Congenital Right Thyroid Hemiagenesis in a Male: A Case Report

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Abstract

Objective: To appreciate the clinical, intraoperative, and post operative implications of thyroid hemiagenesis.

Background: Thyroid hemiagenesis is rare, affecting from 0.02-0.05% of the population. The left lobe is more commonly absent, with a female predominance. Thyroid disorders have a higher incidence in patients with thyroid hemiagenesis.

Case Report: A 50 year old male presented with a left sided compressive neck mass causing dysphagia. Radiographic imaging demonstrated a large, left substernal goiter and absent right thyroid lobe. Laboratory evaluation showed he was euthyroid, but given his compressive symptoms, the gland was removed confirming multiple adenomatous nodules. Post operatively he now requires thyroid supplementation.

Conclusion: Right-sided thyroid hemiagenesis in a male is extremely rare with initial work up and post operative implications discussed.

Background

• Incidence of thyroid hemiagenesis is between 0.02 and 0.05%.1,2
• First described in 1866 with ~330 cases documented3,4
• Various pathologies of the thyroid gland can be associated with hemiagenesis, which often prompts the initial work up and discovery4

Case Presentation

• 50 year old male referred for a left neck mass.
• The mass was present for several years but in the preceding 6 months had become significantly larger.
• With the increase in size the patient had developed compressive symptoms (dysphagia).
• Physical exam showed a 6 cm left neck mass extending below the clavicle which was mobile with deglutition.
• Mirror laryngoscopy showed mobile vocal folds.
• TSH: 1.370 microiU/mL (normal)
• Free thyroxine (T4): 0.95 ng/dL (normal)
• Triiodothyronine (T3): 117 ng/dL (normal)
• Ultrasound: large complex cystic nodule measuring at least 3.8 cm, extending below the clavicle which was mobile with deglutition.

• Computed tomography: Absent right thyroid lobe, 6.6 cm left thyroid lobe extending to the thoracic inlet (Image 1).
• Intraoperative findings confirmed the absent right thyroid lobe (Image 2).

Discussion

• Failure of proper descent of the thyroid most commonly leads to ectopic thyroid tissue or a thyroglossal duct cyst.5
• Thyroid hemiagenesis is rare. The left lobe is absent 80% of the time with females making up 75% of patients.6
• The isthmus is present in 44% of patients with hemiagenesis.7
• Most patients are asymptomatic with a 0.02-0.05% incidence.1,2 In those with hypothyroidism, the incidence is as high as 5.8%.8
• Evaluation of the remaining lobe has been reported for adenocarcinoma, hyperthyroidism, hypothyroidism, adenomas, multinodular goiters, chronic thyroiditis, and Graves’ ophthalmopathy.9
• TSH is significantly higher in children with thyroid hemiagenesis with associated hypothyroidism2 which can explain the finding of an often compensatory hypertrophy of the remaining lobe.4

Discussion Continued

• Diagnosis can be made by various methods including ultrasound,7 CT,10 or thyroid scintigraphy3 with the latter limited by various conditions which can give the appearance of hemiagenesis including a hyperfunctioning nodule on the contralateral side suppressing normal function11 and unilateral thyroiditis.12
• Several important considerations need to be noted in patients with thyroid hemiagenesis:
• First, to prevent unneeded intraoperative morbidity with dissection of the contralateral gland, thyroid hemiagenesis should be recognized preoperatively, especially if the lobe in question is proven to harbor malignancy.
• Second, the operative note should both make it clear that the contralateral gland appeared absent and the specimen should be labeled as the appropriate side but also noted to be a total thyroid specimen. This can prevent unnecessary future surgery to remove a non existent contralateral side if the patient seeks care elsewhere.
• Finally, patients need to be started on thyroid supplementation post operatively as a lobectomy removes all of the thyroid tissue.

Conclusion

Thyroid hemiagenesis is rare, more commonly occurring on the left and in females, which makes this case quite rare. Important post operative considerations include the need for thyroid replacement following a lobectomy, describing the procedure as a complete thyroidectomy to prevent future morbidity arising from attempts at a contralateral thyroid lobectomy, and the need to initiate thyroid supplementation despite only performing a lobectomy.

References