CASE REPORT

Incidental finding of Langerhans Cell Histiocytosis in Thyroid Gland

An Unusual Tumour in the Thyroid Gland

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ABSTRACT

Langerhans cell histiocytosis(LCH) is an uncommon neoplasm arising from the histiocytic cells of the dendritic network[1]. It is commonly seen affecting paediatric population with a systemic involvement (multi-organ involvement), whereas it involves infrequently a single system in the adult population[1, 2]. The organ systems most frequently affected by LCH in adults are pulmonary system, skeletal system, and cutaneous system. Involvement of endocrine system by LCH in adults is not so common. However, the most common manifestation of endocrine involvement is diabetes insipidus due to the involvement of the posterior pituitary gland[1, 2]. LCH involvement of the endocrine gland, thyroid is extremely rare[3, 4]. It is due to the uncommon age group and uncommon organ system of involvement by LCH, we report our case of incidental finding of Langerhans cell Histiocytosis in the thyroid gland in a 29 year old South Indian female.

Case Report

A 29 year old South Indian woman presented with goitre for duration of 6 months with pressure symptoms. She was a known patient with hypothyroidism and was on 50 mcg of Thyroxine. Physical examination revealed Grade 2 Multinodular Goitre which was firm in consistency. Her TFT was normal on 50 mcg Thyroxine and her Ultrasound of Neck showed multiple, hypoechoic areas in both lobes. Antimicrosomal antibody levels and antithyroglobulin antibody levels were elevated. FNAC Thyroid was suggestive of lymphocytic thyroiditis.

A total thyroidectomy was done in view of the pressure symptoms.

Gross pathology of the whole specimen revealed left lobe along with isthmus measuring 4.5 × 4 × 2 cms and right lobe measuring 5.5 x 4.5 × 2.5 cms. The entire specimen weighed about 180 grams. Cut section of both the lobes revealed diffuse firm grey white areas with interspersed grey brown areas (*Fig. 1).*

Fig. 1 : Gross photograph of the right(A) and left lobes (B) of thyroid

The histopathological examination of thyroid parenchyma revealed thyroid follicles with hurthelized cell changes and stroma with prominent lymphoid follicles and fibrosis. Since the morphologic appearance was in favour of Hashimoto thyroiditis, extensive sampling was done to rule out papillary carcinoma thyroid. On further sections, diffuse sheets of discohesive mononuclear cells, with oval nucleus exhibiting grooves and clefts and moderate orangophilic cytoplasm was seen along with abundant mature eosinophils and scatterd small lymphocytes. Focal areas showed the mononuclear cells infiltrating the extra-thyroidal muscular and fibrotic tissue. There was no necrosis and increased mitotic activity *(Fig. 2).*

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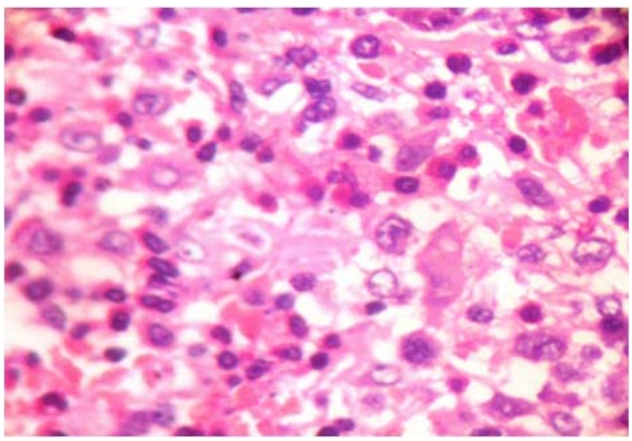

Fig. 2 : Biopsy revealed diffuse sheets of discohesive mononuclear cells, with oval nucleus exhibiting grooves and clefts in a background of eosinophils (Haematoxylin and Eosin X40).

Immunohistochemically, the tumour cells stained strongly with CD1A (diffuse cytoplasmic staining) and S100 P (nuclear staining) and stained negative for CD45, which provided support to the dendritic cell origin of the neoplasm *(Fig.3A, B)*. The thyroid follicles were positive for pan cytokeratin and the lymphoid follicles were positive for CD20. Also the scattered lymphocytes were positive CD3.

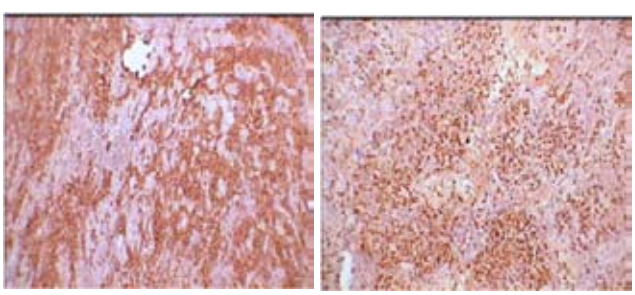

Fig.3 : Immunohistochemical characterization showed (A) CD1A diffuse cytoplasmic staining and (B) S100P nuclear staining.

Hence, with the aid of the morphology and immunohistochemical findings, a diagnosis of LCH with lymphocytic thyroiditis was made. This diagnosis was an incidental finding in a thyroidectomy specimen done for clinically symptomatic goitre.

Following surgery and diagnosis, a whole body scan was done and no evidence of disease involvement in any organs was seen. All the other haematological and biochemical tests were done and all were found to be within normal ranges. Hence a diagnosis of incidental or latent LCH was made. After 2 years of follow up, no definite evidence of systemic involvement of the disease was noticed.

Discussion

Though LCH predominantly affects the paediatric population, thyroid involvement by LCH is mostly seen in the adult population[3]. Almost all the reported thyroid involvement by LCH co-exists with either Lymphocytic thyroiditis or Hashimoto thyroiditis. Moreover, LCH in thyroid may exhibit either diffuse or focal involvement[3,5]. Our case also showed multiple focal LCH in the background of lymphocytic thyroiditis. Hence a strong suspicion and meticulous sampling is warranted in all lymphocytic thyroiditis, to rule out the rare possibility of co-existing LCH. However, a diffuse inflammatory infiltrate may mask the pathgnomonic morphologic features of malignant dendritic cells. Presence of eosinophils along with foci of dendritic cells(coffee-bean like) with clefts and grooves in a background of lymphocytic thyroiditis should always be looked with a high degree of suspicion for the presence of LCH. Also, immunohistochemical characterization of LCH by positive CD1A, S100P is a confirmatory for the diagnosis of LCH[3].

According to the literature, localized thyroid involvement in LCH, has favourable outcome[3]. In line with the literature, our subject had no recurrence or other organ involvement in follow-up.

References

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