

Case Report

Amyloid goiter with diffuse thyrolipomatosis: a rare entity

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Received: 08 March 2024

Revised: 09 April 2024

Accepted: 11 April 2024

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ABSTRACT

Diffuse thyroid lipomatosis with amyloid deposition is a rare entity. We report a case of a 42-year-old lady who presented with progressive enlargement of the thyroid gland with compressive symptoms. Her thyroid hormone levels were within normal limits. Imaging revealed enlarged thyroid gland with multiple fat density lesions. She underwent total thyroidectomy, and histopathology revealed diffuse infiltration of thyroid stroma by mature adipose tissue and an associated amyloid deposition. A diagnosis of amyloid goiter with diffuse lipomatosis of the thyroid gland was rendered. Only a few cases of diffuse lipomatosis with concurrent deposition of amyloid protein have been reported in the literature.

Keywords: Thyroid lipomatosis, Amyloid, Goiter, Thyrolipoma

INTRODUCTION

Diffuse thyroid lipomatosis is a rare histopathological condition characterized by diffuse adipocytic infiltration of the thyroid gland.¹ It can present with neck swelling, difficulty in swallowing, and hoarseness. Patients may be hypothyroid, euthyroid/hyperthyroid. Thyrolipomatosis may also be associated with amyloid deposition. The mechanism of fatty tissue development in thyroid lesions and its association with amyloidosis have not been clearly understood. Less than 40 cases have been documented in literature till date.

CASE REPORT

A 42-year-old lady with long standing type 2 diabetes mellitus presented with a neck mass gradually enlarging since past 6 years, with compressive symptoms recently. Physical examination showed a lobulated and asymmetrical diffuse thyroid enlargement. Her routine lab investigations were within normal limits including thyroid

function tests. USG neck was done which showed features of multinodular goiter. CT scan revealed a diffusely enlarged thyroid gland with multiple fat density lesions causing lateral displacement of bilateral carotid arteries and internal jugular vein. The mass was found to be abutting esophagus and prevertebral fascia (Figure 1). Fine needle aspiration cytology was also done which showed features of colloid goiter.

Patient underwent total thyroidectomy and macroscopic examination showed irregularly enlarged thyroid gland right lobe measuring 9.5×6.0×3.8 cm, left lobe measuring 8.5× 5.5×4.5 cm and isthmus measuring 4.0×2.5×2.0 cm. Cut section of all appeared greyish brown to yellowish and lobulated (Figure 2).

Microscopy revealed variably sized thyroid follicles lined by cuboidal epithelium and interspersed lobules of mature adipocytes with no atypia, spread diffusely throughout the gland. Deposits of pale eosinophilic acellular material was also noted which was congophilic and showed apple green birefringence on polarized microscopy (Figure 3).



Figure 1: Diffusely enlarged thyroid gland with multiple fat density lesions.



Figure 2: Gross photograph of thyroidectomy specimen of bilateral lobes with yellowish cut surface.

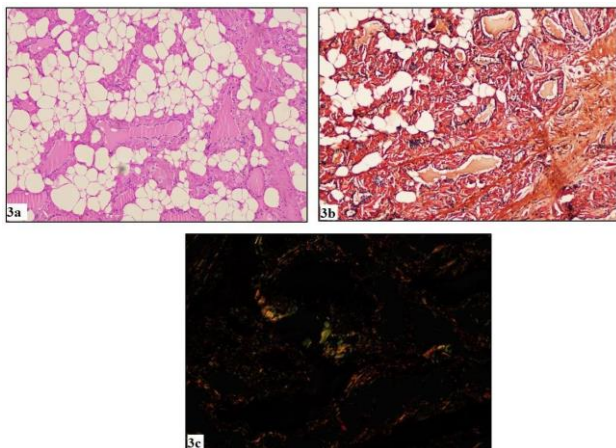


Figure 3 (A-C): Interspersed lobules of mature adipocytes with no atypia, spread diffusely throughout thyroid gland. Congophilic acellular deposits and apple green birefringence on polarized microscopy.

No evidence of systemic amyloidosis was noted in the patient after undergoing a systemic evaluation, including blood and urine parameters. A final diagnosis of amyloid goiter with diffuse lipomatosis of the thyroid gland was made. She is presently doing well on replacement dose thyroxine.

DISCUSSION

Only a few cases of diffuse thyroid lipomatosis have been reported, and still fewer cases have shown association with amyloid deposition. Patients present with a diffuse neck mass with compressive symptoms and a normal thyroid function.

Diffuse infiltration of the thyroid gland with adipose tissue was first described by Dhayagude in 1942 and has since been described in various terminologies.²

Thyrolipomatosis shows characteristic diffuse mature adipocytic infiltration among the non-neoplastic thyroid follicles. Stromal fibrosis and lymphocytic infiltration may be occasionally observed but no fibrous capsule formation has been reported.

The differential diagnosis of thyrolipomatosis include heterotopic nests of adipocytes, adenolipoma, amyloid goiter, lymphocytic thyroiditis, intrathyroid thymic or parathyroid lipoma, encapsulated papillary carcinoma and liposarcoma.³

Fat-containing thyroid lesions may be categorized into i) Nodular pattern (thyrolipoma), ii) diffuse pattern (diffuse distribution of mature adipocytes throughout the thyroid gland with unremarkable follicles, and fat infiltration also present in adenomatous nodules), and iii) combined nodular and diffuse pattern (presence of thyrolipoma, with diffuse fatty infiltration surrounding the thyrolipoma).⁴

In our case, fatty infiltration was noted throughout the lobes of the thyroid gland without any evidence of follicular adenoma. Hence a diagnosis of diffuse thyrolipomatosis was established.

The mechanism underlying thyrolipomatosis is unclear. A hypothesis proposes the existence of a metaplastic process. Adipocytes might have evolved from metaplasia of stromal fibroblasts in response to chronic tissue hypoxia or senile involution.⁵ According to Trites et al, fat lesions may have resulted from disturbed differentiation of the primitive foregut early in embryogenesis, whereas Lau et al suggests a relationship between diffuse lipomatosis and loss of expression of succinate dehydrogenase-subunit B in the follicular or adipose cells.^{6,7}

However, the detailed mechanism has not been elucidated. Additional studies are warranted to determine the mechanism underlying the pathophysiology of this rare lesion.

There is a lack of clear criteria to differentiate thyroid lipomatosis with amyloid deposits from amyloid goiters with fat infiltration.^{8,9} In our case, based on the pathology results which showed equally predominant fat infiltration as well as amyloid content, we concluded with a diagnosis of amyloid goiter with diffuse thyroid lipomatosis.

CONCLUSION

Diffuse fatty infiltration of the entire thyroid gland is a very rare finding, and the coexistence of amyloid protein deposition adds to its rarity. This report describes one such rare case of amyloid goiter with diffuse thyroid lipomatosis without any evidence of systemic amyloidosis. There is no known pathogenetic relationship between the presence of fat and amyloid protein in the thyroid which warrants further investigation.

Funding: No funding sources

Conflict of interest: None declared

Ethical approval: Not required

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Cite this article as: Vallathai S, Kuruvilla S, Lakshmi K, Parameswaran KK, Abdulla FA. Amyloid goiter with diffuse thyrolipomatosis: a rare entity. Int Surg J 2024;11:840-2.