Recurrence of Graves’ disease in the thyroglossal duct after total thyroidectomy

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SUMMARY
Graves’ disease (GD) due to hyperfunction of thyroglossal duct remnants is rare, but recurrence after total thyroidectomy is even rarer. We present a rare case of a patient with recurrence of GD in a thyroglossal duct, after total thyroidectomy, who has been treated by Sistrunk procedure. Patients with a history of GD and difficult thyroid function control after total thyroidectomy should be studied to rule out persistent and functional thyroid tissue. In these cases, surgical treatment is an effective option.

BACKGROUND
Graves’ disease (GD) is typically characterised by diffuse goitre, hyperthyroidism and extrathyroidal manifestations, such as exophthalmos and pretibial myxoedema. It is an autoimmune disease caused by the anti-thyroid-stimulating hormone (TSH) receptor autoantibodies (TRAbs) that activate this receptor, stimulating the diffuse growth of the thyroid and the production of thyroid hormones.

There are three therapeutic options: anti-thyroid drugs, radioiodine, and surgery. The main goal of treatment is to suppress thyroid function and prevent the recurrence of thyrotoxicosis. The recurrence rate after total thyroidectomy is practically nil, and when present, it is mostly iatrogenic (excessive supplementation), due to the persistence of thyroid tissue on the surgical site, or to the existence of functional ectopic thyroid tissue like the persistence of the thyroglossal duct remnants.

The thyroglossal duct tract results from the descent of the thyroid gland during gestation from the foramen cecum to its final position. The tract usually atrophies, but in some patients, remnants of the tract and thyroid tissue may persist.

There are only a few reports of GD due to hyperfunction of thyroglossal duct remnants, and to the best of our knowledge, only three cases of recurrence after total thyroidectomy have been described by searching in PubMed with the MeSH (Medical Subject Headings) terms “graves disease”; “thyroglossal cyst” and “recurrence”.

We present a rare case of recurrence of GD in a thyroglossal duct, after total thyroidectomy.

CASE PRESENTATION
A woman in her 50s with the diagnosis of GD of over 25 years, and no other relevant medical history, refused surgery and was treated with anti-thyroid drugs.

In 2016, the patient accepted surgical treatment and was referred for general surgery consultation.
INVESTIGATIONS
Neck ultrasonography (figure 3) showed an elongated configuration image, of mixed echogenicity and approximately 18×16 mm. Technetium (99m-Tc) thyroid scintigraphy (figure 4) revealed functioning thyroid tissue superior to the thyroid bed, slightly to the right of the midline. TRAbs were increased by about 10 times the normal limit (17.8 U/L).

TREATMENT
The euthyroid state was achieved under levothyroxine 50 µg/day and the patient underwent an uneventful Sistrunk procedure (figure 5). During the operation, a mass connected to the hyoid bone was identified.

OUTCOME AND FOLLOW-UP
The histological examination of the specimen (figure 6) revealed ectopic thyroid tissue (6 g and 4×2×1.5 cm), in the thyroglossal duct, with hyperplasia and lymphocytic infiltrate, compatible with GD, without evidence of malignant involvement. Three years later, the patient continues to be clinically and biochemically euthyroid at a dose of levothyroxine at 100 µg/day. However, due to persistence of exophthalmos, tocilizumab was initiated with significant improvement.

DISCUSSION
Hyperfunction of thyroglossal duct remnants is rare, and relapse of GD by hyperfunctioning ectopic thyroid tissue derived from the thyroglossal duct after total thyroidectomy is even rarer. Theoretically, traces of thyroid tissue, when present in the thyroglossal duct, are subject to the same effect of TSH and TRAb. This ectopic tissue is potentially susceptible to the same thyroid diseases like tumours and, rarely, hyperfunction.9

In the present case, the patient had no symptoms, signs, or other clinical and imaging findings suggestive of a thyroglossal duct remnant containing functional ectopic thyroid tissue before total thyroidectomy. Furthermore, during surgery, all visible thyroid tissue was removed, including the pyramid lobe and Zuckerkandl tubercles. Otherwise, the persistence of thyroid tissue at the thyroid site could be the cause of GD recurrence. It appears that after total thyroidectomy, the ectopic thyroid tissue from the thyroglossal duct became hyperfunctioning and had grown in size stimulated by the presence of TRAb, requiring successively lower doses of levothyroxine. We believe that if the patient was not treated, she would end up in a hyperthyroid state.

In cases of thyrotoxicosis or difficulty in thyroid function control after total thyroidectomy with adequate therapeutically adherence, functional ectopic thyroid tissue or thyroid remnants must be suspected. Whenever functional thyroglossal duct remnants are present, like in this case, patients may present with a visible or palpable cervical midline mass.
Small interstitial lymphocytic aggregates are also identified. Hyperplasia with dilated follicles accompanied by papillary hyperplasia.

Sizes, with colloid. H&E stain 5× (B) showing a typical pattern of diffuse hyperplasia with dilated follicles accompanied by papillary hyperplasia. Small interstitial lymphocytic aggregates are also identified.

Thyroid tissue present in the thyroglossal duct is potentially susceptible to the same thyroid diseases like tumours and, rarely, hyperfunction.

Patients with a history of Graves’ disease and difficult thyroid function control after total thyroidectomy should be studied to rule out persistent or ectopic functional thyroid tissue.

Surgery is an effective option to treat functional thyroid tissue in the thyroglossal duct.

Learning points

- Thyroid tissue present in the thyroglossal duct is potentially susceptible to the same thyroid diseases like tumours and, rarely, hyperfunction.
- Patients with a history of Graves’ disease and difficult thyroid function control after total thyroidectomy should be studied to rule out persistent or ectopic functional thyroid tissue.
- Surgery is an effective option to treat functional thyroid tissue in the thyroglossal duct.