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Amyloid Goiter in a Patient with Progressive Thyromegaly

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Case History

A 56-year-old female who was referred to our institution for an enlarging right-sided goiter associated with pain and pressure. Her past medical history was significant for AA amyloidosis diagnosed by kidney biopsy in 2010 after routine blood work showed an elevated creatinine. A thorough evaluation for an underlying cause, including rheumatologic disorders and chronic inflammatory conditions, has been unrevealing and she has since developed renal insufficiency and is awaiting transplant. In 2012, she presented with thyromegaly and hyperthyroidism, which was treated with radioiodine ablation. Although she was subsequently euthryoid without hormone replacement, her right-sided goiter continued to enlarge.

She was referred for surgical management in 2014; at that time, acute surgical intervention was not recommended because the patient was asymptomatic, euthyroid, and due to the significant risk the procedure posed to her chronic renal failure. A follow-up ultrasound 1 year later showed significant growth of her right thyroid lobe from 10.9 up to 12.1 cm, with a minimally heterogenous echotexture but without focal nodules, and without increased vascularity. The patient denied voice changes, dysphagia, and shortness of breath. However, she was experiencing pain and pressure and had concerns regarding the cosmetic appearance. Therefore, a thyroid lobectomy was recommended. She underwent a right thyroid lobectomy with an uneventful postoperative course.

What Is Your Diagnosis?

Histopathological Diagnosis: Amyloid Goiter

The surgical pathology examination of the specimen showed a 219.4-g right thyroid lobectomy specimen measuring 13.8×9.4×4.8 cm. The thyroid parenchyma was rubbery, yellow tan, and homogeneous without any distinct lesions or masses.

Histological examination revealed thyroid parenchyma diffusely replaced by fatty metaplasia with few remaining atrophic follicles. Rare scattered giant cells and minimal chronic inflammation were seen. An eosinophilic extracellular amorphous material was interspersed throughout the stroma, which was confirmed to be amyloid deposition by orangeo-philic staining with Congo red and positive birefringence under polarized light (Fig. 1).

Comment

We present a case of amyloid goiter in a female patient with a history of AA amyloidosis presumed to be isolated to the kidney. Amyloid goiter is defined as the enlargement of the thyroid gland due to the presence of amyloid deposition and is occasionally associated with diffuse lipomatosis [1]. The condition was first described by von Rokitansky in 1855 and was eventually termed “amyloid goiter” by von Eisenberg in 1904 [2]. Primary amyloidosis involving the thyroid gland is rare and is estimated to occur in 0.04 % of patients with primary systemic amyloidosis [3, 4]. The finding is more common in

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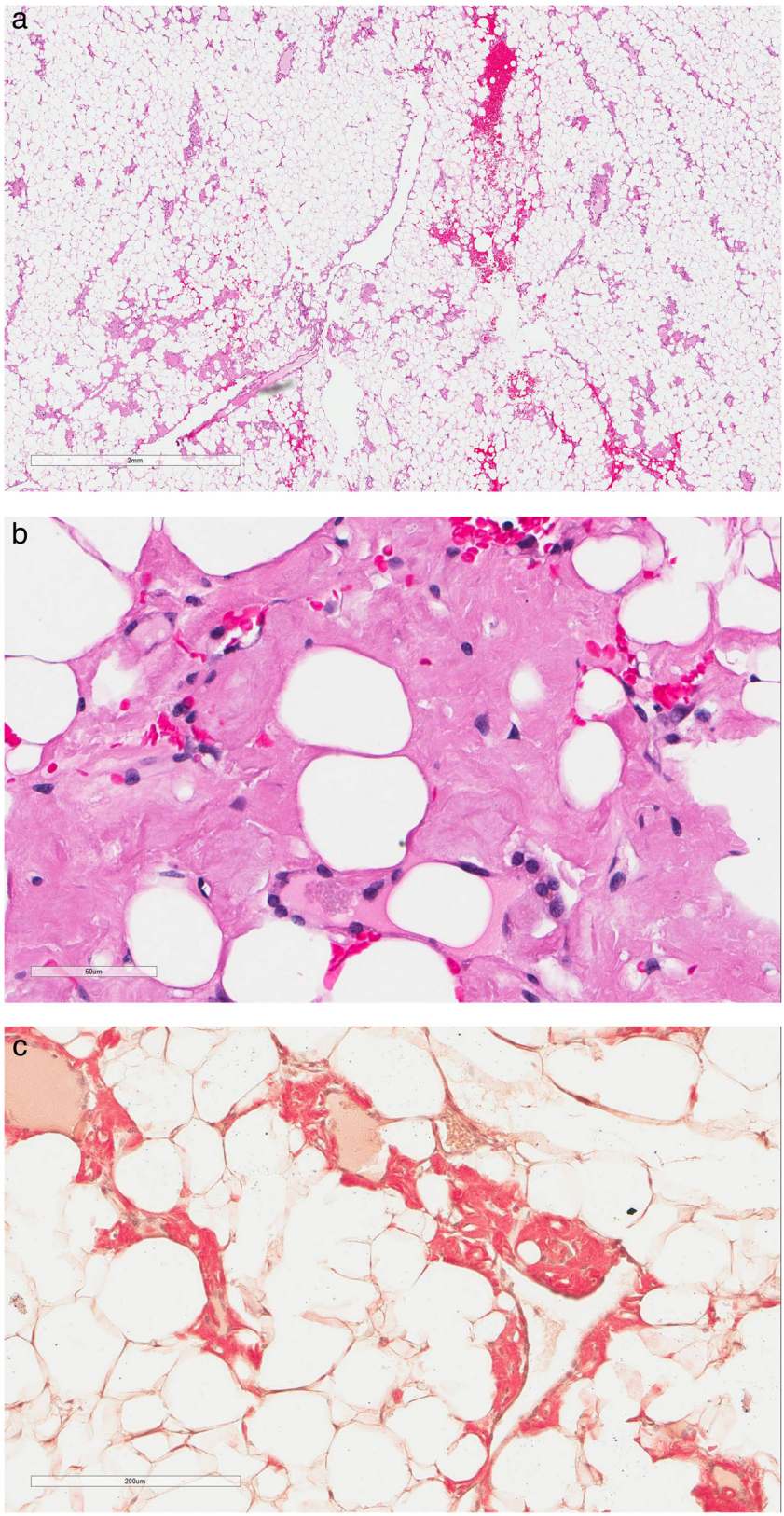

Fig. 1 Amyloid goiter showing thyroid with diffuse fatty metaplasia and scant remaining follicles (**a**). Amorphous eosinophilic material can be seen surrounding atrophic follicles as well as interspersed throughout the stroma (**b**). Congo red staining confirms the presence of amyloid (**c**)

cases of secondary amyloidosis, including long-standing inflammatory disorders, primary thyroid malignancies [5], and Familial Mediterranean fever [6]. Although microscopic amyloid deposits in the thyroid are common in these conditions,

clinically recognized cases of amyloidosis resulting in goiter are rare [7].

Amyloid goiter tends to affect both lobes and develops over a course of weeks to several months [8]. The thyroid is non-tender, and the patient may or may not develop obstructive symptoms, such as airway impingement, hoarseness, and dysphagia [1]. Thyroid function is usually preserved, but case reports have described both hypo- and hyperthyroidism [8, 9]. Imaging findings are characteristic and include diffuse and bilateral thyroid enlargement. Fat deposition leads to increased echogenicity and a fine echotexture similar to ground-glass appearance on ultrasound and low attenuation values on CT [9, 10].

Grossly, the gland appears pale due to fatty metaplasia. Adipose tissue is often found in thyroid amyloidosis. Although the origin remains unclear, the dominating hypothesis in the literature contributes this finding to metaplasia of stromal fibroblasts precipitated by ischemia due to the destruction of capillaries by amyloid deposition [1, 3].

The diagnosis can be made by FNA with Congo red staining. Treatment of amyloid goiter is often surgical, but partial regression has been reported with therapy aimed at the amyloidosis [8].

In summary, clinically significant amyloid goiters are rare but must be considered in a patient with progressive thyromegaly, especially in those with a history of amyloidosis.

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