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CASE REPORT

Amyloid Goiter with Diffuse Lipomatosis

[1]Bulent Citgez, [1]Mehmet Uludag, [1]Gurkan Yetkin, [1]Ismail Akgun, [2]Senay Yener
[2]Nedim Polat, [3]Adnan Isgör

[1]Department of General Surgery, Sisli Etfal Training and Research Hospital, Istanbul, Turkey
[2]Department of Pathology, Sisli Etfal Training and Research Hospital, Istanbul, Turkey
[3]Department of General Surgery, Istanbul Golden Horn University, Istanbul, Turkey

Correspondence: Bulent Citgez, Department of General Surgery, Yavuklular Sokak Gul, Apartment No. 1, D-3 Bakirkoy, Istanbul Turkey, Phone: 90-505-2376847, Fax: 90-212-5713570, e-mail: bcitgez@yahoo.com

ABSTRACT

We report a case of a 37-year-old man who presented with a neck swelling associated with recent onset respiratory distress. The patient was receiving hemodialysis due to chronic renal failure three times a week for 8 years. Local examination showed a diffuse thyroid enlargement. The thyroid function test results of the patient were within normal limits. At the ultrasonography examination, there was a 17 x 11 mm nodule at the isthmus with a hypoechoic halo containing calcified foci. The fine needle aspiration biopsy (FNAB) of the nodule at the isthmus that contained calcification revealed adenomatous nodule rich in cells, which showed degeneration and follicular neoplasia. Total thyroidectomy was planned due to the patient's ongoing symptoms and dyspnea. The pathological examination revealed diffuse lipomatosis and amyloidosis.

Keywords: Lipomatosis, Thyroid, Amyloid, Surgery.

INTRODUCTION

Diffuse lipomatosis of thyroid gland is a rare entity, which was first reported by Dhayagude in 1942.[1] Adipose tissue is occasionally found beneath the thyroid capsule and in the neighborhood of blood vessels in normal thyroid gland.[1] Fat deposition frequently accompanies amyloid storage, but diffuse enlargement of thyroid gland due to lipomatosis is a very rare situation.[2] It may be asymptomatic or cause nonspecific symptoms clinically. In this paper, we report a case that was operated due to goiter, which has caused dyspnea, and diagnosed as diffuse lipomatosis and amyloid goiter by histopathological examination.

CASE REPORT

A 37-year-old male patient presented to our general surgery outpatient clinic with swelling at the neck and dyspnea. The patient was receiving hemodialysis due to chronic renal failure three times a week for 8 years. The thyroid function test results of the patient were as follows—T3:1.41 ng/ml (0.87-1.78), T4:6.66 ug/dl (6.09-12.23), TSH:1.05 uIU/ml (0.34-5.60). The results were within normal limits. At the ultrasonography examination, the dimensions of the thyroid gland were extremely increased; the right lobe measured 48 × 38 mm, the left lobe measured 36 × 41 mm, and the isthmus measured 24 mm. The echogenicity of the parenchyme of the thyroid gland was homogeneous. There was a 17 × 11 mm nodule at the isthmus with a hypoechoic halo containing calcified foci. At the neck computed tomography (CT), the dimensions of both thyroid lobes, the right lobe being more significant, and the isthmus were extremely increased. The fine needle aspiration biopsy (FNAB) of the nodule at the isthmus that contained calcification revealed adenomatous nodule rich in cells, which showed degeneration and follicular neoplasia. Total thyroidectomy was planned due to the patient's ongoing symptoms and dyspnea. Both thyroid glands were found to be enlarged diffusely, weighing 304 gm, and nodules that contained milimetric calcifications were detected at isthmus. The microscopic evaluation of the hematoxylin-eosin (HE) stained sections pointed out that the thyroid follicles were mostly removed, and there was a widespread accumulation of amorphous eosinophilic material (Fig. 1). With the crystal violet dye performed by histochemical technique, this deposit was stained with purple color (Fig. 2) with these findings, the case was diagnosed as secondary amyloidosis. The patient was externed at the 3rd day without any postoperative complication. The pathological examination revealed diffuse lipomatosis and amyloidosis. At the postoperative controls, we learned that the patient had been diagnosed with rheumatological fever, had open kidney biopsy 22 years ago, and the biopsy revealed amyloid deposits.

DISCUSSION

Amyloid infiltration of the thyroid was first reported in a case with systemic amyloidosis by Rokitansky in 1855. Beckmann reported a case with enlargement of the thyroid gland due to amyloid infiltration in 1858 and these cases were defined as amyloid goiter since then.[2,3] Amyloidosis is examined in two major titles ethiopathologically as primary or secondary amyloidosis. In primary amyloidosis, there is an increase in monoclonal plasma cells at the bone marrow, and in some cases it is accompanied by multiple myeloma. Immunoglobulin light chains (AL type amyloidosis or primary amyloidosis) cause

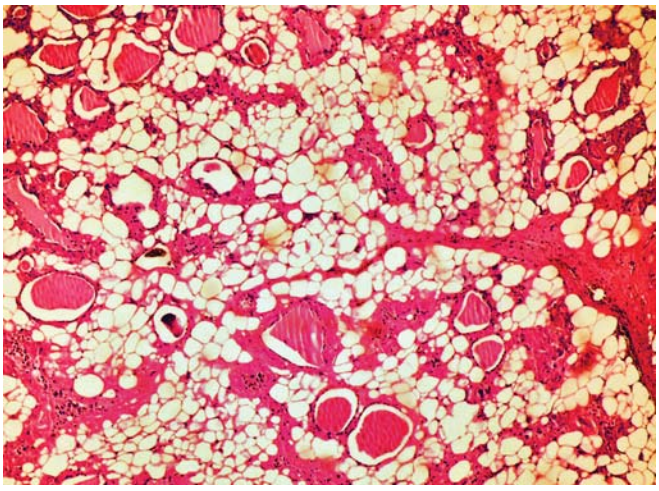

Fig. 1: The microscopic evaluation of the hematoxylin and eosin (H & E) stained sections pointed out that the thyroid follicles were mostly removed, and there was a widespread accumulation of amorphous eosinophilic material (H & E × 20). 203 × 152 mm (160 × 160 DPI)

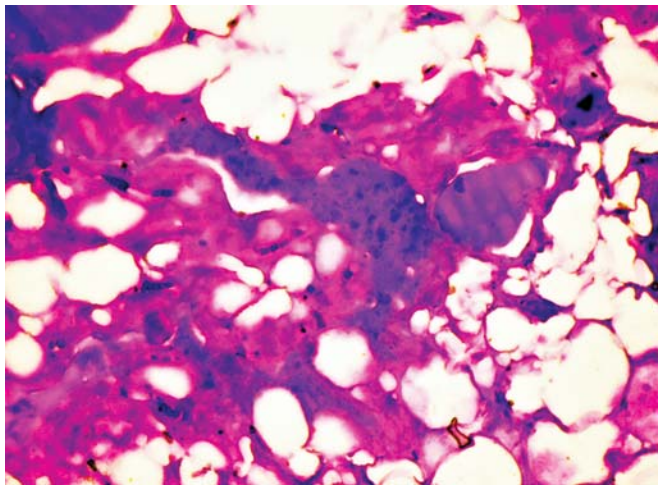

Fig. 2: With the crystal violet dye performed by histochemical technique, this deposit was stained with purple color and with these findings, the case was diagnosed as secondary amyloidosis. 203 × 152 mm (160 × 160 DPI)

amyloid accumulation in tissues. However, the secondary amyloidosis term is used in the case when the disease is related with a chronic inflammatory condition (e.g. tuberculosis, osteomyelitis, rheumatoid arthritis, Crohn disease, bronchiectasy, etc.). Serum amyloid A (SAA) protein, which is an acute phase reactant, causes secondary amyloidosis.[4,5] SAA circulates in blood through the body and in some tissues (e.g. kidney, gastrointestinal system, thyroid, testicles and heart), it cannot be metabolized enough due to the surrounding factors and accumulates as AA (amyloid associated) peptides. These AA peptides join to form the amyloid fibrils.[4,6] Amyloid deposits can be shown by dyes, such as Kongo red and crystal violet. In addition, it is possible to detect the type of the amyloid by immunohistochemical and electron microscopic studies. In our case, AA type amyloid deposits were detected with dyes. Amyloid infiltration of thyroid can be seen in medullary thyroid carcinoma, foreign body reaction and systemic amyloidosis. Systemic amyloidosis may cause dysfunction of the vital organs, such as heart or kidney; in the thyroid gland, amyloid infiltration can be seen 50 to 80% microscopically.

In a few number of cases named as amyloid goiter, which was reported by Beckmann for the first time in 1858, there was a goiter view clinically. The majority of these cases were due to secondary amyloidosis. In the study of Himmetoğlu et al[3] secondary amyloidosis was detected in 11 of 18 cases with amyloid goiter. In our case, secondary amyloidosis developed due to rheumatological fever.

Diffuse lipomatosis develops very likely from the heterotopic foci at the thyroid gland during embryogenesis, and rarely accompanies amyloid goiter.[7] In the literature, although the relationship between the amyloid and fat accumulation is not clear, it is pronounced that the amyloid infiltration may cause CRF (chronic renal failure).[7]

Himmetoğlu et al found CRF in 3 of 18 patients with amyloid goiter, as in our case.[3] Amyloid goiter might be accompanied by fat deposition of varying extent and this fatty infiltration in a previous background of amyloid goiter has been explained in two ways. One hypothesis suggests stimulation of stromal fibroblasts to become adipose cells due to tissue hypoxia caused by amyloid deposition and destruction of thyroid follicles.[4] The other hypothesis suggests progressive capillary destruction by amyloid deposition as the primary occurring event.[4] The present case is a unique example of diffuse lipomatosis probably on a background of secondary amyloidosis due to chronic renal failure.

In the same study, the absence of the fat tissue infiltration at the early stage of the amyloid accumulation is explained by the progressive capillary damage that was caused by the amyloid in the fatty transformation.[7,8]

Amyloid goiter may develop between weeks and years, and usually affects both lobes. It may clinically cause mass effect due to enlargement, as in our case. Despite the diffuse involvement, the patients are usually euthyroid.[7] Our case had also normal thyroid function test results. The FNAB of the nodule may not establish the diagnosis, as in our case, the FNAB of the nodule at the isthmus revealed the cellular smear.

The amount of the fatty infiltration due to amyloid is still unclear. Diffuse fatty proliferation at the thyroid gland is sometimes accompanied with amyloid accumulation. This proliferation is thought to develop from the adipose cells, not migrating and staying in the thyroid gland at the embryogenesis.[8] Thyroid lesions containing adipose tissue are rare, and the reported cases generally fall into two groups: Fat-containing thyroid neoplasms and non-neoplastic conditions; the latter includes amyloid goiter, lymphocytic thyroiditis, goiter, thyroid atrophy, Graves' disease, adenolipoma and diffuse lipomatosis.[3-5] Diffuse lipomatosis is a form of fat deposition, first reported by Dhayagude in 1942[6] and only five cases have been reported in the English literature.[7] It is characterized by goiter due to diffuse proliferation of adipose tissue in the thyroid gland rather than being secondary to proliferation of the follicular epithelium. It might be accompanied by amyloid deposition.

As a result, in the amyloid goiter cases, systematic examination should be performed to bring out the underlying disease, and in the cases with systemic amyloidosis, it should be kept in mind that the amyloid may accumulate in the thyroid gland, and in addition, the amyloid infiltration may be due to benign conditions.

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