

Fast growing goitre as the first clinical manifestation of systemic amyloidosis

J. A. AMADO*
M.D.

R. ONDIVIELA†
M.D.

S. PALACIOS‡
M.D.

D. CASANOVA‡
M.D.

J. MANZANOS‡
M.D.

J. FREIJANES*
M.D.

*Endocrine Unit, Department of Medicine, †Department of Pathology and ‡Department of Surgery, C.M.N. 'Marqués de Valdecilla', Santander, Spain

Summary

A case of systemic amyloidosis presenting as a fast growing goitre is reported. There was neither clinical nor biochemical evidence of hepatic, cardiac or renal involvement, in contrast with previously reported cases. Despite large deposits of amyloid in the thyroid the clinical and biochemical studies demonstrated a normal thyroid function.

Introduction

Systemic amyloidosis is often associated with microscopic deposits in the thyroid gland (Kennedy, Thompson and Buchanan, 1974). Far less frequently the amyloid deposits are large enough to become palpable. When this occurs, there is usually clinical or biochemical evidence of hepatic or renal involvement (Kennedy *et al.*, 1974). A case is reported in which the initial manifestation of amyloidosis was the presence of a fast growing goitre without other clinical or biochemical abnormalities.

Case report

A 73-year-old woman had noticed a fast growing cervical mass for 6 months. Mild dysphagia was present. She had lost 10 kg during this time and had anorexia and asthenia. There were no symptoms of thyroid dysfunction. She had had frequent episodes of acute bronchitis for 10 years.

On examination she appeared in ill general condition and poorly nourished. A four-fold diffuse enlarged painless hard thyroid gland was palpable. No enlarged lymph glands were found. Rhonchi and basal rales were present in both lungs. The remainder of the physical examination was unremarkable.

A complete blood count and routine biochemistry was normal. Electrophoresis showed a normal

pattern as did immunoelectrophoresis of blood and urine. Test for C-reactive protein was positive, and for rheumatoid factor, negative. Urinalysis appeared normal. Chest X-rays were suggestive of bilateral bronchiectasis. A bone radiological survey showed mild osteopenia. Serum thyroxine was 69.5 nmol/l (normal 51-141), serum triiodothyronine was 1.5 nmol/l (normal 1.53-3.07), T₃ uptake index was 1.1% (normal 0.9-1.1), serum TSH was 1 mu./l (normal 0-5 mu./l). Tanned red cells agglutination tests for microsomal and thyroglobulin antibodies gave negative results. Recent ingestion of cough syrups containing iodide prevented a thyroid scintiscan.

Because a thyroid neoplasm was suspected, a subtotal thyroidectomy was performed. The thyroid gland was irregularly enlarged, 10 × 5 × 3 cm, and weighed 65 g. The lobular architecture was distorted by lardaceous yellowish streaks extending far beyond the capsule. There were nodular uninvolved areas. Histology showed areas of almost regular morphology alternating with others where the interfollicular space was stained with congo red and exhibited apple-green birefringence under polarized light. The latter was stained with osmium tetroxide and represented neutral fat. A rectal biopsy showed amyloid deposits. Six months after surgery there were no changes, clinically or biochemically.

Discussion

Amyloid in the thyroid may be associated with systemic amyloidosis (Kennedy *et al.*, 1974), medullary carcinoma (Hazard, Hawk and Crile, 1959), microfollicular thyroid carcinoma (Valenta *et al.*, 1977) or Riedel's thyroiditis (Melato and Mlac, 1978).

Amyloid goitre is considered a rarity, although in systemic amyloidosis microscopic deposits of amyloid are frequently found in the thyroid. The largest published series reported seven cases (Kennedy *et al.*, 1974), and the present authors could not find more than 80 cases from an extensive review of the world literature. From a total of 25 cases of amyloidosis seen in their hospital in the last six years, they have found only two cases with amyloid goitre.

In most of the reported cases, goitre was present in patients with clinical or biochemical evidence of renal or hepatic involvement (Kennedy *et al.*, 1974). The present patient developed a rapidly growing goitre as the first clinical manifestation of systemic amyloidosis, which is unusual. In all other aspects, the clinical presentation was similar to that commonly reported—a rapidly growing, painless, diffuse goitre, absence of clinical and biochemical evidence of thyroid dysfunction and negative antithyroid antibodies. Most of these patients, like the present one, have been operated upon because of suspicion of thyroid cancer; in other cases an iodide goitre was suspected (Kennedy *et al.*, 1974).

The amyloidosis in this patient is probably related to the presence of bronchiectasis. Plasma cell dyscrasia, a common cause of amyloidosis, can be ruled out here in view of the normal findings on blood and urine immunoelectrophoresis.

In recent years amyloid goitre has been recognized as a complication of systemic amyloidosis associated with familial Mediterranean fever, and Danovitch *et al.* (1979) stated 'it is possible that as haemodialysis continues to prolong lives of patients with systemic amyloidosis, organs not usually associated with amyloid deposition will become involved'.

Amyloid deposits usually appear in previously normal thyroids, although they can occur in patients with Hashimoto's thyroiditis (Wuchter and Keller, 1978). The cause of the rapid growth of amyloid goitre is unclear. Rarely, amyloid infiltration can involve laryngeal nerves or cricopharyngeus muscle causing dysphagia and dyspnoea (Shapiro, Kohut and Potter, 1971).

Thyroid function in these patients is usually normal, although there are no detailed biochemical studies (Kennedy *et al.*, 1974). Jaimet (1951) described a case of amyloid goitre with hypofunction of the gland based on a 12% uptake of ¹³¹I in 24 hr and a 6% conversion ratio.

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