

Secondary Amyloidosis with Amyloid Goitre Presenting as a Multinodular Goitre

Multinodüler Guvatr Tanısı ile Başvuran Sekonder Amiloidoza Bağlı Amiloid Guvatr

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Abstract

Amyloid goitre is a rare entity defined by the presence of amyloid within the thyroid gland in such quantities as to produce clinically apparent enlargement of the gland. It affects the thyroid in a bilateral and diffuse manner. Pre operatively it simulates a multinodular goitre and surgical intervention is often necessary to establish a diagnosis and to relieve compressive symptoms of a neck mass. We present a case of 15 year old female with juvenile rheumatoid arthritis (JRA) who presented with a thyroid enlargement diagnosed only post operatively on histopathological examination. *Turk Jem 2007; 11: 119-20*

Key words: Amyloid, goitre, thyroid

Özet

Amiloid guatr klinik olarak tiroid bezinin anlamlı ölçüde büyümesine yol açacak düzeyde amiloid birikimi sonucu ortaya çıkan nadir görülen bir durumdur. Tiroidi bilateral diffüz olarak etkiler. Preoperatif multinodüler guatrı taklit eder ve boyundaki kitlenin bası etkisini kaldırmak ve tanı için cerrahi müdahale gereklidir. Burada 15 yaşında juvenil romatoid artritli tiroid büyümesi ile baş vuran ve postoperatif histopatolojik inceleme ile tanı konan bir kız çocuğu sunulacaktır. *Turk Jem 2007; 11: 119-20*

Anahtar kelimeler: Amiloid, guatr, tiroid

Introduction

Amyloid goitre is a rare entity defined by the presence of amyloid within the thyroid gland in such quantities as to produce clinically apparent enlargement of the gland. It affects the thyroid in a bilateral and diffuse manner. Pre operatively it simulates a multinodular goitre and surgical intervention is often necessary to establish a diagnosis and to relieve compressive symptoms of a neck mass. We present a case of 15 year old female with juvenile rheumatoid arthritis (JRA) who presented with a thyroid enlargement diagnosed only post operatively on histopathological examination.

Case Report

We present a case of 15-year old female, known case of juvenile rheumatoid arthritis (JRA) who presented with symmetrical thy-

roid enlargement and dyspnoea. Because of the obstructive symptoms caused by the diffusely enlarged gland, clinically consistent with goitre, and a fine needle aspirate (FNA) of the thyroid nodule reported as adenomatous goitre, the patient was taken up for an elective thyroidectomy. During surgery, a large multinodular goitre was identified, which circumferentially encased the trachea. Subtotal thyroidectomy was performed.

Grossly both lobes were enlarged weighing 27.7g and completely encapsulated. Cut surface of both lobes and isthmus revealed diffuse involvement of the gland with multiple lobulated nodules, 2 centimeters in greatest dimension. Cut surface revealed multiple hemorrhagic and congested areas simulating a multiple nodular goitre (Figure 1). No preserved normal appearing thyroid could be identified.

Microscopic examination of hematoxylin-eosin-stained sections taken from both lobes of the thyroid revealed extensive infiltration of the parenchyma by eosinophilic amorphous material con-

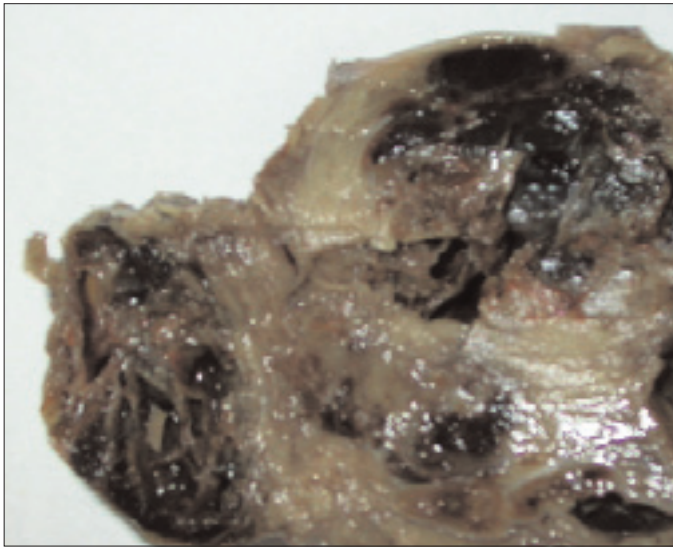


Figure 1. Gross picture thyroid specimen showing variegated cut surface with hemorrhagic, necrotic and congested areas

sistent with amyloid substance (Figure 2). This material stained intensely with Congo red (Inset) and showed apple green birefringence under polarized light. There was no evidence of C-cell hyperplasia or medullary carcinoma. There was extensive fat cell metaplasia in the thyroid interstitium. A bone marrow biopsy and gingival biopsy were done to look for systemic involvement. Both were positive for amyloid confirming widespread involvement.

Amyloid infiltration of the thyroid gland in patients with systemic amyloidosis was first reported by Rokitsky in 1855, and later confirmed by Virchow (2). Focal microscopic deposits of amyloid substance may also be seen in cases of medullary carcinoma and less frequently, primary amyloidosis of the thyroid. Diffuse enlargement due to amyloid infiltration is infrequent. Beckmann first reported the presence of clinically detectable thyroid enlargement because of amyloid deposition in 1858, followed by Eiselberg in 1904, who later coined the term 'Amyloid Goitre' (3). Once amyloid goitre was recognized as a specific entity, around 140 cases had been reported till 1993 (3, 4). It usually presents with a rapidly growing neck mass causing pressure symptoms. This condition has to be distinguished from other types of goitre and malignancy. Patients are usually euthyroid, despite extensive involvement of the gland. In contrast to malignant thyroid tumors that are often unilateral, amyloid goitre affects in a bilateral and diffuse manner. FNAC may facilitate the diagnosis of amyloid goitre, but is inconclusive in some cases (5). Surgical

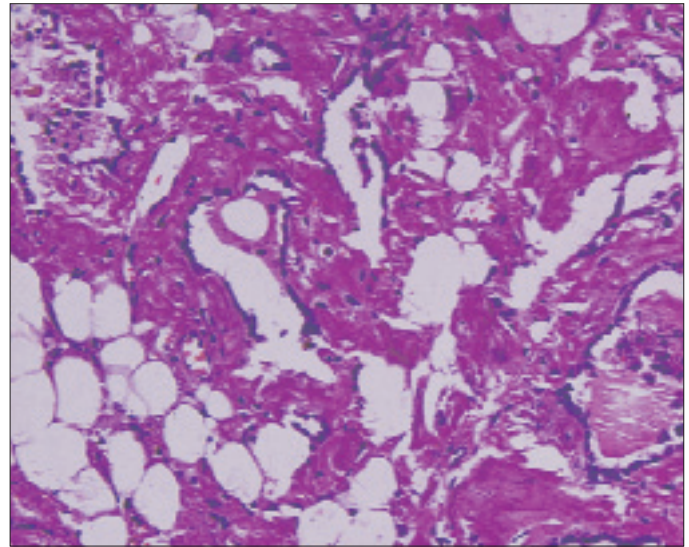


Figure 2. Infiltration of the parenchyma by eosinophilic amorphous material consistent with amyloid along with rare preserved thyroid follicle. (H&E100 Congo red positivity. (Inset)

intervention serves to establish a diagnosis, as well as for relieving the symptoms associated with the large neck mass. Uncommonly, amyloid goitre can present as a first manifestation of systemic amyloidosis (6,7). Further work up in the form of gingival, rectal, bone marrow biopsy or kidney biopsy helps to establish an underlying systemic involvement. Autoimmune disease and chronic remitting infections are the most common causes leading to secondary deposition. No medical treatment has been effective for amyloidosis, though the overall prognosis is better with patients with secondary amyloidosis (7).

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