Amyloid tumor of thyroid as a first manifestation of systemic amyloidosis: A diagnosis to keep in mind

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ABSTRACT

Amyloidosis is an abnormal extracellular deposit, which can occur in several tissues. Although involvement of the thyroid gland by amyloid is a relatively common phenomenon, clinically significant enlargement of the thyroid due to amyloid deposition is an extremely rare occurrence. About 250 cases of amyloid goiter have been reported in the literature. We report a rare case of a 15-years-old girl, diagnosed as a case of Crohn’s and intestinal tuberculosis diseases and has been on treatment. She developed a goiter without compressive complications. Histologic examination concluded to diffuse amyloid deposition surrounding thyroid follicles. Confirmation of amyloid was made by the presence of immunoreactivity that was seen with AA protein. The prognosis depends on the treatment of the amyloidosis and the underlying chronic disease.

Keywords: Amyloid goiter, Thyroid gland, Amyloid, Histology, Crohn’s disease, Tuberculosis.

CASE REPORT

A 15-years-old girl, with a history of intestinal tuberculosis treated with anti-tuberculous Therapy for 12 months was referred to ENT department for cervical tumefaction. She had no previous history of a thyroid disease or the use of medications known to induce thyroid dysfunction, irradiation, and no family history of thyroid disease. Her mother noticed neck swelling with right neck tenderness which had enlarged rapidly two weeks prior to admission. Physical examination found an elastic, firm goiter primarily in the right lobe of the thyroid. The abdominal organs were enlarged. The laboratory data revealed hypochromic anemia and hypoalbuminemia. Her renal function was normal, with the exception of a slightly elevated urinary protein level (0.3 g/l). Her serum free thyroxine (fT4) and serum thyroid stimulating hormone (TSH) were both normal. Ultrasonography of her thyroid gland revealed an enlarged goiter (estimated thyroid volume: 62 ml), particularly in the right lobe, with irregular hypoechoic region in the right lobe. CT showed marked thyroid enlargement with cystic non-enhancing areas surrounded by hyperdense tissue which enhanced markedly after contrast (Figures 1,2). Fine needle aspiration confirmed the intracystic haemorrhage. Renal ultrasonography revealed normal.

The patient underwent a total thyroidectomy. A diagnosis of thyroiditis was initially considered, but was ruled out by the final result of histology which pointed rather to thyroid amyloidosis. The interstitial tissue was expanded by an acellular eosinophil deposit. A few mature adipocyte clusters were observed between the thyroid vessels. The deposit was marked brick-red on Congo red staining, showing yellow-green double refraction under polarized light. Immunohistochemistry classified the deposit as AA. The patient was referred to internal medicine department and she was administered Colchicine one capsule per day.

Figures 1,2 : CT scan showing marked thyroid enlargement particularly in the right lobe with cystic areas separated by septa of tissue which enhanced markedly after the injection.

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DISCUSSION

Chronic serum SAA elevation is the basic determining factor in the onset of amyloidosis in chronic inflammatory disease. But not all patients with chronic inflammatory disease and prolonged serum SAA elevation go on to develop amyloidosis [1]. There must therefore be further genetic and environmental factors at work [2]. As chronic infectious disease becomes rare and many chronic inflammatory diseases are better controlled, the epidemiology of this complication of inflammation is changing. While AA amyloidosis is becoming less frequent, its prognosis, when it occurs, remains poor. Commonly, amyloidosis tends to involve the heart, kidneys, lungs, gastro-intestinal tract or peripheral nervous system. Secondary systemic AA amyloidosis involves the thyroid in 80% of cases [3]. Occasionally in Amyloid Goiter, the amyloid can also involve the parathyroid glands, as reported one review [4]. Infiltration by microscopic amyloid deposits is often discovered serendipitously, in thyroidectomy specimens or on autopsy. The deposits are asymptomatic, causing neither goiter nor dysthyroidism even if in a minority of cases a hypothyroid or hyperthyroid state is detected [5]. The quantity of amyloid deposits is rarely such as to impact thyroid volume. Only some 250 cases of amyloid goiter have indeed been reported worldwide [5]. If symptomatic, it may cause dysphagia, dyspnea, and hoarseness. Classically in patients with amyloid goiter, the enlargement of the gland is relatively rapid, occurring in weeks to several months [6].

Amyloid goiter may be diagnosed by fine needle aspiration which is a sensitive and safe procedure. Main differential diagnosis is medullary carcinoma, in which amyloid deposit is also found in 50% to 80% of cases. Fine needle aspiration can, however, exclude other thyroid cancers, particularly anaplastic one and lymphoma [7]. But this was not the case in our patient, perhaps because samples were obtained from cystic areas and amyloid is mainly present in solid ones.

The fatty infiltration, usually associated to amyloid infiltration, causes hyperechogenicity on ultrasonography and a diffuse hypodensity in the CT attenuation [8]. The most striking features in known cases were the marked thyroid enlargement and the presence of cystic lesions which appeared as discrete hypodense masses on CT with very high signal intensity on T1 and T2-weighted images. Some cysts were hyperdense on CT scan as our patient, with internal liquid-liquid levels due to spontaneous bleeding. This has been considered characteristic finding by some authors [9]. Both CT and MR could show the involvement of the thyroid and neighbouring organs including secondary tracheal compression [9].

Anatomopathology enables positive diagnosis in case of amyloid deposit detected as an amorphous substance showing yellow-green double refraction under polarized light on Congo red staining. Amyloidosis typing is based on immunohistochemistry, as in the present case. Eradicating infectious diseases such as tuberculosis is of preventive value with regard to AA amyloidosis. Regular monitoring of serum SAA could help in controlling inflammation and preventing amyloidosis [10]. Once amyloidosis has taken hold, etiological management of the underlying pathology remains primary, reducing the availability of amyloid protein precursor so as to halt deposit progression is at present the main treatment strategy in all forms of amyloidosis [10].

CONCLUSION

Amyloid goiter should be suspected in all patients with a progressive, rapidly growing, thyroid enlargement and a concomitant history of chronic inflammatory processes. In many cases fine-needle aspiration can be performed to exclude malignant lesions. In order to diagnose amyloid goiter definitively thyroidectomy is often necessary. Positive diagnosis of AA amyloidosis requires combined histologic data, and immunohistochemistry, for differentiation of other varieties, since patients with secondary amyloidosis tend to have a better prognosis than those suffering from primary amyloidosis. Once the disease established, prognosis remains poor. Progress in treatment is needed, and notably the development of drugs targeting the early stages of amylogenesis.

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REFERENCES