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CORRESPONDENCE

Langerhans Histiocytosis Renders a Differential and Complementary Diagnosis for a Sjögren Syndrome Female Presenting with a Hypophyseal Mass, Pulmonary Emphysema and Thyroid Nodules

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Dear editor,

A 35-year-old female with 18 months of mouth and eye dryness, polyuria, polydipsia, amenorrhoea, several papules on the chest skin and 6 months of galactorrhea was referred to the outpatient department. Her peripheral blood sample indicated prolactin 57.84 ng/ml, follicle stimulating hormone 1.42 IU/L, luteinizing hormone 0.22 IU/L, estradiol 15.00 pg/ml; positive antinuclear antibody testing at 1:320 in a speckled pattern and positive anti-SSA. Physical examination revealed diffuse bilateral thyroid enlargement and triggering ivory lactation of bilateral breasts.

MRI demonstrated abnormal mass in the hypophysis funnel-area, about 9.0 × 6.9 × 9.5 mm with compression of optic chiasma. Water-deprivation test was positive. Ophthalmology examinations revealed positive Schirmer's test and shortened tear break-up time (right 4 s, left 4 s). The unstimulated whole salivary flow rate was 0.02 mL/min. Histologic sections of salivary gland indicated the infiltration of lymphocytes among the gland alveolus which demonstrated Sjögren syndrome (SS). Chest CT revealed diffuse pulmonary emphysema and pneumatocele in both lungs. Fine needle aspiration biopsy (FNAB) of the thyroid nodule showed numerous Langerhans-type cells within a background of eosinophils and plasma cells (Fig. 1a, b). Immunophenotypic analysis of the thyroid nodule (Fig. 1c, d) showed that the lesion cells were positive for CD1a and S-100, which further confirmed the diagnosis of LCH. The lesion was also stained positive for CD163, CD68 and Ki-67 (index 30%). The radionuclide bone imaging of the patient showed abnormal radioactive concentration at the right mandibular angle. Bone marrow biopsy revealed decreased hemopoietic tissue with normal ratio of granulocytes to erythrocytes. Bone marrow aspiration indicated active proliferation of granulocytes and erythrocytes with normal ratio and morphology. The histopathology of the hypophyseal mass was not performed on the patient because diagnosis of LCH had already been confirmed by histopathological and immunophenotypic analysis of thyroid nodules. Besides, the patient was also worried about the relatively high risk of intracranial histopathology. Systemic chemotherapy of methotrexate and cytosine arabinoside has been administrated for 3 months and the patient's symptoms have apparently alleviated.

According to 2016 SS classification criteria [1], the patient can be firmly diagnosed as SS. SS is an autoimmune disease affecting a wide range of tissues and organs. SS patients co-occurring with autoimmmune hypophysitis, a rare autoimmune disorder with the estimated prevalence of 1 in 9 million/year [2], can result in the typical MRI manifestations—pituitary enlargement and Meningioma Dural tail sign. The most common abnormal HRCT findings of SS include bronchial wall thickening (8–68%), bronchiectasis (5–46%), air trapping (32%) and nodules (6–29%) [3]. Pulmonary emphysema and pneumatocele are

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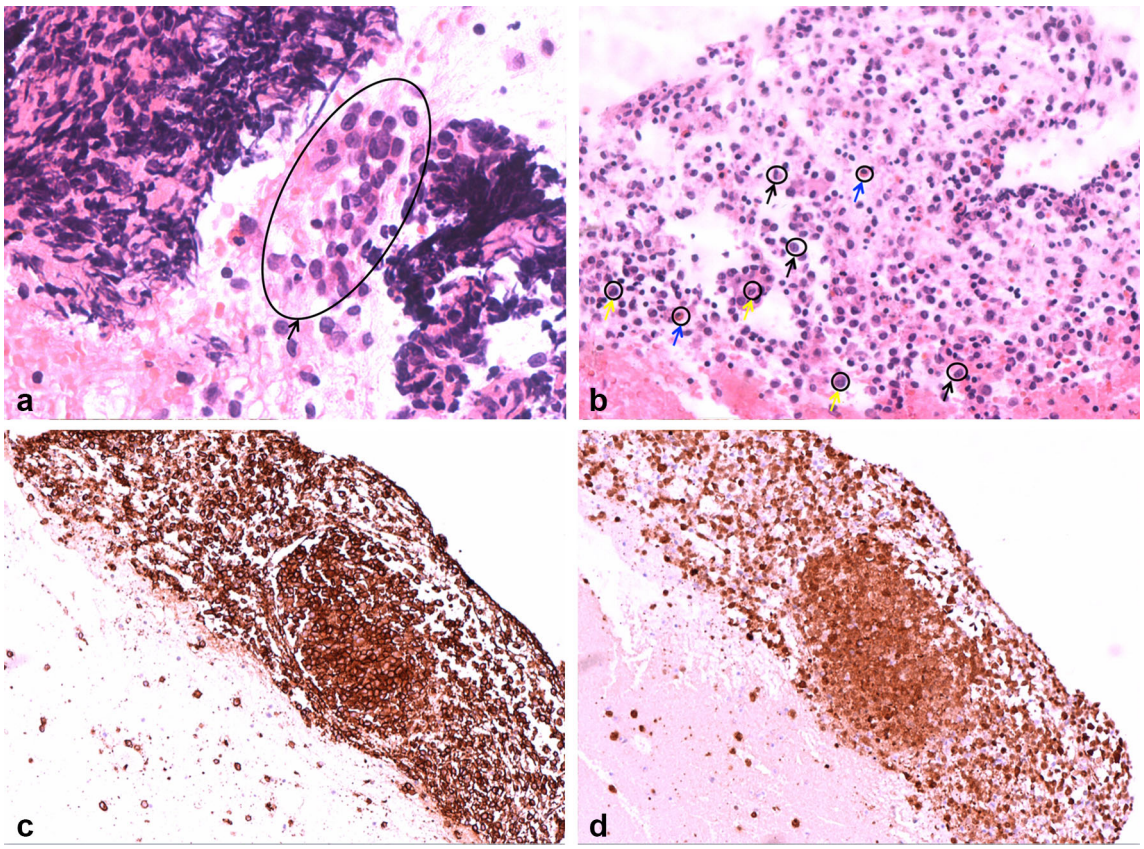

Fig. 1 Hematoxylin and eosin staining of the thyroid nodule showed numerous Langerhans-type cells (black arrow) (400 × original magnification)(Fig. 2a), within a background of eosinophils (blue arrow) and plasma cells (yellow arrow) (hematoxylin and eosin 200 ×)(Fig. 2b). Immunohistochemical stains (immunoperoxidase 100 ×) of the thyroid nodule were strongly positive for CD1a (Fig. 2c) and S-100 (Fig. 2d)

rare in SS individuals. SS complicating with autoimmune thyroid disease can lead to destruction of thyroid follicles and present higher levels of anti-thyroglobulin (ATG) and anti-thyroid peroxidase antibodies (anti-TPO). However, the patient had no typical MRI findings of AH and no detectable serum anti-TPO and/or ATG. The clinical and laboratory manifestations of the patient led us to the consideration of LCH.

With an estimated prevalence of 5 per 1 million individuals, LCH is a rare monoclonal disease caused by Langerhans-type cells originating from bone marrow [4]. It can be divided into uni-system and multi-system LCH. Although case with uni-system LCH complicated with SS has been reported before [3], we herein report the first case with concomitant of multi-system LCH and SS. Moreover, LCH cases with thyroid involvement are extremely rare [5].

In conclusion, for patients with SS involving pituitary gland, thyroid gland and both lungs, multi-system LCH should be considered as a differential diagnosis. Histopathological and immunophenotypic analysis should be implemented as early as possible to obtain the correct diagnosis and to avoid erroneous therapy. The pathogenesis connection between SS and multi-system LCH necessitates further investigation.

Compliance with Ethical Standards

Conflict of interest Authors declare that they have no conflict of interest.

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