ECTOPIC LINGUAL THYROID PRESENTING WITH MASSIVE HEMATEMESIS

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Abstract

Ectopic lingual thyroid is a rare developmental abnormality caused by aberrant embryogenesis during thyroid migration. Even though, most patients are asymptomatic, uncommonly the mass can be enlarged and cause dysphagia, dyspnea, upper airway obstruction, dysphonia, hypothyroidism. We report a very rare case of ectopic lingual thyroid presenting with massive hematemesis.

Key words: ectopic, lingual thyroid, massive hematemesis.

INTRODUCTION

Ectopic thyroid tissue is a rare embryological anomaly which appears during the migration of the thyroid anlage from the primitive foregut to its final position in the pre-tracheal region of the neck. According to autopsy series the prevalence of ectopic thyroid tissue varies between 7% and 10%. It can be found in the midline, laterally in the neck, mediastinum, or even under the diaphragm and other sites such as axillary, pituitary and adrenal gland, genitourinary and gastrointestinal tract. Approximately 90% of ectopic thyroid tissue is found in the lingual thyroid area that is usually localized in the posterior third of the base of tongue (1). The pathogenesis of lingual thyroid caused by migration defect is not fully known but maternal immunoglobulins leading to block TSH induced thyroid growth may play a role (2). Ectopic lingual thyroid is an incidence of 1 in 3000 of the thyroid cases with prevalence of 1 in 100,000. In 75 % of cases lingual thyroid is associated with an absence of the normal cervical thyroid and the unique source of thyroid hormone production. Lingual thyroid is most diagnosed in the first three decades of life and has a predominance of female as in other thyroid diseases (3). Even though, most patients are asymptomatic, infrequently the mass can be enlarged and cause several symptoms such as cough, dyspnea, dysphonia, dysphagia and upper airway

obstruction, hypothyroidism. However, bleeding is a very rare manifestation which may cause life-threatening massive hemorrhage (4).

CASE REPORT

A 33-year-old man was referred to emergency department with complaint of coffee ground emesis for two days. On admission his blood pressure was 110/70 mm Hg with a pulse rate of 96 bpm. Direct physical examination of oral and nasal mucosae failed to reveal a bleeding focus. Examinations of chest and abdomen were within normal limits. Patient was informed about the admission rules and procedures and signed the informed consent, according to the Ethics Committee regulations. Upper gastrointestinal endoscopy examination revealed fresh blood in the stomach, without any evidence of mucosal abnormalities in the esophagus and stomach. His serum hemoglobin and hematocrit levels were 7 g/dL and 25% respectively. Two units of erythrocyte suspension were transfused. Four hours later, hematemesis stopped spontaneously. On laryngoscopic examination, a smooth mass obstructing the visualization of the larynx with reddish surface covered with vessels and bleeding focus was found at the base of the tongue that was considered as ectopic lingual thyroid (Fig. 1). Thyroid ultrasonography revealed no orthotropic thyroid gland in normal region. Thyroid scintigraphy with technetium-99m (Tc-99m) showed an increased focal uptake in the tongue base consistent with lingual thyroid without any Tc-99 m uptake in the normal thyroid location (Fig. 2). Magnetic resonance imaging (MRI) exam revealed a mass, 40 x 36 x 51 mm in size with no invasive features indicated a lingual thyroid without a thyroid gland in the normal location (Fig. 3). Thyroid hormone tests showed elevated TSH: 39 mIU/L (reference value 0.3–5.5 mIU/L), low FT3: 0.5 ng/dL (reference value 0.7–1.8 ng/dL) and normal FT4: 1.52 ng/dL (reference value 0.89–1.78 ng/dL).

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Figure 1. A. Videolaryngoscope (VLS) imaging of oropharynx showing a smooth lingual thyroid mass at the base of tongue with a bleeding focus on it (Dashed arrow), *: Uvula. B. Flexible fiberoptic nasopharyngoscope (FFN) imaging of lingual thyroid mass with intense vascularization (Arrow). C. Lingual thyroid mass (**) pushing epiglottis (*) and obstructing air passage on FFN imaging.

Figure 2. Tc-99m thyroid scan showing evidence of lingual thyroid (arrows) without any thyroid tissue at its normal location in the neck.

Figure 3. Magnetic resonance imaging showing thyroid tissue at the base of tongue (arrows).

He was diagnosed as a case of lingual thyroid with hypothyroidism and thyroid hormone replacement therapy (levothyroxine 1.6 microgram/

kg/day) was started and surgical removal was recommended due to the risk of re-bleeding but our patient refused to get operated. Six weeks later TSH was found to be 8 mIU/L with normal FT3 and FT4 levels. Although he was warned about the risks of nonadherence to therapy, the patient was lost to follow-up for one year. He was called by the physician and invited for a control visit. In the latest examination he reported recurrent hemoptysis episodes and dyspnea. Laboratory examination revealed persistent overt hypothyroidism (TSH 40 mIU/L and FT4 0.5ng/dL) due to noncompliance with treatment. There was no shrinkage in the lingual thyroid mass on repeat MRI imaging. He was persuaded for trans-oral robotic surgery after euthyroid state was achieved due to the risk of re-bleeding.

DISCUSSION

Lingual region is the most common site of the ectopic thyroid lesions. Lingual thyroid is often asymptomatic, but increased levels of thyroid stimulating hormone (TSH) for physiologic demands can lead to symptoms due to hypertrophy of ectopic lingual thyroid. The lingual thyroid tissues may exist as a painless, non-tender, reddish mass with a smooth or irregular surface at the base of the tongue with or without prominent vessels. The symptom severity may depend on the size of the ectopic lingual tissue and may be presented as the sensation of a foreign body, cough, dysphagia, dysphonia, and dyspnea or bleeding. In the literature only a few cases leading to life-threatening massive hemorrhage have been described so far of which the hemorrhage was attributed due to their prominent large blood vessels on its surface as in our case (4).

For a lingual thyroid case, initial laboratory studies should include thyroid function tests to find out whether the gland is functional or not. A great part of cases of lingual thyroid without a co-existing eutopic thyroid gland will develop subclinical hypothyroidism that may become clinically obvious during periods of physiological stress, puberty, pregnancy especially in iodine deficient regions or lithium therapy. Hyperthyroidism arising from lingual thyroid is less common than hypothyroidism. It is thought that thyroid-stimulating immunoglobulins have a role in etiology (1).

Various diagnostic modalities can be used to confirm the diagnosis, such as thyroid scintigraphy, computed tomography (CT) scan and MRI scan. Scintigraphy should be planned in cases of non-visualized normal thyroid gland on US. Additionally, hybrid imaging systems such as single-photon emission computed tomography imaging using either iodine-123 or Tc-99 m pertechnetate scan can provide both anatomical and functional information (5).

Lingual thyroid is frequently benign therefore histologic confirmation is usually not necessary, but rarely can harbor malignancy. Incidence of carcinoma in lingual thyroid is estimated to be approximately 1 in 100 cases that majority of these tumors are follicular type not papillary in contrast to typical thyroid tumors (6). In our case, we did not performed biopsy owing to its non-invasive nature on MRI imaging and risk of re-bleeding.

The management of treatment of a lingual thyroid depends on the size, symptoms, and the presence of complications. Asymptomatic and euthyroid patients do not usually need the treatment, and just follow-up is enough. In order to suppress TSH level to correct hypothyroidism, reduce gland size and local symptoms, levothyroxine replacement therapy should be given. Surgery is considered when the mass produces obstructive symptoms, ulcerations, malignancy or repeated severe bleeding. Unless emergency surgery is needed, thyroid hormone therapy should be given before surgery to decrease the size of the gland (7). While trans-oral route is more suitable for small lesions, the external approach is more appropriate for bigger lesions. However, scarring and fistula formation on the neck are the undesirable complication of external approach. Trans-oral robotic surgery (TORS) is the latest innovative application (8). If the lingual thyroid is the sole functional thyroid tissue in the body, transposition or autotransplantation may be considered (9). The embolization of the main feeding artery before

surgery of massive hemorrhagic lingual thyroid may reduce the amount of bleeding. Thyroid ablation with radioactive iodine is an alternative to surgery for unfit patients or those who refuse surgical intervention. However, ablative radioactive iodine should be avoided in children and young adults, because of the potential side effects on the gonads (7).

In conclusion, lingual thyroid presenting with massive hematemesis is a rare situation. Bleeding ectopic lingual thyroid could be considered after excluding common causes of acute gastrointestinal bleeding by gastroendoscopic examination. If the thyroid gland is not in its normal location, ectopic thyroid should be considered and should be further investigated. Most lingual thyroids are asymptomatic and no therapy is necessary. For the patients with mild symptoms and elevated TSH concentration, thyroid hormone replacement therapy is usually sufficient. Surgery is recommended in serious cases.

Conflict of interest

The authors declare that they have no conflict of interest.

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