Ectopic lingual thyroid tissue and acquired hypothyroidism: case report

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INTRODUCTION

Ectopic thyroid tissue is the result of abnormal migration of the thyroid gland from the foramen caecum to its final pre-tracheal position. It may be found in the midline anywhere from the base of the tongue to the porta hepatitis [1]. Together with thyroid agenesis and hypoplasia, thyroid ectopy is classified as thyroid dysgenesis. Lingual thyroid was first described by Hickman in 1869 and accounts for 90% of ectopic thyroid tissue [12]. The presence of lingual thyroid tissue does not absolutely preclude the presence of thyroid in its normal location even if this association is extremely rare [3, 4, 21]. The majority of patients with lingual thyroid is asymptomatic but obstructive symptoms, related to mass effect, and bleeding as well as congenital hypothyroidism have been observed [5, 20]. Dysthyroidism in an ectopic thyroid tissue is rare and becomes an exceptionally finding in a patient with both ectopic thyroid tissue and thyroid gland. To our knowledge, only a few cases have been reported in the literature data [3, 4, 11, 21] but an acquired hypothyroidism in a patient with both thyroid ectopy and thyroid gland has not been reported.

CASE REPORT

A 38-year-old female was seen in an outpatient endocrinology clinic on March 2003 because of symptoms
of hypothyroidism. She complained of a one-year history of a progressive weight gain and asthenia. She showed normal growth and development. Personal past medical history was not significant. She was engaged and she had two daughters. There was no history of thyroid disease in her family. She had never taken any medication, which would affect thyroid function. In November 2001 during investigations for anemia, laboratory findings showed an euthyroid state: TSH 2.5 mU/L (0.2-4.2 mU/L) and fT4 12 pg/mL (7-18 pg/mL) without thyroglobulin antibody (TgAb) and thyroid peroxidase antibody (TPOAb) titres. She was evaluated in December 2002 in another Hospital because of asthenia. Laboratory findings revealed a sub-clinical hypothyroidism TSH 8 mU/L, fT4 11.5 pg/mL, fT3 4.1 pg/mL (2.4-4.7 pg/mL) with high titres of TgAb and TPOAb. Neck ultrasonography performed at that time showed “a small thyroid tissue with the characteristic of Hashimoto thyroiditis”. No replacement therapy has been started.

At our evaluation her general condition appeared almost normal. Her height was 168 cm and weight was 71.3 Kg. Blood pressure was 118/72 mmHg and pulsate rate was 54/min and regular. Her skin was slightly dry. Her voice was normal. Thyroid gland was not palpable on physical examination. Laboratory findings confirmed clinical hypothyroidism (TSH 9 mU/L, fT4 6.1 pg/mL, fT3 3.1 pg/mL with TgAb 570 UI/mL (<34) and TPOAb >1.000 UI/mL (<12).

Ultrasonography of the neck revealed a small mass with echogenicity similar to that of thyroid tissue with the characteristic of Hashimoto thyroiditis (fig. 1). A Tc-99m pertechnetate scan yielded focal accumulation at the oropharynx, whereas no uptake was seen at the usual thyroid location (fig. 2). Our diagnosis was acquired hypothyroidism due to Hashimoto’s thyroiditis in lingual thyroid with concomitant non functional cervical thyroid. The patient has been treated with L-tiroxina (50 μg/day) that improved her symptoms and normalized her thyroid tests.

**DISCUSSION**

Lingual thyroid is a rare development anomaly first described by Hickman in 1868 and defined as the presence of thyroid tissue at the base of the tongue [12]. Thyroid primordium begins as a thickening of the epithelium in the pharyngeal floor that later forms a diverticulum and starts its descent caudally towards its final pretracheal position [7, 18]. Functional thyroid differentiation and development require coordinate expression of TSH and its receptor [2, 10], as well as transcription factors TTF-1, TTF-2 and PAX8. Up to now, only rare patients with thyroid dysgenesis have been found to bear mutations in one of these genes [1], so the etiopathogenesis of the majority of the cases of thyroid dysgenesis remains to be identified [15, 17, 22].

Thyroid dysplasia (ectopy, hypoplasia, or aplasia) is a common cause of congenital hypothyroidism. Essentially, all cases of thyroid aplasia and most cases of thyroid ectopy become clinically evident in infancy or childhood [25]. Scintigraphy by Tc-99m pertechnetate can be used
safely in children with hypothyroid disease as in functional orthotopic and ectopic thyroid tissue [13, 16, 23]. Lingual thyroid is a rare entity found in 1/100,000 people with a female preponderance [8]. In the literature, cases with ectopic thyroid gland developing hypothyroidism in adulthood are quite rare [6, 24, 25, 27]. Even in such cases, most of them had features to suggest that hypothyroidism was present from birth [25].

Ectopic thyroid tissue with thyroid gland in its normal location is an extremely rare phenomenon [4, 9, 19]. Dysthyroidism in an ectopic thyroid tissue is rare and become an exceptionally finding in a patient with both ectopic thyroid tissue and thyroid gland. To our knowledge only a few reports have been reported in the literature date. Neinas et al reported fifteen cases of lingual thyroid and only one female patients had both lingual thyroid and thyroid gland. She was not in hypothyroidism and she had an “irritation” at base of tongue and occasionally blood in saliva [21]; GoK et al. reported a 65-year-old white female with both Hashimoto’s thyroiditis that developed from the ectopic thyroid and thyroid gland, but she was not in hypothyroidism [19] and Andrieux S et al a case of a 20-year-old female with hypothyroidism and both ectopic and orthotopic thyroid but without anti-thyroid antibodies [4]. In our report acquired hypothyroidism due to lymphomatous thyroiditis was well-documented and confirmed by laboratory findings: in November 2001 our patient was in euthyroidism with no titres of thyroid peroxidase antibody and thyroglobulin antibodies while in March 2003 laboratory findings showed a clinical hypothyroidism with high titres of anti-thyroid antibodies. Two neck ultrasounds (one of them in our institute) showed “a small thyroid tissue with the characteristic of Hashimoto thyroiditis”. A Tc-99m pertechnetate scan revealed uptake at the oropharynx and no significant uptake in the area of normal thyroid.

Treatment of the lingual thyroid depends on its size, the presence or absence of symptoms, and concomitant factors such as ulceration, bleeding, or malignancy.

Kansal et al suggested that all these patients should have lifelong thyroxine suppression, even those who are asymptomatic and who have an initially small lingual thyroid, as it will prevent its subsequent enlargement, and prevent the onset of hypothyroidism [14]. Patients with obstructive symptoms, suspected malignancy, ulceration, and haemorrhage, should undergo surgical excision of thyroid tissue [11]. In the present case local complications were absent, but there was an hypothyroidism so replacement with thyroxine was the only treatment required. It may be emphasized that in any primary adult hypothyroidism, careful examination also of the base of tongue is necessary even if oropharyngeal symptoms are lacking. We recommend performing a thyroid scan not only when TSH levels are suppressed, but also in all hypothyroid patients, especially when ultrasound investigation shows a small thyroid tissue.

REFERENCES

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