

## THYROID HEMIAGENESIS: CASE REPORT AND REVIEW OF LITERATURE

Mohammed K. Alam, FRCSEd

Thyroid hemiagenesis is believed to be one of the rarest developmental anomalies of the thyroid gland.<sup>1</sup> The thyroid gland begins to develop as a median thickening of endoderm on the floor of the pharynx between the first and second pharyngeal pouches. This area later invaginates to form the median diverticulum, which appears in the latter half of the fourth week. This diverticulum grows caudally as a bifurcating tubular duct, from which the isthmus and the lateral lobes of the thyroid are developed.<sup>2</sup> The cause of unilateral agenesis of the thyroid gland is not known, but it is believed to arise from the failure of the original anlage to become bilobed and spread out laterally to both sides.<sup>1</sup>

The total number of cases of thyroid hemiagenesis reported is uncertain.<sup>1</sup> This report probably presents the first reported case from Saudi Arabia. The aim of the report is to increase readers' awareness of this rare anomaly of the thyroid gland, and thus help to make the preoperative diagnosis possible. A brief review of the literature is also presented.

### Case Report

A 45-year-old Saudi female presented with a gradually increasing swelling on the left side of the neck. She had noticed this painless swelling two months earlier. She complained of occasional palpitation, but no other toxic or pressure symptoms were present. On physical examination, her pulse was 76/minute. She had a left-sided swelling which moved with swallowing, and was nodular on palpation. No swelling was palpable on the right side of the neck, and there were no palpable lymph nodes on either side of the neck. Examination of the vocal chords was normal.

Hematological, biochemical and hormonal assessments were normal. Isotope thyroid scan (Tc99) showed patchy uptake in the left thyroid lobe due to hypoactive areas. There was no uptake on the right side (Figure 1).

Fine-needle aspiration cytology (FNAC) of the left lobe nodules showed aggregates of benign follicular cells with foci of papillary structure. Biopsy was recommended for better evaluation. The neck was explored through a standard thyroid incision. An enlarged left lobe with two well-defined nodules and an absent right lobe were found. Tracheal rings were not covered with any thyroid tissue suggestive of absent isthmus of the thyroid gland. No parathyroid glands were seen in the usual place of the right lobe. The nodular left thyroid lobe was removed, leaving posteriorly a thin rim of thyroid tissue to protect the parathyroids. Histopathological examinations revealed well-capsulated, benign follicular adenomas. The postoperative period was uneventful.

### Discussion

Thyroid agenesis may be complete, unilateral or isthmic.<sup>3</sup> The true incidence of unilateral agenesis is not known. In a study of over 7,000 thyroid scans, unilateral agenesis was found in only four patients.<sup>4</sup> Another study involving 12,456 patients who underwent thyroidectomy reported seven cases of thyroid hemiagenesis.<sup>5</sup> Unilateral agenesis is reported to be more common in the left side, and in females.<sup>6,7</sup> The patient in this report, although a female, had a right-sided hemiagenesis.

Most cases of thyroid hemiagenesis are discovered when patients present with a lesion in a functioning lobe. Clinical examination is of little help in diagnosing hemiagenesis. When the isthmus and the lobe are both absent, tracheal rings may be easily palpable and the edge of sternomastoid on the affected side is nearer the midline overlying the trachea.<sup>1</sup> In the present case, although the isthmus and the right lobe were absent, these physical signs were not appreciated preoperatively. Absence of uptake on the affected side in a thyroid scan may raise suspicion, but usually it may be passed over as a suppressed lobe by an overactive enlarged lobe, or big cold nodule. TSH-stimulated thyroid scan has been used to differentiate between thyroid hemiagenesis and a suppressed lobe. It will show the total absence of uptake in congenital hemiagenesis, but activity will reappear in the suppressed lobe.<sup>4</sup> However, routine use of this investigation is not recommended. Ultrasound examination

---

From the University Unit-Department of Surgery, Riyadh Medical Complex, Riyadh, Saudi Arabia.

Address reprint requests and correspondence to Dr. Alam: University Unit-Department of Surgery, College of Medicine, P.O. Box 7805, Riyadh 11472, Saudi Arabia.

Accepted for publication 05 July 1997. Received 18 March 1997.

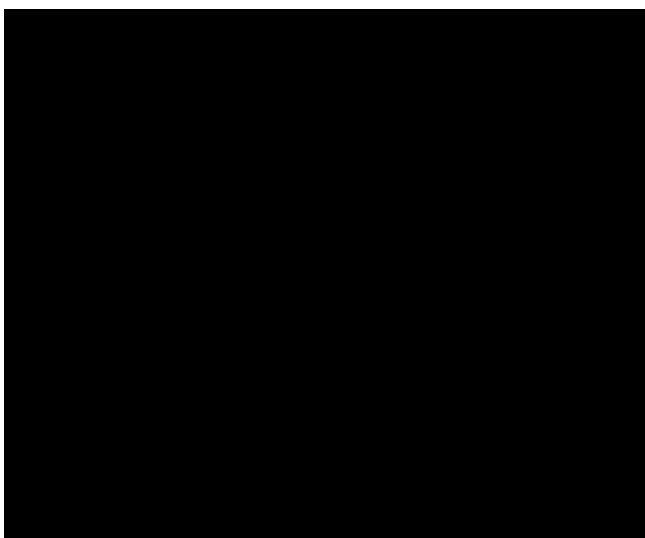


FIGURE 1. Isotope thyroid scan (Tc99) showing uneven uptake in the left thyroid lobe and absent right lobe.

can easily establish the diagnosis in suspected cases.<sup>7</sup> Ultrasound examination is cheaper, easy to perform and there is no radiation risk to the patient.

The functioning lobe in patients with thyroid hemiagenesis could be the site of pathological changes similar to a normally developed thyroid gland. Primary hyperthyroidism, toxic multinodular goiter, hyperfunctioning follicular adenoma, colloid goiter, multinodular goiter, lymphocytic thyroiditis, papillary and follicular carcinoma, and hypothyroidism involving the remaining lobe have been reported.<sup>6-8</sup>

A right aortic arch and unilateral absence of the superior and inferior thyroid artery and the thyrocervical trunk have been reported in association with thyroid hemiagenesis.<sup>9</sup> The superior and inferior thyroid artery were not seen on the right side in this patient during

surgery. Duh et al. have reported hyperparathyroidism due to parathyroid hyperplasia in a patient of thyroid hemiagenesis.<sup>10</sup> Serum calcium was normal and there were no features of hyperparathyroidism in the present case.

Preoperative recognition of thyroid hemiagenesis in patients undergoing thyroidectomy is helpful in avoiding unnecessary exploration of one side of the neck and potential morbidity stemming from this.<sup>7</sup> A degree of suspicion may help in establishing a preoperative diagnosis. Thyroid hemiagenesis should be suspected when isotope thyroid scan shows no uptake on one side, particularly in a euthyroid patient. Ultrasound examination may help in establishing the diagnosis.

### Acknowledgement

I would like to thank Cora Rivera for her secretarial help in typing this manuscript.

### References

1. Greening WP, Sarker SK, Osborne MP. Hemiagenesis of thyroid gland. *Br J Surg* 1980;67:446-8.
2. Warwick R, editor. Williams PL. *Gray's Anatomy*, 36<sup>th</sup> edition. Edinburgh: Churchill Livingstone, 1980;198.
3. Leatherdale BA. An unusual thyroid gland. *Br J Surg* 1973;60:410-3.
4. Hamburger JI, Hamburger SW. Thyroidal hemiagenesis. *Arch Surg* 1970;100:319-20.
5. Harada T, Nishikawa Y, Ito K. Aplasia of one thyroid lobe. *Am J Surg* 1972;124:617-9.
6. Melnick JC, Stemkowski PE. Thyroid hemiagenesis (hockey stick sign): a review of the world literature and report of four cases. *J Clin Endocrinol* 1981;52:247-51.
7. McHenry CR, Walfish PG, Rosen IB, Lawrence AM, Paloyan E. Congenital thyroid hemiagenesis. *Am Surg* 1995;61:634-9.
8. Burman KD, Adler RA, Wartofsky L. Hemiagenesis of thyroid gland. *Am J Med* 1975;58:143-5.
9. Konno N, Kanaya N. Thyroid hemiagenesis. *J Endocrinol Invest* 1988;11:685-7.
10. Duh Q, Ciulla T, Clark OH. Primary parathyroid hyperplasia associated with thyroid hemiagenesis and the agenesis of the isthmus. *Surgery* 1994;115:257-63.