Isolated Langerhans Cell Histiocytosis of the Thyroid gland: Diagnosed on FNAC

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ABSTRACT

Langerhans cell histiocytosis (LCH) is a rare illness with an incidence of 0.5–5.4 per million individuals. Isolated thyroid gland involvement is an unusual presentation of LCH that can mimic malignancy and pose a diagnostic challenge for clinicians. We report a case of unifocal LCH involving thyroid gland in a 12 year boy. Fine Needle Aspiration Cytology (FNAC) smears were highly cellular comprising of numerous eosinophils, lymphocytes, macrophages, neutrophils along with large Langerhans cells (LCs) having abundant granular cytoplasm, irregular nuclei and longitudinal nuclear grooves. On immunocytochemistry these cells were S-100 positive. Our case discusses the characteristic cytological features and differential diagnoses of unifocal LCH involving the thyroid gland, emphasizing the diagnostic utility of FNAC in such an unusual clinical presentation, thus obviating the need of biopsy.

Keywords: Langerhans cell histiocytosis, Langerhans cell,Thyroid,FNAC

INTRODUCTION

Langerhans cell histiocytosis (LCH) is a rare monoclonal proliferation of abnormal dendritic cells (Langerhans cells) originating from bone marrow.[1] The solitary form of the disease usually affects the bone and is indolent, while the multisystem form of the disease is aggressive and involves many organs including bone, skin, liver, spleen, and lymph node. [2] Amongst the endocrine organs, pituitary gland is most commonly involved and presents as diabetes insipidus.[1]Thyroid gland involvement is very rare in patients with multisystemic disease. Isolated involvement of thyroid gland is even rarer with around 75 cases being reported in literature till now.[3] Though the cytological features of LCH have been well documented in literature, very limited number of cases with thyroid involvement have been diagnosed on FNAC. We report an unusual case of LCH with isolated thyroid gland involvement in a 12 year old boy which was clinically mimicking multinodular goiter and was diagnosed on FNAC.

CASE REPORT

An 18 year old female presented with abdominal A 12 year boy presented with an anterior neck swelling for the last 2 years. The swelling was diffuse, painless and gradual in onset. On examination, the swelling measured 5x5 cm and appeared to involve both the thyroid lobes (Figure. 1). It was soft, nontender and moved with deglutition. The child had no other significant complains except mild pressure symptoms like difficulty in deglutition. No cervical lymphadenopathy or organomegaly was noted. Provisional diagnosis of multinodular goitre was made and the child was referred to the FNA Clinic. Thyroid profile showed normal thyroid hormone levels. Serum antithyroid peroxidase and thyroglobulin antibodies were negative. Complete hemogram and other biochemical parameters were within normal limits. Ultrasound and radionuclide scan were suggestive of multinodular goitre with multiple large cold nodules in both lobes of thyroid.

FNAC was done from both thyroid lobes using 23 G needle which yielded blood mixed aspirates. Ethanol fixed smears and air-dried smears were prepared and stained

with Papanicolaou and Giemsa methods, respectively. Aspirate smears were highly cellular and comprised of a polymorphous population of numerous eosinophils, lymphocytes, foamy histiocytes, and neutrophils admixed with thyroid follicular cells (Figure 2). Admixed were many singly scattered large round to oval cells, having moderate to abundant granular cytoplasm, central oval nucleus with vesicular chromatin and inconspicuous nucleoli. Prominent nuclear indentations, infoldings and longitudinal grooves giving a coffee bean appearance were observed within these histiocytic cells (Figure 3). No mitosis was identified. Few binucleate and multinucleated histiocytic giant cells were also seen. Papanicolaou smear was destained and immunocytochemistry was performed. These atypical histiocytic cells showed strongly positivity for S-100 (Figure 3 inset). Cytological features were suggestive of LCH. Skeletal survey and chest Xray was requested, which did not show any involvement of bones. No other systemic involvement was found.

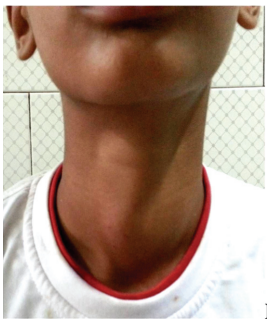

Figure 1

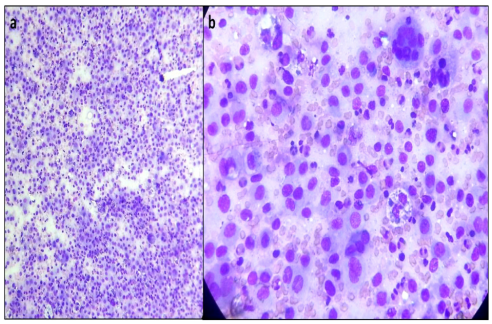

Figure 2

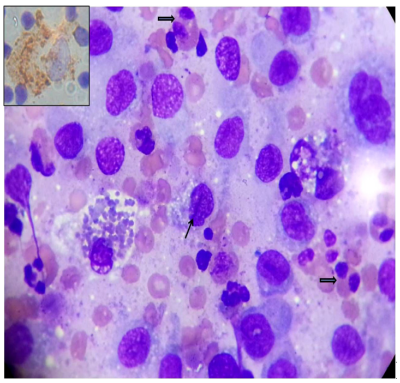

Figure 3

Figure 1: Clinical photograph shows diffuse anterior neck swelling measuring 5x5 cm and involving both the thyroid lobes.

Figure 2: May Grunwald Giemsa stained aspirate smears are cellular and show a polymorphous population of numerous eosinophils, lymphocytes, plasma cells, tingible body macrophages, histiocytes, neutrophils admixed with thyroid follicular cells. Few binucleated and multinucleated histiocytic giant cells are also seen. a) 40x and b) 200x

Figure 3: Higher magnification shows large round to oval Langerhans cells having moderate to abundant cytoplasm, central oval nucleus with vesicular chromatin, inconspicuous nucleoli and prominent nuclear infoldings and longitudinal grooves (thin arrow), giving a coffee bean appearance. Few eosinophils (thick arrows) and a multinucleated histiocytic giant cell are also seen in the background. (May Grunwald Giemsa x400). Inset- Langerhans cells show cytoplasmic positivity for S-100 (x1000).

DISCUSSION

LCH is a rare disease caused by clonal proliferation of antigen-presenting dendritic cells referred to as Langerhans cells (LC).[3]

The vast majority of LCH patients are children under the age of 10 years, but thyroid LCH is much more common in adults and has a relatively indolent course.[4] Isolated involvement of thyroid by LCH is believed to be even rarer, although in a case series by Thompson et al, isolated thyroid involvement was slightly more common than being part of multifocal disease.[4] Thyroid LCH is seen in only 1 out of 238 LCH cases. [5] It is important to distinguish isolated thyroid LCH from multisystemic disease because of the favourable prognosis and an excellent survival of around 100% in the former.[6] The usual first symptom of thyroid LCH is diffuse or nodular thyroid enlargement.[7] However, there are instances in which the microscopic involvement by LCH had been found incidentally only on histological examination, and in these cases, thyroid enlargement might be due to coexisting pathology such as adenomatous goiter, lymphocytic thyroiditis, or papillary carcinoma. [4,7] Not only the clinical presentation but also the thyroid hormone status, antithyroid antibodies, sonographic and scintigraphic findings of LCH may be similar to other thyroid disorders including malignancy. [8,9]

The diagnosis of LCH in our patient was made on the basis of characteristic Langerhans cells (LCs) admixed with polymorphous population of numerous eosinophils, neutrophils, lymphocytes, plasma cells and multinucleated giant cells on aspirate smears. The key to diagnosis is identification of LCs by their characteristic longitudinal nuclear grooves, indentations and nuclear pseudoinclusions. Number of eosinophils in cytology smears varies in different areas of a LCH lesion as well as in different organs. Their presence can help in drawing attention towards the diagnosis. Differential diagnoses considered in our case were autoimmune thyroiditis, papillary carcinoma of thyroid and malignant melanoma. Intense lymphoid inflammatory infiltrate in autoimmune thyroiditis can overshadow the histiocytes and eosinophils of LCH, leading to a misdiagnosis. Lymphocytic thyroiditis has been seen in association with LCH.[4] Presence of abundant granular cytoplasm with low nuclear-cytoplasmic ratio, vesicular bland chromatin and prominent nuclear foldings and grooves in the LCs, cytoplasmic immunopositivity for S-100 and the concomitant presence of eosinophils helped in excluding the diagnosis of papillary thyroid carcinoma. Rarely, papillary

thyroid carcinoma can coexist with LCH. [9,10] Lack of prominent nucleoli, nuclear pleomorphism, intracytoplasmic pigment and mitoses helped in excluding the possibility of melanoma.

Confirmation is done by immunopositivity for CD1a, langerin and/or S-100 and by demonstration of Birbeck's granules on electron microscopy. Electron microscopy is costly and not an easily available modality, therefore, not considered mandatory for diagnosis.[11] For localized thyroid LCH, resection by subtotal or total thyroidectomy along with long term follow up by ultrasound and computed tomography scan is the treatment of choice.[6] The patients with apparently unifocal LCH need thorough clinical work up and staging of their disease to ensure that these are not part of a multisystemic involvement. This is important because unlike unifocal disease, multifocal LCH is associated with disease progression, recurrence and an aggressive course, therefore, warrants chemotherapy. [6]

Diagnosis of LCH on FNAC may be missed due to lack of familiarity with the cytological features. The presence of Langerhans cells having prominent nuclear indentations and grooves along with numerous eosinophils should raise the suspicion of this rare entity on FNAC smears.

To conclude, isolated LCH of the thyroid gland is a very rare entity. Owing to non specific clinical and radiological findings, it poses a diagnostic dilemma for clinicians. The present case highlights the diagnostic role of FNAC in such a clinical scenario. A high index of suspicion along with awareness of the cytological features and differential diagnoses of LCH can help in making an accurate diagnosis on FNAC. This can obviate the need of biopsy and electron microscopy.

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