AMYLOID GOITRE: A RARE CASE STUDY
Usharani R1, Madhusudhan BV2, Sandeep Patil3, Ashok A4, Lakkanna S5

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ABSTRACT: Involvement of the thyroid gland by amyloidosis is a common phenomenon but clinically significant enlargement of the thyroid owing to amyloid deposition is an extremely rare occurrence. Amyloid Goitre is defined as a presence of amyloid within the gland so as to produce a clinically detectable enlargement of the gland. This is an extremely rare entity, in association with both primary and secondary amyloidosis. Primary amyloidosis occurs spontaneously in the absence of apparent predisposing illness. The diagnosis of amyloid goiter should be considered in any patient with an enlarging diffuse goiter and euthyroid state. Rarely, amyloid goiter may occur as the first sign of systemic amyloidosis. Approximately 200 cases have been reported in English literature. In 50 – 80% of case, it is seen in Medullary carcinoma thyroid. It is an important etiological factor of End Stage Renal Disease. We describe the clinical and pathological feature of amyloid goiter in a patient which was insidious in onset with no significant influence on thyroid function as seen with the thyroid function tests

KEY WORDS: Amyloidosis, thyroid, Primary, secondary.

CASE SUMMARY: A 48 year old female presented to the surgical OPD with slowly progressive enlargement of thyroid since the past 2 years which was insidious in onset. The patient was asymptomatic. There were no secondary obstructive symptoms like dyspnoea, dysphagia or hoarseness of voice. On examination, her vitals – heart rate, blood pressure, temperature, pulse rate were normal. The thyroid was diffusely enlarged and non tender. Cardio-pulmonary and abdominal examination were unremarkable, including thyroid function tests. On fine needle aspiration cytology and sonological evaluation, a provisional diagnosis of multinodular goiter was made.

The patient underwent total thyroidectomy. The gland was found to be highly vascular and fragile during the intra-op period. The patient had an uneventful post-op period and recovered well. The thyroidectomy specimen weighed 380 gms, measuring 9.0 X 5.0 X 3.0 cm, with smooth surface. Cut section showed diffusely enlarged thyroid with homogenous yellow tan appearance.

Histopathological examination showed atrophic and dilated follicles lined by low cuboidal epithelium surrounded by abundant acellular homogenous pink substance along with fat infiltration, which stained positive with congo red and exhibited apple green birefringence under polarized light. No immunoreactivity was seen with calcitonin.

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. The patient subsequently developed renal failure necessitating hemodialysis.

DISCUSSION: Amyloidosis is a condition characterized by the deposition in various body tissues and organs of an amorphous, predominantly extracellular eosinophilic material called amyloid. It is seen as microscopic deposits, as plaques or as confluent masses that may progressively replace the
parenchyma of affected organs, resulting in gradual loss of function and eventually death. Such organs are enlarged, firm and pale in macroscopic appearance, with a waxy texture to their cut surface, a feature which led VonRokitansky in 1842 to refer this condition as “waxy degeneration” or “lardaceous disease” and subsequently named by Virchow in 1853 as AMYLOID, on the basis of color after staining with iodine and sulfuric acid, under the mistaken belief that the material was starch like (AMYLON-STARCH).

Amyloidosis may be primary or secondary. Primary amyloidosis occurs spontaneously in the absence of apparent predisposing illness and often affects tissues of mesodermal origin such as muscle, heart, skin, spleen, liver, kidney, thyroid, tongue etc. Secondary amyloidosis occurs with a wide range of predisposing or co-existent pathology like chronic infection-tuberculosis, inflammatory condition – rheumatoid arthritis cystic fibrosis, bronchiectasis, and Crohn’s disease, Chronic renal failure, Multiple myeloma.

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. The present case is apparently due to primary amyloidosis. In a review by Levillain et al., 56% of cases of amyloid goiter were caused by secondary amyloidosis.

Commonly amyloid goiter manifest clinically as a rapidly enlarging neck mass, firm to hard in consistency which suggests suspicion of malignancy 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 an enlarging diffuse goiter and euthyroid state. Rarely, amyloid goiter may occur as the first sign of systemic amyloidosis. Medullary carcinoma of the thyroid is an important diagnostic consideration, and calcitonin staining is helpful in establishing or excluding its presence.

Fine needle aspiration cytology facilitates the diagnosis but may be inconclusive in some cases.

The definitive diagnosis is by histopathological evaluation. Amyloid is usually present extracellularly as an amorphous, eosinophilic proteinaceous substance on light microscopy, 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, and rarely, squamous metaplasia.

Histochemical stains aid in the confirmation of amyloid which include Congo red, thioflavin T, and crystal violet. Congo red, the most frequently used and the accepted gold standard for amyloid detection, imparts a unique apple-green birefringence when viewed under polarized light (Divery and Florkin in 1927) and is considered a pathognomonic feature of amyloid. Immuno-histochemical techniques may help differentiating amyloid A from other types of amyloid.

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 the type by immunohistochemical techniques is important, since patients with secondary amyloidosis tend to have a better prognosis than those suffering from primary amyloidosis\textsuperscript{1,2}, due to controllable underlying condition.

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**REFERENCES:**
PHOTOMICROGRAPH SHOWING ATROPHIC AND DILATED FOLLICLES SURROUNDED BY PINK HYALINE MATERIAL WITH FAT INFILTERATION. LM, H & E (10 X).

PHOTOMICROGRAPH SHOWING POSITIVE STAINING WITH CONGO RED. LM (40 X).

APPLE GREEN BI-REFRINGENCE ON POLARIZATION. (200 X)
### CASE REPORT

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