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 hepatis [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 func-
tional orthotopic and ectopic thyroid tissue [13, 16, 23].
Lingual thyroid is a rare entity found in 1/100,000 peo-
ple with a female preponderance [8]. In the literature,
cases with ectopic thyroid gland developing hypothyroid-
ism 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 be-
come an exceptionally finding in a patient with both
ectopic thyroid tissue and thyroid gland. To our know-
ledge only a few reports have been reported in the liter-
ature 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 occa-
Sionally 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 An-
drieux S et al a case of a 20-year-old female with hy-
pothyroidism and both ectopic and orthotopic thyroid
but without anti-thyroid antibodies [4]. In our report ac-
quired hypothyroidism due to lymphomatous thyroiditis
was well-documented and confirmed by laboratory find-
ings: in November 2001 our patient was in euthyroidism
with no titres of thyroid peroxidase antibody and thyro-
globulin antibodies while in March 2003 laboratory find-
ings 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-99
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 lin-
gual thyroid, as it will prevent its subsequent enlarge-
ment, and prevent the onset of hypothyroidism [14].
Patients with obstructive symptoms, suspected mali-
gnancy, 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 orophar-
yngeal symptoms are lacking. We recommend per-
forming a thyroid scan not only when TSH levels are
suppressed, but also in all hypothyroid patients, espe-
cially when ultrasound investigation shows a small thy-
roid tissue.

REFERENCES

1. Abdallah-Matta M, Dubarry P, Peey J, Caron P. Lingual thyroid
and hyperthyroidism: a new case and review of the literature.
2. Abramowiz J, Duprez L, Parma J, Vassart G, Heinrichs C. Fami-
liar congenital hypothyroidism due to inactivating mutation of
the thyrotropin receptor causing profound hypoplasia of the
3. Aktolun C, Demir H, Berk F, Kir M. Diagnosis of complete ecto-
cpic lingual thyroid with Tc-99m pertechnetate scintigraphy.
of three cases. Thyroid 1998 ; 8 : 1055-1057.
6. Borgoni F, Liberatori E, Giambagli M. Lingual thyroid and
hypothyroidism: report of a case in an middle aged woman.
7. De Felice M, Ovitt C, Biffali E. A mouse model for hereditary
thyroid dysgenesis and cleft palate. Nat Gene 1998 ; 19 : 395-
398.
search for the possible molecular mechanisms of thyroid dys-
genesis: sex ratios and associated malformations. J Clin Endo-
ocrinol Metab 1999 ; 84 : 2502-2506.
9. Feller UK, Mavros A, Gaertner HJ. Ectopic submandibular thy-
roid tissue with a coexisting active and normally located thy-
10. Gok U, Keles E, Cobanoglu B, Yildiz M, Donder E. Ectopic thy-
roid and Hashimoto’s thyroiditis arising from a thyroglossal
duct cyst: a case report. Kulak Burun Bogaz Ihtis Der 2003 ;
10 : 29-32.
11. Hickman W. Congenital tumor of the tongue pressing down
the epiglottis on the larynx and causing death by suffocation
sixteen hours after birth. Trans Path Soc London 1869 ; 20 :
160.
thyroid glands in euthyroid children. Pediatrics 1996 ; 38 :
647.
14. Kansal P, Macchia E, Chiovato L. Mutations in the gene encoding
thyroid transcription factor-1 (TTF-1) are not a frequent cause
of congenital hypothyroidism with thyroid dysgenesis. Thyroid
16. Lecklntner ML. Neonatal and pediatric thyroid imaging. In: Saan-
der MP, Patton JA, Partain CL, eds. Thyroid and Parathy-
roid Imaging. Norwalk, CT: Appleton-Century-Croft, 1986 ;
pp 159-162.


