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Case Report

Ectopic thyroid - two case reports

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Abstract:

Bachground: Ectopic thyroid is a rare entity resulting from maldescent of the thyroid gland & can be found anywhere between the foramen cecum & the normal pretracheal position of the thyroid gland, as well as in distant places such as the mediastinum and the subdiaphragmatic organs. Although most cases are asymptomatic, obstructive symptoms may appear. Any disease affecting the thyroid gland may also involve the ectopic thyroid, including malignancy. Hypothyroidism is frequent but hyperthyroidism is an exceptionally rare finding. The clinician must distinguish between ectopic thyroid and metastatic deposits emerging from an orthotopic gland, as well as other benign or malignant masses. Thyroid scintigraphy plays the most important role in diagnosing ectopy, but ultrasonography contributes as well. Treatment is indicated in the presence of symptoms & consists initially of full replacement thyroid hormone therapy. Severe or unresponsive cases require excision.

Case reports: We present here two case reports & review of the literature.

Conclusion: This review provides current understanding about the wide clinical spectrum of this rare condition, optimal diagnostic approach, differential diagnosis, and management strategies.

Key words: *Thyroid; ectopic*

Introduction:

Ectopic thyroid is a rare developmental abnormality resulting from aberrant embryogenesis of the thyroid gland during its passage from the floor of the primitive foregut to its final pre-tracheal position[1]. Lingual site is the most common cause >90% of ectopic thyroid. Extralingual thyroid tissue is commonly located in the anterior cervical area along the path of the thyroglossal duct[2]. Other rare locations of ectopic thyroid are the submandibular region, trachea, esophagus, mediastinum, heart, lung, abdomen etc[3].

Its prevalence is about 1 per 100 000–300 000 people, rising to 1 per 4000–8000 patients with thyroid disease[1]. However, in autopsy studies, the prevalence ranges from 7 to 10%[4]. More than 440 cases have been reported to date. In 70–90% of cases, it is the only thyroid tissue present[1,4,5]. Ectopic thyroid is most common in females, especially in populations of Asian origin[5]. It

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may occur at any age, from 5 months to 40 years, but it is most common at younger ages.

Case report 1:

A 15-year-old female presented with the history of not gaining height since her earlier operation in the neck 8 years back in 2003 for thyroglossal duct cyst. Upon referral to the ENT clinic she was seen in the ENT department of SSMC, Dhaka in May 2011. The patient gives the history of slow thought & coarse dry skin but has normal intelligence & menstruation. Oropharyngeal examination showed a spherical mass at the back of the tongue but covered with intact mucosa measuring 3-2-2 cm. Thyroid hormone study reveals elevated TSH 43.54 mIU/L but with normal serum T_3 4 pml/L & T_4 9.3 pml/L level suggestive of hypothyroidism. Thyroid ultrasonogram report shows no thyroid tissue in thyroid bed, Thyroid spect scan by Tc99 shows thyroid gland not visualized in thyroid area. There is a focal area of radiotracer concentration in the oral cavity-suggesting thyroid tissue in ectopic position at the tongue base. she was started on thyroid hormone replacement therapy, levothyroxine, 50 ug daily & hypothyroidism including the size of ectopic thyroid is reducing in size.

Case report 2:

A 13-year-old male child presented to the ENT outdoor with a painless mass in the anterior neck that had been present for at least 8 years & is gradually increasing in size. Upon referral to the otolaryngology clinic the patient was found to have a smooth, soft to firm, mobile, 3 cm mass in the anterior neck above the hyoid region. This mass elevated with swallowing but movement on tongue protrusion was difficult to evaluate. A presumptive diagnosis of thyroglossal duct cyst (TGDC) was made. Fine needle aspiration biopsy of the neck

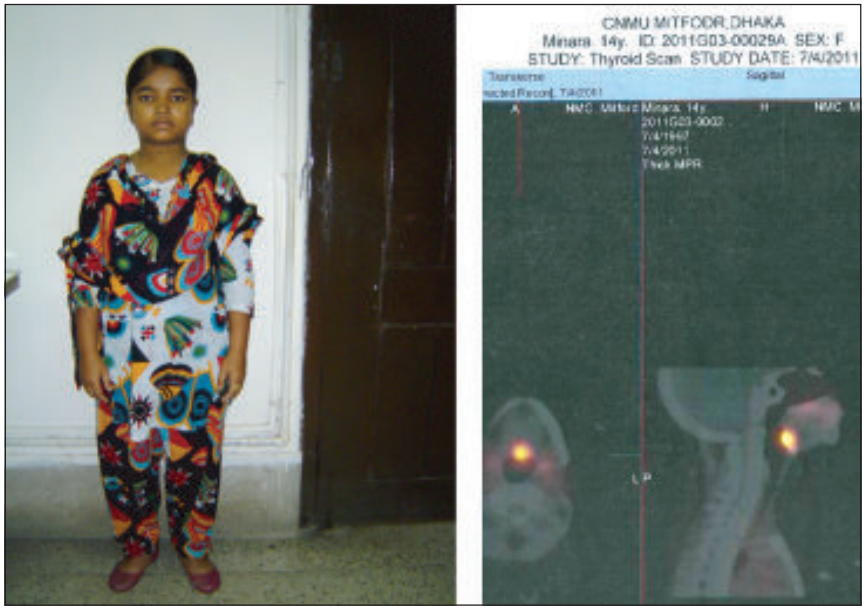

Figure 1: *First case with Spect CT (a & b)*

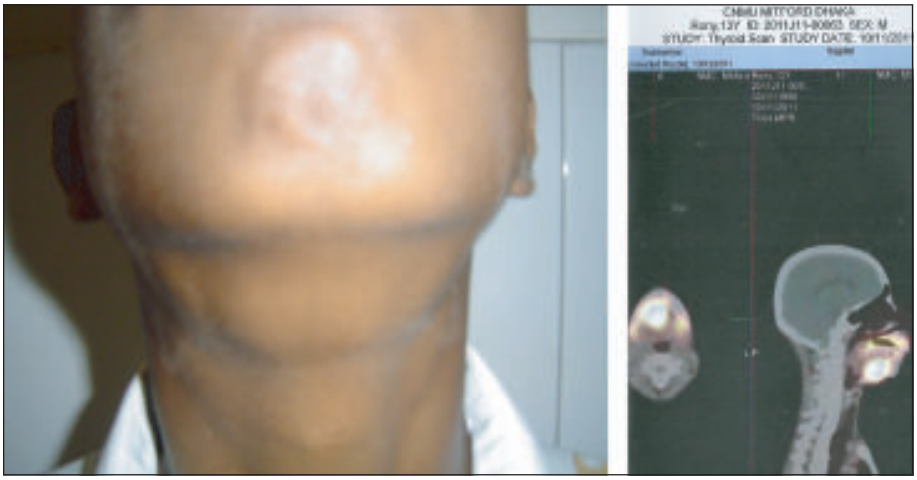

Figure 2: *Second case with Spect CT (a & b)*

mass was performed in Rajshahi & was consistent with TGDC and negative for malignancy. He was otherwise completely well and has normal intelligence. Ultrasonogram reveals no thyroid tissue in thyroid position. Radioisotope scan shows a focal increased radiotracer concentration in the oral cavity but no uptake in the thyroid bed suggestive of ectopic thyroid tissue in sublingual region & the midline neck mass was in fact the only functioning thyroid tissue.

Spect CT shows increased radiotracer concentration in submental region corresponds to the area in between the mandible and hyoid bone a bit right from the midline below the level of the tongue. Thyroid hormone study reveals elevated TSH 28.98 mIU/L but with normal serum T_3 2.34pml/L & T_4 57.54pml/L level suggestive of hypothyroidism. He was started on thyroid hormone replacement therapy, levothyroxine, 50 ug daily & the size of the ectopic thyroid is reducing in size.

Discussion:

A larger study of 49 cases of ectopic thyroid in Korea found lingual thyroid in 23 patients, sublingual thyroid in 17 patients, combined type in 7 patients and an intratracheal thyroid in one patient. Only four cases had thyroid in orthotopic position, and 62% demonstrated hypothyroidism[5]. We report two patients, one with lingual and another is sublingual and both of them showed absent thyroid in normal thyroid bed & demonstrated hypothyroidism. We found lingual thyroid in female and sublingual thyroid in male.

Clinical presentation of ectopic thyroid tissue:

Lingual thyroid:

Lingual thyroid is the most frequent cause of ectopic thyroid, accounting for about 90% of the reported cases[6]. In 70-75% of cases, lingual thyroid is the only thyroid tissue present as it is found in our case. Our patient gives the history of excision of thyroglossal cyst 8 years back & now she presented with lingual thyroid along with

symptoms of hypothyroidism in the absence of orthotopic thyroid gland. Related papers of thyroglossal cyst excision is not avialabe. Treatment of thyroglossal cyst involves Sistrunk's operation & include excision of the body of the hyoid and en block dissection in the line of descent of the throid gland as far as the foramen caecum[7]. Probably the tract was not excised as far as the foramen cecum in our patient. In terms of thyroid function, most patients with lingual thyroid present with hypothyroidism, Hyperthyroidism is extremely rare.

Normal glands hypertrophy during puberty, menstruation and pregnancy. Relative deficiency of thyroid hormones with these events will also cause the ectopic thyroid to enlarge and become symptomatic. Onset of symptoms of lingual thyroid is often associated with these events[8]. This situation is more common in women (seven times)[9]. We found it in female patient as is found in other study in our country[10].

The diagnosis is usually made by the incidental discovery of a mass on the back of the tongue in an asymptomatic patient. The mass may enlarge and cause a mild sensation of fullness in the throat to dysphagia, dysphonia, dyspnea, or a sensation of choking, cough, snoring, sleep apnea[11], and, in more severe cases, respiratory obstruction and hemorrhage[12]. In our patient there is no obstructive symptoms.

On examination a mass is seen in the base of the tongue, often visible without the aid of a laryngeal mirror. Its characteristics are very variable in size, shape, color, consistency, vascularity and mucosal continuity, and are of minor importance in making a diagnosis. Physical examination is usually sufficient for diagnosis, although CT or nuclear imaging may provide additional information.

Extralingual ectopic thyroid tissue:

Extralingual ectopic thyroid most commonly located in a midline position above or below the hyoid bone, the region of the thyroglossal duct cyst. We report a case found above the hyoid bone (Sublingual). It must be differentiated from thyroglossal duct cyst, because it is usually located in the same anatomic position[13] & frequently represents the only source of thyroid tissue, removal of which has definite physiologic as well as possible medicolegal implications. To prevent total thyroid ablation, a thyroid scan or ultrasound examination must be performed in all cases of thyroglossal duct cyst before its removal so as to be certain that a normal thyroid gland is present. Our patient was initially diagnosed as thyroglossal duct cyst on FNAC but on USG, Thyroid scan & Spect scan it was proved to be ectopic thyroid.

Hypothyroidism is commonly present as in our case because of the absence of a normal thyroid gland in most instances. An enlarging mass commonly occurs during infancy, childhood or later life. Our patient presented at 13 years of age. Extralingual ectopic thyroid tissue can also undergo the same pathological changes as the ectopic thyroid gland, and can be benign or malignant.

Ectopic thyroid tissue and its clinical presentation[14].

Location	Symptomatology	Age at diagnosis	Thyroidal status
Lingual	Dysphagia, dysphonia, stomatolalia, cough, sensation of foreign body, snoring and sleep apnea, respiratory obstruction, and hemorrhage Asymptomatic	0–83	Usually hypothyroid, less often euthyroid
Submandibular	Palpable, mobile, usually painless mass	4–81	Euthyroid
Thyroglossal duct cyst	Asymptomatic	4–75	Hypothyroid or euthyroid
Intratracheal	Dyspnea, cough, difficulty in swallowing, hemoptysis, and stridor Asymptomatic	30–50	Euthyroid
lung, and Intra-thoracic (med-iastinal, heart)	Dry cough, dyspnea, and hemoptysis	40–77	Euthyroid
	Asymptomatic		
Struma ovarii	Usually asymptomatic	45	Euthyroid, hyperthyroid in 5–15%
	Lower abdominal pain, palpable lower abdominal mass, and abnormal vaginal bleeding		
Adrenal glands	Asymptomatic (incidental finding)	50–67	Euthyroid
Duodenum, pancreas and intestine	Asymptomatic (incidental finding)	50–63	Euthyroid
	Rarely abdominal pain		
Dual ectopy (usually lingual and subhyoid)	Midline neck swelling or asymptomatic	4–45	Euthyroid or hypothyroid

Investigations:

Scintigraphy, using Tc-99m, I-131, or I-123, is the most important diagnostic tool to detect ectopic thyroid tissue and shows the absence or presence of thyroid in its normal location. Thyroid scan can also unmask additional sites of thyroid tissue. Color Doppler US, computed tomography (CT), and magnetic resonance imaging (MRI), may help in designating the extension and location of ectopic tissue, thus contributing to a better pre-surgical evaluation of these cases [4,11,15].

Thyroid hormone tests may reveal the thyroid function status.

CT and MRI are also useful modalities in cases when radioiodine uptake by normal thyroid gland masks the uptake of the ectopic thyroid tissue, especially in the midline.

Fine needle aspiration cytology (FNAC) helps in confirming the diagnosis of ectopic thyroid. It is the only modality to differentiate between a benign and a malignant lesion[4].

Differential diagnosis:

Ectopic thyroid tissue can pose difficult diagnostic and management problem. Ectopic thyroid tissue should be considered in the diagnosis of a cervical mass even in the presence of a eutopic thyroid gland. The clinician should always take into account the potential of this rare entity and differentiate it from other masses in the neck and distant sites.

In general, differential diagnosis depends on the location. Lingual and submandibular thyroid must be differentiated from adenomas and cysts in the midline, including angiomas, fibromas, lymphangiomas, lipomas, salivary gland tumors, thyroglossal duct cysts, midline branchial cysts, and epidermal or sebaceous cysts[11,15]. Thyroid cancer metastases should always be excluded, as these can manifest as an ectopic thyroid tissue[6]. Lingual thyroid is an important cause of a mass on the posterior third of the tongue & should be differentiated from other swellings at the base of the tongue, such as hypertropic lingual tonsil, vallecular cyst, and mucous retention cyst[15].

Management:

There is no consensus about the management due to the rarity of this clinical entity. Most authors agree that surgical treatment of ectopic thyroid in the neck (mainly lingual, sublingual, submandibular, and lateral cervical) depends on size and local symptoms (airway obstruction, dysphagia, and dysphonia), as well as on other parameters, such as patient's age, functional thyroid status, and complications of the mass (ulceration, bleeding, cystic degeneration, or malignancy)4,11,12,15. The usual treatment of ectopic thyroid is thyroid hormone therapy to suppress the ectopic thyroid and reduce its size. Only rarely is surgical excision necessary. Indications for extirpation include failure of suppression therapy to reduce its size, ulceration, hemorrhage and suspicion of malignancy.

Medical treatment:

For mild symptoms and hypothyroid states, levothyroxine replacement therapy may be effective, leading to considerable mass reduction[4,11,12,15]. Ablation with radioactive iodine has been recommended. Its long-term carcinogenic effects & potential deleterious effect on the gonads and other organs[11] render it unsuitable for use with these predominantly young to middle-aged patients.

Surgical treatment:

Patients with severe symptoms, or who do not respond to medical therapy will require complete excision of the lesion. Inability to exclude carcinoma, for example a sudden or recent enlargement with no obvious precipitating factor, is an absolute indication.

Several surgical approaches for lingual thyroid have been described, such as the transoral route and the transhyoid, suprahyoid, or lateral pharyngotomy. Trans-oral approach has been most popular in the past[16]. Dissection of an extensive lesion may still be difficult. Lateral pharyngotomy provides the most satisfactory approach in terms of exposure and dissection. The former is usually preferred for small lesions since it does not affect deeper structures; thus complications are avoided. The latter approach with or without preoperative tracheotomy is chosen for larger masses providing better control of bleeding[11,16].

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