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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.¹

There are three therapeutic options: anti-thyroid drugs, radioiodine, and surgery.² 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,^{3 4} 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.⁵

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".⁶⁻⁸

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.



Figure 1 Exophthalmos (A,B).

She presented with diffuse toxic goitre and severe exophthalmos (figure 1). Previous ultrasound and radionuclide scintigraphy confirmed the diagnosis and excluded developmental anomalies. The patient underwent a total thyroidectomy, including the removal of the pyramid lobe and Zuckerkandl tubercles. Pathological examination revealed multinodular hyperplasia compatible with GD. The immediate postoperative period was uneventful and she was treated initially with T4 substitution therapy (100 µg/day). At the end of the first year of follow-up, the patient had persistence of exophthalmos and was persistently hyperthyroid necessitating progressive reduction in levothyroxine dose for supplementation. One year later, she presented with a 3 cm non-tender cervical midline mass, mobile during swallowing, and protrusion of the tongue, suggestive of a thyroglossal cyst (figure 2).



Figure 2 Cervical midline mass (arrow).



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Figure 3 Neck ultrasonography, demonstrating a well-circumscribed rounded nodule (arrow) of mixed echogenicity, approximately 18×16 mm.

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.



Figure 4 Technetium (^{99m}Tc) thyroid scintigraphy, showing homogeneous uptake thyroid tissue superior to the thyroid bed, slightly to the right of the midline.

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 therapeutic 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,

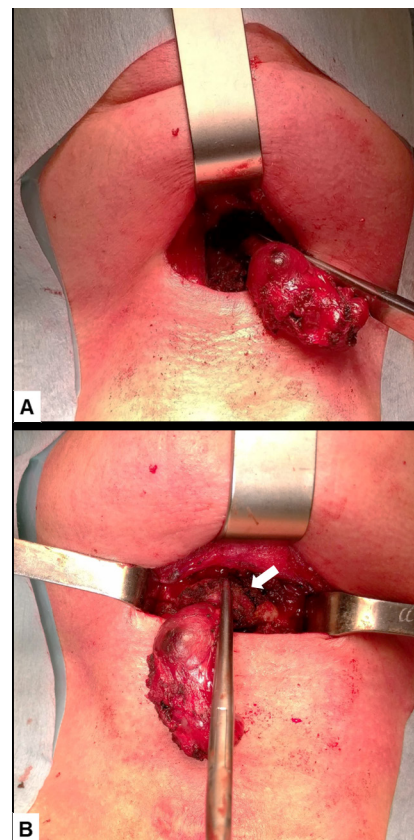


Figure 5 Sistrunk procedure (A,B), during which was observed a mass connected to the hyoid bone (arrow), compatible with thyroglossal duct remnants.

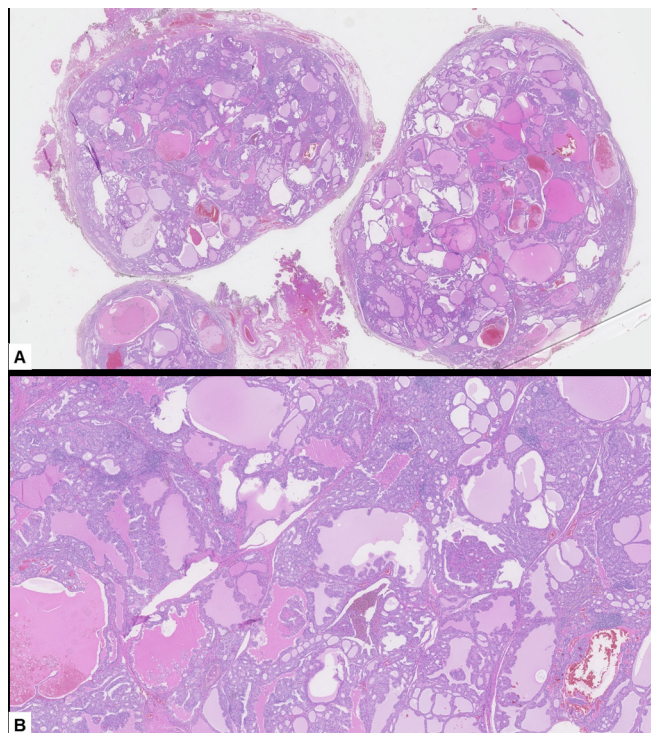


Figure 6 Photomicrographies. H&E stain lower magnification (A) showing thyroid tissue nodules with multiple dilated follicles, of varying sizes, with colloid. H&E stain 5× (B) showing a typical pattern of diffuse hyperplasia with dilated follicles accompanied by papillary hyperplasia. Small interstitial lymphocytic aggregates are also identified.

mobile during swallowing, and protrusion of the tongue. Thyroid scintigraphy with ¹³¹I or technetium is useful for identifying functional thyroid tissue, whereas cervical ultrasound and CT can provide better insight into the location and the relationship with neck structures. Analytically, the patients may present high TRAb levels and an elevated thyroglobulin level, in addition to classic findings of low TSH and high free T4 and T3 concentrations.

Once the diagnosis of functional ectopic thyroid tissue has been established, it is difficult to conclude the best treatment due to the few cases described. Extrapolating from the experience of GD, the same options of treatment are available: anti-thyroid drugs, radioiodine, and surgery. Anti-thyroid agents have the disadvantage of serious adverse effects like agranulocytosis and fulminant liver failure. Radioiodine may worsen the orbitopathy, present in this patient, and surgery

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.

may be the best option for patients with low surgical risk. The fact that the potential for carcinoma is greater than that of hyperfunction⁹ leads us to believe that surgical treatment should be considered the first line, as was proposed to this patient. It is important that pathology findings must discard functional thyroid cancer metastasis. In the present case, the histological examination of the specimen revealed hyperplasia without malignant involvement, compatible with GD.

On patients with a history of GD and difficult thyroid function control after total thyroidectomy, persistent and functional thyroid tissue should be suspected and verified.

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Case reports provide a valuable learning resource for the scientific community and can indicate areas of interest for future research. They should not be used in isolation to guide treatment choices or public health policy.

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