Langerhans Cell Histiocytosis of Thyroid—A Diagnostic Dilemma

R. A. Pandyaraj · K. Sathik Mohamed Masoodu ·
S. Maniselvi · S. Savitha · H. Divya Devi

Received: 16 May 2014 / Accepted: 26 May 2014 / Published online: 17 June 2014
© Association of Surgeons of India 2014

Abstract Langerhans cell histiocytosis (LCH) is a rare disease of antigen presenting cells and involvement of thyroid is really uncommon. The thyroid if involved is usually seen in multisystem disease but isolated thyroid involvement is very rare. The diagnosis of Langerhans cell histiocytosis of the thyroid is very challenging due to its rarity and is usually misdiagnosed as benign goiters, undifferentiated carcinoma, lymphoma, etc. Management of Langerhans cell histiocytosis of the thyroid also remains controversial. Prognosis in an isolated Langerhans cell histiocytosis of the thyroid is usually good but as it may precede a multisystem involvement, prolonged follow-up is required. We present a rare case of Langerhans cell histiocytosis of the thyroid, with variable diagnoses on fine needle aspiration cytology.

Keywords Thyromegaly · Langerhans cell histiocytosis · Thyroid tumor · Lymphoma thyroid

R. A. Pandyaraj
Kilpauk Medical College, No.11, Begum Sahib 3rd Street,
Mount Road, Chennai 600 002, India

K. Sathik Mohamed Masoodu (✉)
Kilpauk Medical College, 32/615 F2, 24th Cross Street,
Shanthi Nagar, Palayamkottai, Tirunelveli 627 002, India
e-mail: sathik.world@gmail.com

S. Maniselvi
Kilpauk Medical College, 22/1, New no. 14, 6th Main Street,
Saurashtra Nagar, Choolaimedu, Chennai 600 094, India

S. Savitha
Kilpauk Medical College, No.48, Gowdia Mutt road,
Royapettah, Chennai 600 014, India

H. Divya Devi
Kilpauk Medical College, GF.3, Vasanth Apartments, No.1,
Srikrishnapuram Street, Royapettah, Chennai 600 014, India

Case Study

A 45-year-old male presented with thyroid swelling for 1 year duration, which was indolent in its course. He had no other complaints. He had no symptoms of hypo or hyperthyroidism. His other medical problems were diabetes mellitus and hypertension. On general examination, the patient was obese, PR-78/min, Bp-160/100 mmHg. Examination of the neck revealed diffusely enlarged, firm thyroid of size 15×8 cm with no nodules. Cervical lymphnodes were not palpable.

On investigating, complete hemogram and renal parameters were normal except for elevated blood sugar values for which the patient was started on insulin therapy. Thyroid profile showed normal levels of T3 and T4 with mild elevation of TSH. Antithyroid antibodies were negative. USG of the neck showed goiter with altered echoes. FNAC reports were indecisive, varying between colloid goiter and anaplastic carcinoma. CT of the neck showed diffusely enlarged thyroid with heterogenous attenuation, likely malignant. USG of the abdomen was normal. Both vocal cords were mobile on video-laryngoscopy.

The case was discussed in the tumor board and planned for total thyroidectomy for definitive diagnosis. Intraoperatively, the thyroid gland was found to be diffusely enlarged and densely adherent to the strap muscles and trachea. Total thyroidectomy was completed with difficulty after preserving recurrent laryngeal nerves on both sides. The specimen was sent for histopathology (Fig. 1). During immediate postoperative period, patient developed stridor, probably due to tracheomalacia, so he was reintubated. The patient was gradually weaned from ventilatory support and extubated on the 6th postoperative day.

Initially, the histopathology of the specimen was reported as non-Hodgkin's lymphoma. With this diagnosis, he was further evaluated for involvement of other sites. CT of the chest, CT of the abdomen, and bone marrow aspiration were

Fig. 1 Total thyroidectomy specimen

normal. The specimen was sent for immunohistochemical analysis which surprisingly showed only focal positivity for CD45, CD20, and CD 3 but a strong positivity for S100 and CD1a. The specimen was re-evaluated which showed oval, plump histiocytes with eosinophil, and lymphocyte aggregates (Fig. 2) and, hence, a final diagnosis of Langerhans cell histiocytosis (LCH) was obtained. With a diagnosis of LCH of the thyroid, he was investigated further to rule out multisystem involvement. Bone scintigraphy did not reveal any bone involvement. Thus, a final diagnosis of solitary LCH of the thyroid was made for which he is now on regular follow-up.

Discussion

LCH is a rare neoplastic disease of the langerhans cell, an antigen presenting cell with an incidence rate of 4.0–5.4 per 1 million individuals [1]. LCH is most commonly a pediatric disease, but it can affect the adult population. It usually presents as a multisystem granulomatous infiltrate (in two thirds of cases) in adults. Bone involvement predominates (80 % of cases) with osteolytic and asymmetric lesions, most often of the skull. Other systems involved include the skin, pituitary gland, liver, spleen, and orbits [2].

Thyroid involvement in LCH is rare with only around 65 cases being reported [1]. The thyroid gland is involved mostly as part of a multisystem disease with solitary involvement being extremely rare. Diagnosis of LCH is found to be extremely challenging [2]. It is usually misinterpreted as a benign disorder or poorly differentiated carcinoma/anaplastic carcinoma or lymphoma [1–3]. Similar difficulties have been reported by Wang W-S et al., and Patten DK et al. Such difficulty has been attributed to its rarity [2]. The disease is confirmed by electron microscopy or immunohistochemical reactivity of histiocytes to CD1a and/or S100. Electron microscopy shows characteristic birbeck granules [1, 2]. Even in our case, preoperatively cytological examination was reported as anaplastic carcinoma. Postoperatively, it was reported initially as non-Hodgkin's lymphoma, but after

Fig. 2 Histopathology showing histiocytes with nuclear grooving (a)

immunohistochemical analysis it was confirmed as langerhans cell histiocytosis.

LCH cases involving the thyroid gland present with diffuse (59 %) or nodular thyroid (25.8 %) enlargement. Thyroidal pain and obstructive symptoms secondary to a primary LCH goiter are rare. Our patient had no specific symptoms other than the neck swelling and was found to be subclinically hypothyroid.

Management of LCH of the thyroid is controversial. With diagnosis of thyroid LCH, further investigations need to be done to rule out other system involvement which includes CT chest, bone scintigraphy, and abdominal USG [1]. Solitary involvement of the thyroid has an indolent course with a good prognosis [4]. Treatment of choice for primary thyroid LCH is surgical resection by hemi-, subtotal, or total thyroidectomy. Adjuvant therapies of chemo-radiation have also been tried. As per Patten DK et al., majority of adults had surgery alone (57.5 %) followed by surgery and chemotherapy or radiotherapy (21.3 %) [1].

Conclusion

Langerhans cell histiocytosis is a rare disease with thyroid involvement being extremely rare. Diagnosis of thyroid LCH is quite challenging; hence, LCH being a differential diagnosis in thyroid swelling needs to be considered. It is currently considered as a low grade malignancy and because it can rarely precede or reveal a multisystem disease, additional investigations as well as a prolonged follow-up are justified.

References

1. Patten DK, Wani Z, Tolley N (2012) Solitary langerhans histiocytosis of the thyroid gland: a case report and literature review. *Head Neck Pathol* 6(2):279–289
2. Braithe F, Kurzrock R (2006) Langerhans cell histiocytosis of the thyroid. *J Clin Oncol* 24(3):522–523

3. Wang W-S, Liu J-H, Chiou T-J, Hsieh R-K, Yen C-C, Chen P-M (1997) Langerhans' cell histiocytosis with thyroid involvement masquerading as thyroid carcinoma. *Japan J Clin Oncol* 27(3):180–184
4. Foulet-Roge A, Josselin N, Guyetant S, Gardet J-J, Besancon A, Saint-Andre J-P et al (2002) Incidental langerhans cell histiocytosis of thyroid: case report and review of the literature. *Endocr Pathol* 13(3):227–233