Case Report / Olgu Sunumu

Coexistence of thyroid hemiagenesis, nodular goitre and papillary carcinoma

Tiroid hemiangenez, nodüler guatr ve papiller karsinom birlikteliği

Gülden Yenice Karatağ, M.D.,[1] Zeynep Kırkali Albayrak, M.D.,[2] Hülya Kapran Önay, M.D.,[2] Ozan Karatağ, M.D.,[3] Önder Peker, M.D.[4]

[1]*Department of Radiology, Çanakkale State Hospital, Çanakkale, Turkey*
[2]*Neoson Imaging Center, Istanbul, Turkey*
[3]*Department of Radiology, Medicine Faculty of Çanakkale Onsekiz Mart University, Çanakkale, Turkey*
[4]*Nişantaşı Pathology Group, İstanbul, Turkey*

In this article, we report a very rare case of left thyroid lobe with agenesis, adenomatous hyperplasia and coexisting papillary carcinoma of the right lobe. A 59-year-old asymptomatic woman with no previous thyroid surgery was evaluated sonographically. Ultrasonography and scintigraphy revealed an agenesis in the left lobe and multiple right lobe nodules with one of them diagnosed with papillary carcinoma.

Key Words: Fine needle aspiration biopsy; papillary thyroid carcinoma; scintigraphy; thyroid hemiagenesis; ultrasound.

Bu makalede çok nadir bir agenezili sol tiroid lobu, adenomatöz hiperplazi ve sağ lobda papiller karsinom birlikteliği olgusu sunuldu. Daha önce tiroid cerrahisi geçirmemiş 59 yaşında asemptomatik bir kadın hasta sonografi ile değerlendirildi. Ultrasonografi ve sintigrafide sol lobda agenezi ve sağ lobda birine papiller karsinom tanısı konulan çoklu nodül tespit edildi.

Anahtar Sözcükler: İnce iğne aspirasyon biyopsisi; papiller tiroid karsinomu; sintigrafi; tiroid hemiagenezi; ultrason.

Thyroid hemiagenesis (TH) resulting from the failure of embryologic development of one thyroid lobe is a very rare anomaly. Thyroid ultrasound studies report the prevalence of this morphological abnormality at between 0.02-0.05% in healthy children.[1,2] It is usually discovered incidentally during the investigation of accompanying thyroid disorders or routine medical checkup. The coexistence of TH and papillary carcinoma is an extremely rare entity. We report a case of left thyroid lobe agenesis and accompanying adenomatous hyperplasia and papillary carcinoma of the right lobe which were all incidental findings in a patient.

CASE REPORT

A 59-year-old female without any complaints and no history of previous thyroid surgery was evaluated sonographically during a routine medical checkup. Serum thyroid hormone (T_3, T_4), thyroid

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Correspondence / İletişim adresi: Ozan Karatağ, M.D. Onsekiz Mart Üniversitesi Tıp Fakültesi Radyoloji Anabilim Dalı, 17100 Kepez, Çanakkale, Turkey.
Tel: +90 532 - 352 77 28 e-mail *(e-posta):* okaratag17@gmail.com

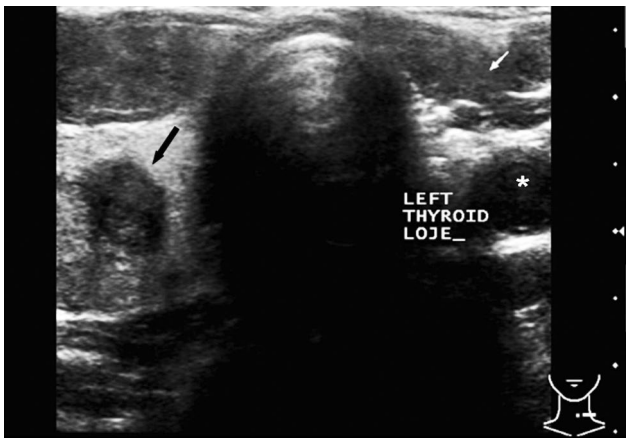

Figure 1. *Transverse ultrasound image shows that the left thyroid lobe is absent (white arrow). Ipsilateral common carotid artery (asterisk) and a hypoechoic nodule of the right thyroid lobe (black arrow) are also visible.*

stimulating hormone (TSH) and thyroid antibody values were within normal limits.

On ultrasound examination, the right lobe and isthmus were present while the left lobe was absent (Figure 1). Technetium 99m (Tc-99m) scintigraphy confirmed the absence of the left thyroid lobe. On the subtraction images, hot spot uptake was observed in the inferior pole of the right lobe with suppression in the rest of the parenchyma (Figure 2).

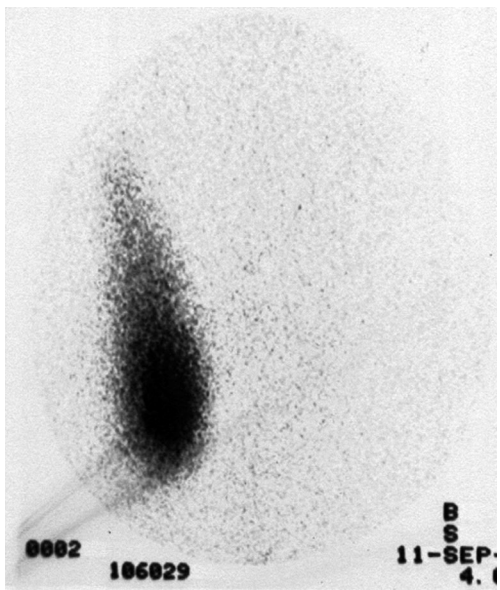

Figure 2. *Tc-99m scintigraphy demonstrates the absence of the left thyroid lobe and hot spot uptake at the inferior pole of the right lobe.*

Sonography revealed multiple isoechoic and hypoechoic nodules in the right lobe. One of the nodules measuring 0.9x0.7 cm showed spiculated margins and marked hypoechogenicity including microcalcific foci with irregular and increased vascularization on color Doppler ultrasonography indicating probable thyroid malignancy (Figure 3).

Fine needle aspiration biopsy of this nodule revealed papillary thyroid carcinoma which was also confirmed histopathologically following a right total thyroidectomy. The remaining nodules of the right lobe were histopathologically diagnosed as adenomatous hyperplasia.

DISCUSSION

Thyroid hemiagenesis is a rare variant of thyroid dysgenesis in which one of the thyroid lobes fails to develop due to failure of embryological development.[3] The occurrence of some cases of TH among members of the same family suggests the role of genetic factors.[4,5] The molecular mechanisms leading to the formation of the two thyroid symmetrical lobes, which are impaired in the case of hemiagenesis, are not known.[6] Recently several genes have been found to be involved in thyroid morphogenesis and descent. Thyroid transcription factors (TTF-1 and TTF-2) and PAX-8 are reported to be responsible genes for ectopy.[7,8] However, these have yet to be studied in hemiagenesis.

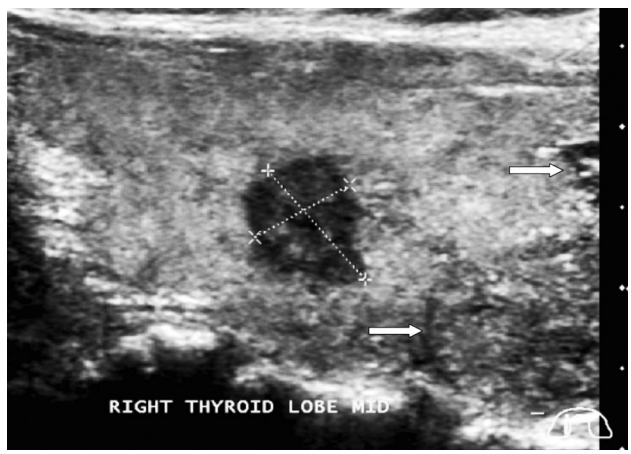

Figure 3. *A marked hypoechogenic nodule with spiculated margins at mid-zone of the right lobe is visible. Multiple hyperechoic punctate foci within the nodule represent microcalcifications. Additionally another two nodules are partially included in the sonographic image at the inferior pole of the right lobe with relatively well defined borders (white arrows).*

The detection of TH in the normal population was expected to increase with the widespread use of ultrasound, and recent ultrasound studies have reported 0.05% prevalence of this abnormality.[1,9,10] Shabana et al.[2] estimated the prevalence of TH in normal children at 0.02%. Mikosch et al.[10] reported the prevalence of TH between 1:1900 and 1:2675 with a left to right ratio of 3.6:1 and isthmus present in 44%. The absence of the left lobe was detected in 80% of cases and agenesis of isthmus in 44-50%. The female-to-male ratio was reported as 3:1;[10,11] however, the larger number of females may be biased due to female predominance of the populations investigated.[10]

A coexistent thyroid disorder is common in patients diagnosed with TH. Because of this, it is frequently diagnosed during investigation of accompanying thyroid disorders.[12] When the remaining thyroid function is euthyroid as seen in our case, it is found incidentally.[3] Among the pathologic conditions commonly occurring in the remaining lobe are thyroid adenoma, nodular goiter, hyperfunctioning adenoma, Graves disease, and chronic lymphocytic thyroiditis.[12,13] In the literature, the most common pathology involved in TH is hyperthyroidism,[11,14,15] but a recent study by Berker et al.[16] reported nodular goiter as the most frequent thyroid disease accompanying TH.

Papillary thyroid carcinoma is the most common histological type of differentiated thyroid cancer and accounts for 80% of all thyroid cancers.[17] In a retrospective study the malignant sonographic features of a thyroid nodule was defined as spiculated margin, marked hypoechogenicity and microcalcification content.[18] In our case the nodule diagnosed with papillary carcinoma showed all of these malignant sonographic features consistent with the literature.

The coexistence of TH and papillary thyroid carcinoma is extremely rare,[13] with few reported cases to our knowledge.

Thyroid hemiagenesis can be diagnosed by imaging methods including ultrasonography, computed tomography, magnetic resonance imaging, and thyroid scintigraphy.[13] However there are several clinical conditions mimicking TH on scintigraphic examination. Autonomously functioning nodules with suppressed normal thyroid tissue, primary or secondary neoplasms, infiltrative diseases such as amyloidosis, and unilateral inflammations of one lobe can mimic thyroid agenesis.[11] In such a situation additional imaging modalities can be helpful in differential diagnosis. Ultrasonography is mostly used for this purpose because it is cost effective, can be performed easily and does not expose patients to radiation.[12] If there is any associated nodular thyroidal disease, fine needle aspiration biopsy should be performed in order to rule out malignancy.

Declaration of conflicting interests

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