Editorial

Thyroid hemiagenesis

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Thyroid hemiagenesis (TH), characterized by the total absence of one lobe, is a very rare congenital anomaly of the thyroid gland. The remaining lobe generally has normal function, such that patients with TH are usually asymptomatic. The pathogenesis and clinical significance of this malformation remain undefined, and specific clinical recommendations are lacking, especially for asymptomatic cases. Therefore, discovering TH in asymptomatic cases is only possible using a screening program with various imaging modalities (1). In clinical practice, TH is usually established during evaluation of patients with symptomatic thyroid pathology (2,3). It is difficult to establish the true prevalence of TH in the normal population due to the normal functioning nature of the remaining lobe of the thyroid gland. Establishing this prevalence would require a screening program on a very large population. Suzuki *et al.* (4) performed a cross-sectional study of 3.5 years of duration on approximately 300,000 children and young adults to screen for congenital anatomic abnormalities of the thyroid and establish the prevalence of structural changes such as hemiagenesis and agenesis. They reported the prevalence of TH as 0.02% and of agenesis as 0.004% (4). This was the largest study reported in the literature for asymptomatic cases of TH. Some previous studies using ultrasound screening reported a prevalence of TH between 0.025% and 0.5% in the asymptomatic population (1,5-7). The incidence of TH is different in patients presenting with thyroid disorders. Gursoy *et al.* (5) reported an incidence of 0.25% in 4,833 patients evaluated for thyroidal pathologies. Berker *et al.* (8) identified ten cases (0.16%) of hemiagenesis in 6,242 patients with thyroid disorders. These results indicate an increase in the incidence of TH in patients with thyroid disorders when compared with the asymptomatic normal population. In our patients who underwent primary thyroid surgery for the treatment of various disorders in the last 4 years, the incidence of TH was 1% (4/405). This finding suggests an increase in the TH incidence among patients with thyroid diseases treated by surgical procedures when compared with both the asymptomatic normal population and patients with medical disorders. The prevalence of TH is up to 5.7% in children with congenital hypothyroidism (1).

Previous reports have shown that in TH, left lobe absence significantly outnumbers right lobe absence. Suzuki *et al.* (4) determined left lobe absence in 55 of 67 (82.1%) TH patients. Previous manuscripts have also reported the incidence of left hemiagenesis to be between 70% and 87.5% of all TH cases (1,8-11). All of our four identified TH patients had left-sided agenesis.

Gender was found to have a significant effect in TH cases with right lobe absence in that 10 of 12 (83.3%) patients were female (4). On the other hand, gender does not appear to affect left hemiagenesis; Suzuki *et al.* (4) have reported that 56.4% (31/55) of patients with left hemiagenesis were female; comparable to our finding of half of the cases of left hemiagenesis (50%; 2/4) being female.

An interesting and important finding of the study of Suzuki *et al.* is the increasing volume of the remaining lobe in cases of hemiagenesis. The volumes of the intact thyroid lobes in subjects with TH were significantly larger than those of ipsilateral lobes in normal subjects (4). This suggests significant compensatory voluminous changes occur in TH. TH without glandular disorders is generally asymptomatic due to production and secretion of thyroid

hormones that is comparable to that of a bilobar thyroid gland. Therefore, enlargement of unilobate gland may be the result of expected compensatory growth aimed at maintaining physiological status and normal serum levels of thyroid hormones. Average thyroid stimulating hormone (TSH) and free triiodothyronine (FT3) serum concentrations were significantly higher in unilobate groups than in bilobate glands. In most cases, a compensatory hypertrophy of the remaining lobe was described that was relative to half the normal total thyroid volume (1).

In their screening study of thyroid structure on a young population, Suzuki *et al.* (4) established 67 cases of TH using ultrasound examination. Noninvasive imaging of the anterior cervical region can successfully identify structural anomalies of the thyroid gland. A combination of ultrasound examination and isotope scintigraphy is sufficient to establish and confirm the diagnosis of TH (1). Ultrasound is the imaging modality of choice to assess structural features of the gland. Beside lobe absence, ultrasound can reveal the structural changes in the remaining lobe, and demonstrate underlying pathology. Nuclear scan is the modality that establishes functional anatomy of the thyroid. This modality can confirm the absence of one lobe detected by ultrasound. Nuclear scan has also established functional status of the remaining lobe. These two imaging modalities are complementary tools to assess structural and functional features of the thyroid and to establish any anatomic abnormality. Many previous reports have also shown that ultrasound is the first tool for evaluation of thyroid anatomy (5,8,12). Previous studies have also emphasized the importance of nuclear scan to assess functional status and to establish functional abnormality of the gland (8,12-14).

Suzuki *et al.* (4) incidentally diagnosed TH during a screening program in an asymptomatic normal population. TH has no specific symptoms and signs leading to its diagnosis. The remaining lobe of the gland generally has normal function. Usually, patients with TH are biochemically euthyroid and clinically asymptomatic (10). The remaining lobe is usually capable of sufficient hormonal synthesis and secretion to sustain clinical euthyroidism. The abnormality is much more likely to be discovered incidentally during the course of imaging examinations of concomitant cervical pathologies. Most of the cases of TH are diagnosed when the patients present a lesion in the functioning lobe. The remaining lobe of the thyroid gland can be a site of pathological changes similar to a normally developed gland and may present with a spectrum of diseases (11). TH is generally established during the clinical workup of symptomatic patients with thyroid disorders. Several thyroid diseases are associated with TH, including: benign or malignant, hyper-, normal-, hypo-functioning disorders, and autoimmune diseases (2,3,8,10,15-18). A higher incidence of associated functional, morphological, and autoimmune thyroid disorders in patients with TH was observed when compared to subjects with bilobate thyroid (10).

The unique lobe of the unilobate gland generally has normal activity and continues to produce and secrete thyroid hormones to sustain euthyroidism. Therefore, TH itself does not require specific treatment. Indications for medical treatment and surgical procedures in TH are not different from those for normal bilobate glands. The remaining lobe may harbor a variety of lesions, and present with disorders indicating surgical intervention (2,3,10,16,17). Thyroid surgery for TH cases involves total excision of the remaining lobe, that is, total excision of thyroidal tissue. Total lobectomy in a bilobate thyroid does not generally require postoperative replacement therapy, because the proper function of the remaining lobe maintains normal serum level of thyroid hormones. In TH, total excision of the remaining lobe is performed. Hemithyroidectomy in TH cases technically becomes a total thyroidectomy with a need for postoperative replacement therapy.

In conclusion, TH is a very rare congenital anatomical anomaly of the thyroid gland with a reported prevalence between 0.02% and 1%. This rate has shown some variation across screening studies performed on asymptomatic populations, imaging studies of patients with thyroid disorders, and preoperative imaging and/or operative findings in patients with surgical thyroidal diseases. TH can be successfully diagnosed using noninvasive imaging methods such as ultrasound and nuclear scan. Left lobe absence is significantly more predominant than right lobe absence. Usually, TH patients without glandular disorders are biochemically euthyroid and clinically asymptomatic, as the remaining lobe is capable of sufficient hormone synthesis and secretion to sustain normal physiology. TH itself does not require specific treatment. However, in some cases, the unique lobe may have medical disorders and harbor surgical diseases. Unilateral exploration and total excision of the remaining tissue achieves definitive treatment of various disorders. Total lobectomy in TH cases technically becomes a total thyroidectomy with a need for postoperative thyroid replacement therapy.

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Footnote

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