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Clinical Record

Thyroid hemiagenesis: clinical significance in the patient with thyroid nodule

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Abstract

Thyroid hemiagenesis is a rare abnormality in which one thyroid lobe fails to develop. Most of the patients diagnosed have an associated thyroid disease. We report the case of a 32-year-old woman who presented with a left thyroid mass which had been gradually increasing in size over a period of two years. A thyroid scan revealed the absence of the right thyroid lobe and isthmus, and a hypoactive nodule was evident in the lower pole of the left lobe. Ultrasonography confirmed the right lobe and the isthmus agenesis. The operative findings confirmed hemiagenesis of the right lobe and nodular hyperplasia in the left lobe.

The purpose of this report is to present a patient with hemiagenesis of the thyroid gland who concomitantly had nodular hyperplasia; we also stress the importance of pre-operative differentiation between benign lesions and malignancy in order to aid critical decision making and to facilitate preservation of thyroid function if possible.

Key words: Thyroid Gland; Abnormalities, Congenital; Diagnostic Imaging

Introduction

Surgeons have recognized that asymmetry of the thyroid lobes is quite common; the right lobe is usually larger and longer than the left lobe. This observation can readily be confirmed by physicians experienced with thyroid scanning.[1] Thyroid hemiagenesis, a developmental failure of a unilateral thyroid lobe, is an uncommon anomaly. The first case was described by Hansfield-Jones in 1866, and approximately 300 cases have been reported in the literature.[2] Because most patients remain unknown until they become symptomatic, the true prevalence of thyroid hemiagenesis is not known; however, it is estimated to be 0.02 per cent in normal children.[3]

This article describes a patient with hemiagenesis of the right thyroid lobe and isthmus who concomitantly had a left thyroid lobe tumour; we also discuss management dilemmas.

Case report

A 32-year-old woman was referred to our hospital for evaluation of a painless, irregular mass in the left anterior neck. This had become noticeably enlarged over the previous two years.

On physical examination, the entire left lobe of the thyroid gland was enlarged. The patient's past medical history did not indicate any other thyroid problems and was essentially noncontributory. There was no history of previous radiation to the neck and no significant family history of thyroid disease. The rest of the physical examination was unremarkable.

The patient was clinically in a euthyroid state, and thyroid function tests were within the normal range. Antithyroglobulin antibody and antimicrosomal antibody were not detected. Thyroid scintigraphy with Tc-99 m pertechnetate revealed the absence of the right thyroid lobe and isthmus and the presence of a hypoactive nodule in the lower pole of the left lobe (Figure 1). Ultrasonography and computed tomography (CT) confirmed agenesis of the right lobe and isthmus and revealed a cystic, approximately $1.9 \times 1.07 \times 2.09$ cm lesion in the inferior pole of the left thyroid lobe (Figure 2). Fine needle aspiration biopsy revealed many colloid-laden macrophages in a colloidal and bloody background.

The patient underwent surgery in order to confirm the diagnosis as well as to treat the thyroid tumour. Intraoperative findings confirmed the presence of a firm nodule in the left lobe of the thyroid and the absence of the right thyroid lobe and isthmus (Figure 3). The left superior and inferior parathyroid glands were normally located. On surgical observation as well as on clinical evaluation, the left thyroid nodule was thought to be a benign tumour. Therefore, a left thyroid nodulectomy was performed, and nodular hyperplasia with cystic degeneration was confirmed on the frozen section and the final pathologic diagnosis.

The patient tolerated the surgery well and recovered with no complications.

Discussion

Failure of development of an entire thyroid lobe is an extremely uncommon anomaly. The discovery rate of this entity by imaging has been reported by Hamburger and Hamburger to be four in 7000 thyroid patients.[1] The true incidence of thyroid hemiagenesis is difficult to determine, since the diagnosis is made in a population being evaluated for some other thyroid pathology. The actual prevalence of this anomaly is probably closer to that recently reported by Shabana *et al.*, who evaluated 2845 school children in a systemic thyroid ultrasound study and found six children with left thyroid lobe agenesis.[3] Their study showed the

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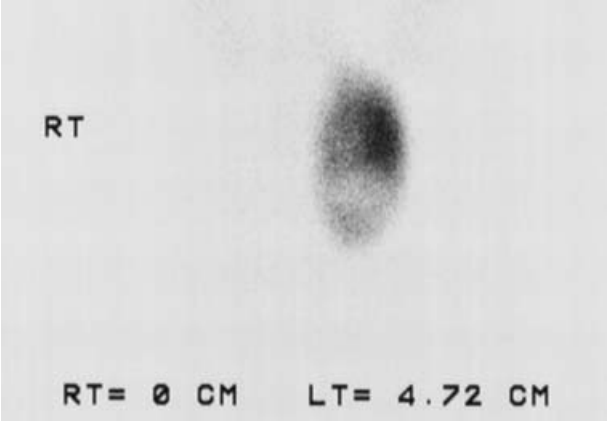

FIG. 1
Thyroid scan showing an absent right lobe and isthmus and a 'cold' nodule involving the lower pole of the left lobe.

estimated prevalence of thyroid hemiagenesis to be 0.02 per cent and confirmed the female predominance and higher incidence of agenesis of the left lobe which have been observed in previous reports.

The thyroid gland develops from a duct-like invagination of the ectoderm in the primitive pharynx between the first and second pharyngeal pouches immediately dorsal to the aortic sac. At 16–17 days of gestation, it continues to expand ventrally, with the most rapid proliferation being in the distal tip, whilst remaining attached to the pharyngeal floor by what is termed the thyroglossal duct. Under the effect of thyroid progenerator, the thyroid rudiment becomes spherical and then bilobed as it grows more caudally, descending in front of the pharynx.[4] It is unclear whether a lobulation defect or a descent disturbance is responsible for hemiagenesis. The absence of compensatory growth of the other lobe is suggestive of a lobulation defect and not of a local problem interfering with descent.

A coexistent thyroid disorder is common in patients diagnosed with thyroid hemiagenesis; such disorders include Graves' disease, chronic lymphocytic thyroiditis, subacute thyroiditis, nodular goitre, adenoma and thyroid carcinoma. Hyperthyroidism has been reported to be the major reason for diagnosis.[1,2,5–14]

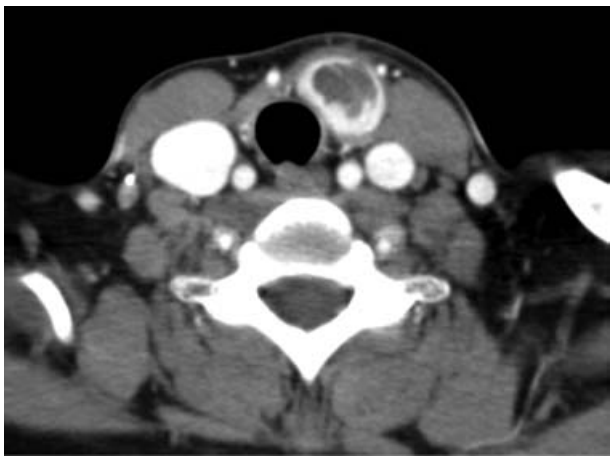

FIG. 2
Pre-operative axial computed tomography scan of the thyroid gland, showing the left thyroid lobe with low-attenuation lesions and an absent right thyroid lobe and isthmus.

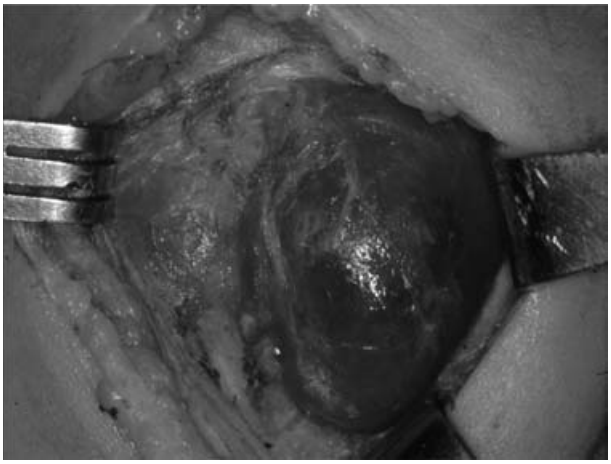

FIG. 3
Intra-operative view of the absent right thyroid lobe and isthmus.

Two physical signs may be of assistance. On the affected side, the edge of the trachea is easily palpable and the edge of the sternomastoid muscle is much closer to the midline and overlies the trachea instead of being separated from it.

Thyroid hemiagenesis is usually diagnosed by thyroid scan via an obvious accumulation of tracer on just one side within the thyroid region. However, there are several clinical conditions mimicking thyroid hemiagenesis on scintigraphic evaluation, and thyroid scans should be confirmed by other methods revealing thyroid morphology. Ultrasound is cost-effective, can be performed easily and does not expose patients to radiation. Magnetic resonance imaging and CT can also visualize thyroid hemiagenesis. As the lobe associated with the underlying disorder is the only thyroid tissue, fine needle aspiration and intra-operative frozen biopsy must be performed, as well as haematologic and radiologic evaluation, in order to rule out malignancy. We emphasize the need for pre-operative differential diagnosis of the underlying thyroid disorder in order to assist critical decision making regarding surgical intervention and preservation of function of the only thyroid lobe.

Our patient displayed a rare case of an absent right thyroid lobe in a 32-year-old woman. Ultrasound and fine needle aspiration cytology were combined with a thyroid scan to enable diagnosis of the hemiagenesis, and frozen biopsy was performed to confirm the nodular hyperplasia. Disease control with preservation of thyroid function was achieved via carefully performed pre-operative tests and intra-operative frozen biopsy.

- **Thyroid hemiagenesis is a rare abnormality in which one thyroid lobe fails to develop. Most patients with this condition have associated thyroid disease**
- **This paper describes the case of a 32-year-old woman presenting with a left thyroid mass which had gradually increased in size over the previous two years. A thyroid isotope scan revealed the absence of the right thyroid lobe and isthmus and the presence of a hypoactive nodule in the lower pole of the left lobe**
- **Several clinical conditions mimic thyroid hemiagenesis on scintigraphic evaluation. Other methods should be used to confirm the diagnosis and reveal the morphology of the thyroid. Ultrasonography is cost-effective, can be performed easily and does not expose patients to radiation**

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