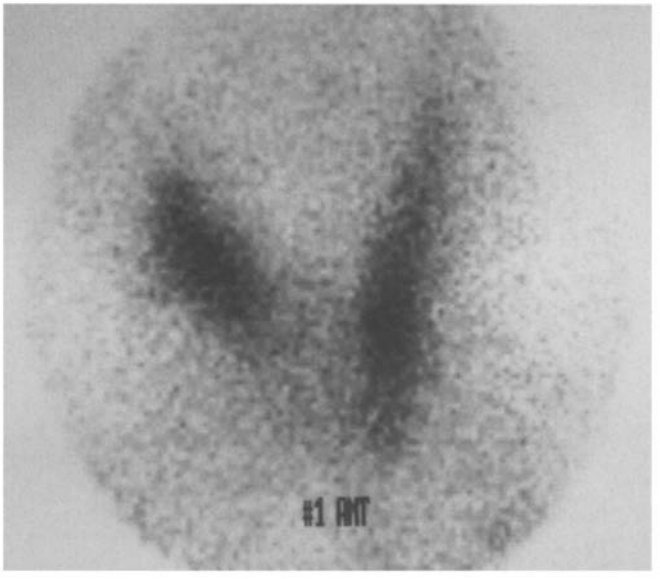

Fig. 4 Radionuclide scan with radioactive ^{131}I labelling demonstrating a cold nodule of the right lobe of the thyroid gland that was proven histologically to be Langerhans cell histiocytosis

fusion with papillary carcinoma. The typical infiltrate may also be obscured by the lymphocytic infiltration that occurs with lymphocytic thyroiditis. Careful histologic examination as well as confirmatory testing for S-100 protein and lysozyme or another macrophage-associated marker by immunohistochemical staining [such as KP-1 (CD68)] will lead to an accurate diagnosis.

The usual treatment for systemic disease associated with LCH often involves radiation or chemotherapy, while it would seem from the reported case in the literature and our case that localized disease is best managed successfully through surgery alone. It is therefore important to ascertain whether the thyroid involvement by LCH is part of systemic disease; if it is localized, curative surgical intervention may be all that a patient with localized thyroid disease requires.

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