Case Report

18F-fluorodeoxyglucose positron emission tomography/computed tomography for primary thyroid langerhans histiocytosis: A case report and literature review

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ABSTRACT

Langerhans cell histiocytosis (LCH) is a rare clonal proliferative disease, with an incidence rate of 4.0–5.4/1 million individuals. LCH encompasses a spectrum of disorders with diverse clinical presentations ranging from a single organ to multiple organ involvement. LCH rarely involves the thyroid gland. We presented a case with LCH of thyroid gland. The patient had painless progressive neck enlargement and then diabetes insipidus. Ultrasonic scan and magnetic resonance imaging scan revealed nodular goiter and pituitary stalk enlargement, respectively. Histopathological analysis revealed features of histiocytoid cells. 18F-fluorodeoxyglucose (18F-FDG) positron emission tomography/computed tomography (PET/CT) was performed in order to rule out the presence of whole body infiltration. 18F-FDG PET/CT also demonstrated increased uptake in the thickening pituitary stalk and maxillofacial skin lesion, in addition to the bilateral thyroid nodules, CT showed the left lung nodule and the skull destruction without 18F-FDG uptake. This report emphasizes the role of 18F-FDG PET/CT in multiple organs involvement of patients with LCH.

Keywords: 18F-fluorodeoxyglucose, langerhans histiocytosis, positron emission tomography/computed tomography, thyroid

INTRODUCTION

Langerhans cell histiocytosis (LCH) is a rare neoplastic disease of antigen-presenting cells, with an incidence rate of 4.0–5.4/1 million individuals.[1] The most frequent sites of occurrence are skin, bone, central nervous system (CNS).[2] LCH rarely involves the thyroid gland, and management of such disease is controversial.[3] Computed tomography (CT), magnetic resonance imaging (MRI), ultrasonic scan (USS) are mainly for the individual part, while multiple systems involvement need the whole body imaging. Therefore, a whole body scan is useful. We present an unusual case of LCH of the primary thyroid gland, which was confirmed by fine needle aspiration cytology. In addition to the thyroid gland, 18F-fluorodeoxyglucose (18F-FDG) positron emission tomography (PET)/CT discovered multiple organs involvement such as pituitary stalk, maxillofacial skin lesion, lungs, and skull.

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CASE REPORT

A 35-year-old man complained with painless progressive asymmetric enlargement of the bilateral thyroid lobes for a year. In the recent 3-month, he was suffered from diuresis and polydipsia. No hereditary syndromes, family history of cancer, and personal history were disclosed. There was no additional past medical or surgical history. Blood tests revealed a normal full blood count, electrolytes, urea, creatinine, calcium, and phosphate levels. Thyroid function tests reported a thyroid stimulating hormone level of 0.97 (normal range 0.3–4.6 uIU/mL), free T4 of 21.78 (normal range 6.6–24.8 pmol/L), and free T3 of 5.80 (normal range 3.8–8.5 pmol/L), thyroglobulin antibodies of 53.20 (normal range <30 U/mL), thyroid peroxidase antibody of 64.00 (normal range 0.16–10 IU/mL). In addition, serum calcitonin was recorded at <1.0 (normal range <11.8 ng/L) and

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urine catecholamine levels were within normal range. Water pressure test revealed explicitly for central diabetes insipidus, and desmopressin acetate was given for 1-month. USS of the neck, conducted at the referring hospital, reported an enlarged (3.1 cm and 2.8 cm in diameter) bilateral thyroid lobes [Figure 1]. Subsequently, a needle biopsy from the thyroid mass revealed features of langerhans cell with the tumor cells staining positive for CD1a, S-100, negative for cytokeratin [Figure 2]. MRI sections of the head revealing pituitary stalk enlargement [Figure 3]. 18FDG PET/CT was performed for further evaluation of LCH in the whole body [Figure 4]. The maximum projection image of the PET revealed increased uptake in thyroid gland (standardized uptake value [SUVmax] = 9.4) [Figure 4a], pituitary stalk (SUVmax = 4.9) [Figure 4b], and maxillofacial skin lesion (SUVmax = 4.9) [Figure 4c], which corresponded well to bilateral thyroid nodule, the thicking of pituitary stalk, and skin lesion on the CT. However, CT showed the left lung nodule [Figure 4d] and the skull destruction [Figure 4e] without 18FDG uptake.

DISCUSSION

Langerhans cell histiocytosis is a type of histiocytic disorders, which was first reported in 1893. The age of onset of LCH from infant to adult and it frequently occur in childhood. Final diagnosis of LCH is made by histopathology and immunohistochemistry,

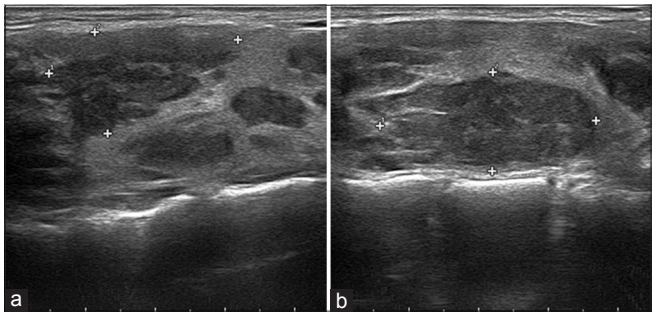

Figure 1: (a) An Ultrasonic scan (USS) of the neck, conducted at the referring hospital, reported multiple keyed acoustic area, basic clear boundary in both thyroid lobes [b: 3.1, and a: 2.8 cm in diameter]. (b) An USS of the neck, conducted at the referring hospital, reported multiple keyed acoustic area, basic clear boundary in both thyroid lobes [b: 3.1 and a: 2.8 cm in diameter]

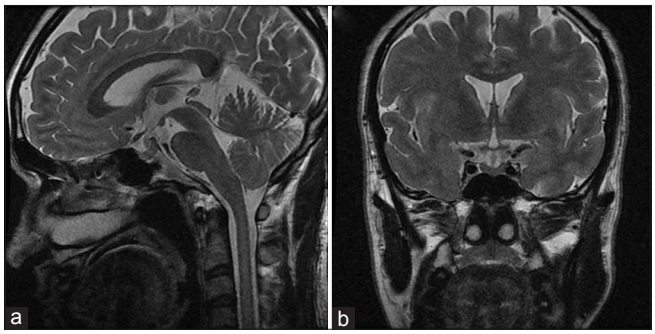

Figure 3: (a) Magnetic resonance imaging (MRI) sections of the head revealing pituitary stalk enlargement. (b) MRI sections of head revealing pituitary stalk enlargement

and there are no specificities only depending on the clinical features or radiologic findings of LCH. The radiologic imaging may provide some characteristics, such as "punched-out" lesion

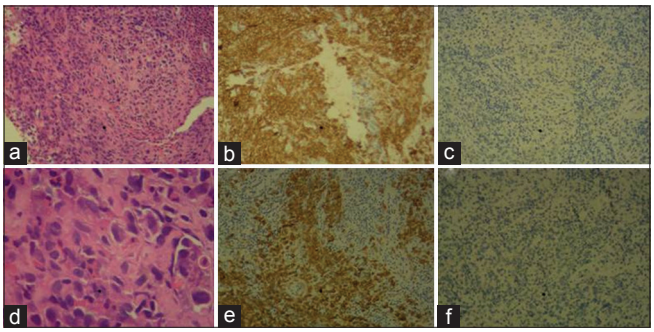

Figure 2: (a) H and E staining [a: H and E ×100; b: H and E ×400] and immunostaining (CD1a, Langerin and S-100) of lesions from the thyroid gland lesions. The lesions from the thyroid gland exhibited positive expression patterns for CD1a [c: ×100], Langerin [d: ×100] and S-100 [e: ×100], and negative for cytokeratin (CK) [f: ×100]. (b) H and E staining [a: H and E ×100; b: H and E ×400] and immunostaining (CD1a, Langerin and S-100) of lesions from the thyroid gland lesions. The lesions from the thyroid gland exhibited positive expression patterns for CD1a [c: ×100], Langerin [d: ×100] and S-100 [e: ×100], and negative for CK [f: ×100]. (c) H and E staining [a: H and E ×100; b: H and E ×400] and immunostaining (CD1a, Langerin and S-100) of lesions from the thyroid gland lesions. The lesions from the thyroid gland exhibited positive expression patterns for CD1a [c: ×100], Langerin [d: ×100] and S-100 [e: ×100], and negative for CK [f: ×100]. (d) H AND E staining [a: H and E ×100; b: H and E ×400] and immunostaining (CD1a, Langerin and S-100) of lesions from the thyroid gland lesions. The lesions from the thyroid gland exhibited positive expression patterns for CD1a [c: ×100], Langerin [d: ×100] and S-100 [e: ×100], and negative for CK [f: ×100]. (e) H and E staining [a: H and E, ×100; b: H and E, ×400] and immunostaining (CD1a, Langerin and S-100) of lesions from the thyroid gland lesions. The lesions from the thyroid gland exhibited positive expression patterns for CD1a [c: ×100], Langerin [d: ×100] and S-100 [e: ×100], and negative for CK [f: ×100]. (f): H and E staining [a: H and E × 100; b: H and E × 400] and immunostaining (CD1a, Langerin and S-100) of lesions from the thyroid gland lesions. The lesions from the thyroid gland exhibited positive expression patterns for CD1a [c: ×100], Langerin [d: ×100] and S-100 [e: ×100], and negative for CK [f: ×100]

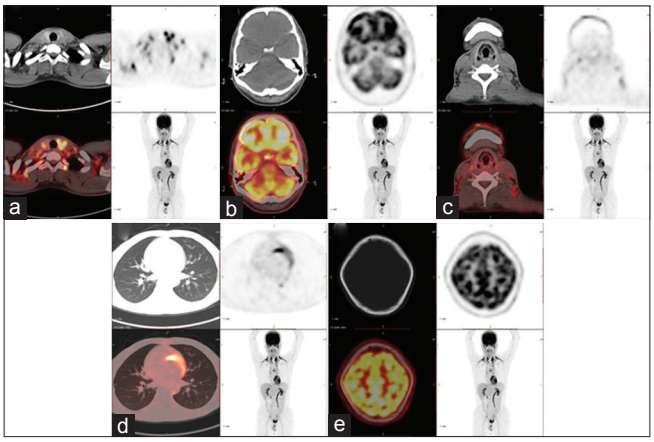

Figure 4: (a) 18F-fluorodeoxyglucose positron emission tomography/computed tomography (18FDG PET/CT) revealed increased uptake in the thyroid gland and maxillofacial skin, in addition to the left lung and skull. (b) 18FDG PET/CT revealed increased uptake in the thyroid gland and maxillofacial skin, in addition to the left lung and skull. (c) 18FDG PET/CT revealed increased uptake in the thyroid gland and maxillofacial skin, in addition to the left lung and skull. (d) 18FDG PET/CT revealed increased uptake in the thyroid gland and maxillofacial skin, in addition to the left lung and skull. (e) 18FDG PET/CT revealed increased uptake in the thyroid gland and maxillofacial skin, in addition to the left lung and skull

or beveled edge in skull, symmetric flattening of vertebral body, lytic lesion with sclerotic rim and surrounding areas of sclerosis more common in iliac bone, diaphyseal or metaphyseal of long bones involved in children.[4] Otherwise, approximately 70% of patients will also show thickening of the pituitary stalk on MR images.[5]

The mechanism of FDG uptake by malignant tissue or metastatic lesions is due to the enhanced rate of glucose utilization by neoplastic cells. Furthermore, it was found that granulation tissue and macrophages may show FDG uptake.[6,7] LCH as a type of histiocytic disorders, most of the lesions display a high level of FDG uptake.[8,9] The lesions of LCH can range from a single system involvement to a multisystem disease. It may present as a neoplasm, skin rash, lytic bone lesion, emphysematous bullae, interstitial lung disease or present with multiple affected organ systems within the human body. The most common clinical CNS manifestation of LCH is diabetes. In this case, the patient underwent a whole-body PET/CT, which was positive for suspicious lesions, PET revealed increased FDG uptake not only in thyroid nodule, but also in the pituitary stalk and maxillofacial skin. In the region of these areas, the intense FDG activity on PET corresponded well to CT and the fusion images. We also found that the left lung nodule has slighter metabolic (SUVmax 0.5) and the skull destruction even nonmetabolic in PET. It has been reported that PET may showed false-negative in some small bone lesions, such as skull and cerebrum involvement. Hence, it was suggested that combined multiple analysis methods would improve sensitivity and specificity of the whole body LCH.[8]

Positron emission tomography/computed tomography is a useful tool to rule out the presence of other organ infiltration. As the intent by our case, it also provides a reference basis of classification, staging, treatment plan, and evaluation therapy effect. FDG PET/CT not only provides the characteristics of lesions in CT scan, but also finding the lesions activity FDG uptake. Previous reports of LCH in FDG PET/CT imaging mostly shows bone, lung, lymph node or skin invasion. Few cases of the primary thyroid LCH showed in FDG PET/CT have been reported.[10-12] In the patient of this case, the thyroid involved LCH was the first symptoms, and clinical considered as thyroid cancer. However, the result of biopsy pathology was thyroid LCH. Whole-body PET/CT was performed to evaluate the involvement of disease and make an individualized treatment plan after the biopsy pathology. FDG PET/CT showed bilateral involvement of multiple low-density nodules, the boundary of nodules are clear and some breakthrough thyroid capsule. FDG uptake was significantly increased in the nodules; which were mostly higher than primary thyroid cancer in addition to low-differentiated cancer and medullary carcinoma. In CT, these nodules had rare calcification and the density mostly lower than primary thyroid cancer. However, to confirm all these characteristic performances, needs more patients' data accumulation.

In summary, we recommend the use of PET/CT as a meaningful noninvasive tool to evaluation scope of LCH lesions involving. It also useful in treatment decision establish and curative effect evaluation.

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