

Amyloid Goiter: A case of primary thyroid amyloid disease

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Citation

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Abstract

Amyloid goiter can be defined as the presence of amyloid within the thyroid gland in such quantities as to produce a clinically apparent enlargement of the gland. It is a rare disease which occurs in association with both primary and secondary amyloidosis. Focal microscopic deposits of amyloid substance may often be found within the thyroid gland in association with systemic (generalized) amyloidosis, medullary carcinoma, and, less frequently, primary amyloidosis of the thyroid (1). It is a rare entity and should be suspected in patients who are known to have diseases predisposing to amyloid deposition. This condition has to be distinguished from other types of goitre and malignancy. In spite of extensive involvement, patients are usually euthyroid and diagnosis is established on histopathological evaluation. Surgical intervention is necessary to relieve the symptoms of neck mass and establish a diagnosis. The diagnosis of amyloid goitre should be suspected in patients with a diffusely enlarging thyroid gland and in those with an appropriate clinical history.

CASE SUMMARY

A 47-year-old male presented to the hospital with slowly progressive enlargement of the thyroid since 2 years. No secondary obstructive symptoms were present. The patient complained of increased sweating and decreased appetite. His thyroid function tests were, however, normal. His vital signs, including heart rate, blood pressure, and temperature, were normal. The results of cardiopulmonary and abdominal examinations were unremarkable. No neurologic deficit was found. The thyroid gland was diffusely enlarged, non-tender with no apparent nodularity. Fine-needle aspiration of the thyroid revealed features of colloid goiter. Per-operatively the gland was found to be diffusely enlarged.

The thyroidectomy specimen weighed 350 g, and measured 8.0 x 4.5 x 3.0cm. The surface was smooth. Cut section showed a diffusely enlarged thyroid with homogenous yellow to tan appearance

Figure 1

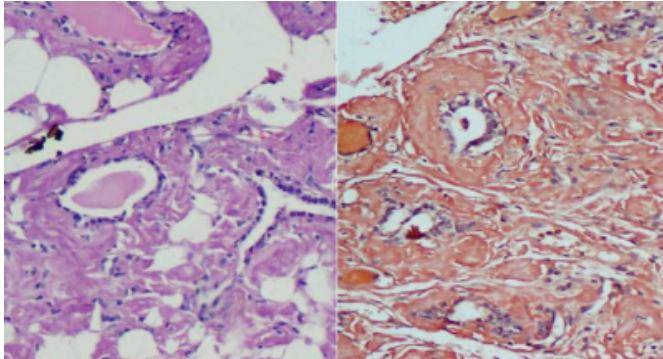
Fig 1: showing enlarged thyroid with diffuse yellow appearance



Histopathological examination showed atrophic and dilated follicles lined by low cuboidal epithelium, surrounded by abundant acellular homogenous pink substance, along with fat infiltration. These eosinophilic deposits stained positive with Congo red stain.

Figure 2

Fig 2: Photomicrograph showing atrophic thyroid follicles surrounded by abundant pink hyaline material on H&E stain (left) and positive staining with Congo red stain (right) at 200x



A diagnosis of amyloid goiter was made. Since a thorough search for the cause of amyloid deposition did not reveal any illness, a diagnosis of primary thyroid amyloidosis was considered.

DISCUSSION

We have described a patient with diffuse involvement of the thyroid gland by amyloid substance, characteristic of amyloid goiter.

Amyloid goiter may be associated with either primary or secondary amyloidosis. Primary amyloidosis involving thyroid gland is rare and is limited to case reports. It is seen in approximately 0.04% of patients with primary systemic amyloidosis. Although this entity has been identified most often in patients with amyloidosis secondary to chronic infections and other inflammatory processes, including tuberculosis, rheumatoid arthritis, cystic fibrosis, bronchiectasis, and Crohn's disease, the present case is apparently due to primary amyloidosis. In a review by Levillain et al., 56% of cases of amyloid goitre were caused by secondary amyloidosis (2).

Commonly in patients with amyloid goiter, the enlargement of the gland is relatively rapid, occurring in weeks to several months, but in our case the onset was insidious in nature. It usually presents as diffuse thyroid involvement, affecting both lobes, with a slight nodularity. The process is usually non-tender and may be accompanied by obstructive symptoms such as hoarseness, dyspnea, dysphagia, and lymphadenopathy. Thyroid function tests are often non-specifically altered, and most patients are clinically euthyroid despite the diffuse involvement by the disease. Occasionally they may be associated with symptoms of

hyperthyroidism and hypothyroidism.

The diagnosis of amyloid goiter should be considered in any patient with systemic amyloidosis presenting with an enlarging diffuse goiter and an euthyroid state. Rarely, amyloid goiter may occur as the first sign of systemic amyloidosis (3). Medullary carcinoma of the thyroid is an important diagnostic consideration, and calcitonin staining is helpful in establishing or excluding its presence.

The definitive diagnosis is by histopathological evaluation. Amyloid is usually present extracellularly as an amorphous, eosinophilic proteinaceous substance on light microscopy. In cases of amyloid goiter, amyloid material is commonly seen infiltrating the parenchyma, distorting the normal tissue architecture and eliciting a variable degree of foreign body giant-cell reaction with associated moderate to severe, often focal lymphocytic thyroiditis. Other histologic features occurring in amyloid goiter include large foci of fatty metaplasia, as in our case, and, rarely, squamous metaplasia.

Histochemical stains aid in the confirmation of amyloid. These stains include Congo red, thioflavin T, and crystal violet stains. Congo red, the most frequently used technique, imparts a unique apple-green birefringence when viewed under polarized light and is considered a pathognomonic feature of amyloid. Immunohistochemical techniques may help differentiating amyloid A from other types of amyloid (4).

In the case of amyloid goiter, careful investigation is warranted to exclude multiple myeloma or other plasma cell dyscrasias. Although no effective medical treatment exists for either form of amyloidosis, confirmation of type by immunohistochemical techniques is important, since patients with secondary amyloidosis tend to have a better prognosis than those suffering from primary amyloidosis(5)

References

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