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Ectopic Thyroid Tissue – A Case Series

ABSTRACT

Objective: To describe the clinical presentation and course of seven ectopic thyroid patients.

Methods:

Design: Retrospective Chart Review

Setting: Tertiary Government Teaching Hospital

Patient: Seven patients

Results: Five patients were female and two were male (ratio of 5:2). Three belonged to the 20 to 30 year-old age group, whereas two were below 10 years of age. All seven were biochemically hypothyroid and ectopic thyroid was found to be the only functioning thyroid tissue. Three patients were managed medically with levothyroxine, while ectopic thyroid was excised in four. Ectopic thyroid tissue was autotransplanted in two cases following excision.

Conclusion: The ages of presentation in the present series correspond with the increased physiological demand of thyroid hormone. Thyroid substitution therapy is a must in the presence of clinical and/or biochemical hypothyroidism. Surgical excision should be avoided as far as possible especially if the ectopic tissue is the only functioning thyroid in the body. Surgery is required in selected cases presenting with obstructive symptoms or hemorrhage which are unresponsive to substitution therapy. Auto transplantation of the ectopic thyroid may not provide significant benefit to the patient and more research is warranted in this aspect.

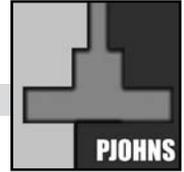
Keywords: *ectopic thyroid, lingual thyroid, thyroid replacement therapy, ectopic thyroid surgery*

When thyroid tissue is not in its normal anatomical location, it is called an ectopic thyroid. Ectopic thyroid tissue can be found anywhere between the *foramen caecum* and the normal position of the thyroid gland, and may be the sole functioning thyroid tissue of the body. It is most frequently found in the region of the *foramen caecum* in patients where the gland fails to descend. Extralingual thyroid tissue is most commonly located in the anterior cervical area in the region of thyroglossal duct.¹ Sometimes, ectopic thyroid may be found in unusual anatomic locations like the heart, gall bladder and in the trachea.^{2,3,4} We describe the clinical presentation, location, functional status, course and management of seven patients with ectopic thyroid.

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METHODS

A retrospective chart review was conducted in a tertiary care institution, Medical College Hospital, Kolkata after obtaining institutional ethical board clearance. All patients who were clinically suspected to have an ectopic thyroid tissue and were subjected to radiotracer scanning along with conventional radiological and biochemical investigations between January 1999 and December 2007 were included in our search protocol. Among these cases, those patients where documentary proof of ectopic thyroid was present were included in the study. Patients having any malignant change in the ectopic tissue demonstrated by fine-needle aspiration cytology (FNAC) were excluded from the study.

Demographic data, clinical presentation and location of ectopic thyroid, thyroid function status (Free T3, Free T4 and TSH) and results of Iodine ¹³¹ thyroid scans and CT scans were recorded. Medical and surgical treatments as well as follow ups were also obtained and analyzed.

RESULTS

Of the 13 patients suspected to have ectopic thyroid within the study period, five patients were excluded (one with tongue base leiomyoma, two with secondarily-infected vallecular cysts, two with cervical lymphadenitis masquerading as ectopic thyroid tissue). Eight patients were confirmed to have ectopic thyroid, but one of the eight had follicular neoplasm reported by FNAC and was further excluded from the study. Thus a total of seven patients were included in the review, of whom five patients were female and two were male (ratio of 5:2). Three belonged to the 20- to 30-year-old age group, whereas two were below 10 years of age. (Table 1)

Four of these seven patients presented with lingual masses and three had midline neck masses. All four patients presenting with lingual thyroid complained of dysphagia. (Figure 1) In addition, two of these patients also had bleeding from the mouth and one had respiratory difficulty. The patients with midline neck masses usually complained of a cosmetic problem. (Figure 2) Biochemically, all patients were hypothyroid and thyroid scans showed ectopic thyroid as the only functioning thyroid tissue in each patient. Midline-neck ectopic thyroids were diagnosed by FNAC, whereas lingual thyroids were diagnosed by clinical and radiological examination. Thyroid scintigraphy was performed in all cases. (Figure 3) CT scans were only performed in the cases of lingual thyroid. Three out of four lingual thyroids were excised because of obstructive symptoms. Of these, one had auto-transplantation of excised thyroid tissue. Only one patient with midline-neck ectopic thyroid was surgically treated and received auto-transplantation. Younger patients with little or no compressive

or other symptoms were treated with thyroxin suppression therapy only. Regardless of treatment modality all patients received life-long levothyroxine replacement therapy.

Table 1.

| No | Sex | Age | Presentation/ Symptoms | Site | Treatment |
|----|-----|-----|--|----------------|---|
| 1 | F | 7 | Midline neck swelling | Suprahyoid | Thyroid substitution therapy |
| 2 | F | 22 | Midline neck swelling; Pain | Subhyoid | Excision & auto-transplantation |
| 3 | M | 18 | Lingual mass, Dysphagia, Bleeding, Respiratory Difficulty. | Base of tongue | Excision & auto-transplantation |
| 4 | F | 23 | Lingual mass, Dysphagia | Base of tongue | Excision followed by Thyroid substitution therapy |
| 5 | M | 8 | Midline neck swelling | Subhyoid | Thyroid substitution therapy |
| 6 | F | 26 | Lingual mass, Dysphagia | Base of tongue | Excision followed by Thyroid substitution therapy |
| 7 | F | 17 | Lingual mass Dysphagia, Bleeding | Base of tongue | Thyroid substitution therapy |

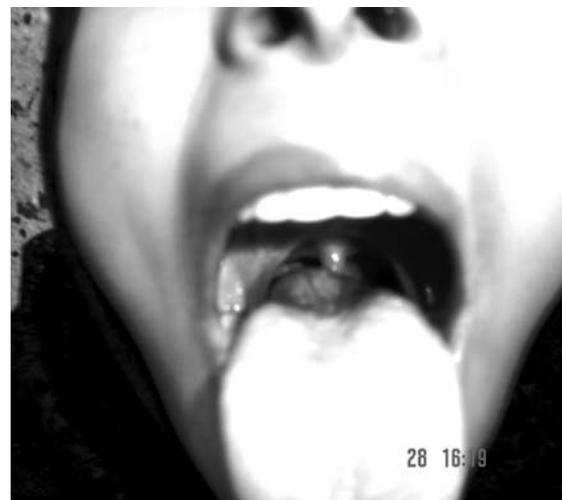


Figure 1. Clinical photograph showing ectopic thyroid tissue at the base of tongue in subject number 6, who presented with dysphagia.



Figure 2. Axial Contrast enhanced CT Scan at the level of the oropharynx showing the location of ectopic thyroid in the tongue base in subject number 6.

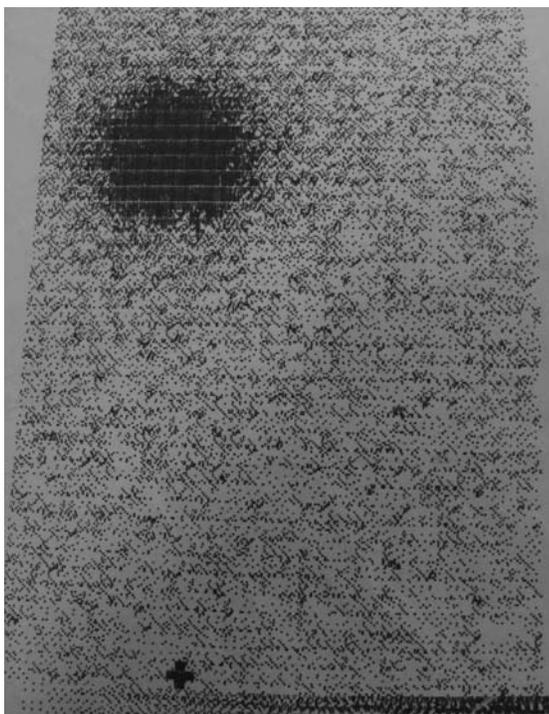


Figure 3. I¹³¹ scan showing uptake of the radiotracer superior to the normal location of the thyroid without any uptake at the normal site. The + sign in the picture denotes the suprasternal notch which proves absence of normal thyroid tissue.

DISCUSSION

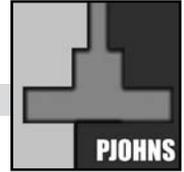
A MEDLINE search through PubMed using the keywords “ectopic” and “thyroid” yielded 1603 results. Most of the available literature featured case reports with one or two cases each. Only four case series or reviews were found with more than five cases.^{4,5,6,7} Apart from this, two case series of congenital hypothyroidism were found with more than five cases each of ectopic thyroid.^{8,9}

Late in the first month of life the anlage of the thyroid gland descends from the posterior dorsal midline of the tongue (actually the floor of the pharyngeal gut) to its final position in the lower neck. The developing thyroid gland descends ventral to the hyoid bone and laryngeal cartilages. The initial site of descent eventually becomes the foramen caecum, located in the midline at the junction of the dorsum of tongue and the tongue base. If the embryonic gland does not descend normally, ectopic or residual thyroid tissue (technically *ectopic thyroid*) may be found between the foramen caecum and the epiglottis.¹⁰

Kaplan *et al.* studied 30 children with ectopic thyroid gland with a female: male ratio of 2:1. Nine children were diagnosed within their first year of life.⁵ In the KEM Hospital study comprised of 36 patients, female preponderance was also noticed (female: male ratio of 11:7). Age of presentation was from five months to 40 years (mean age 14.3 years) in the same study.⁶ Yoon *et al.* reviewed 30 cases of ectopic thyroid reported in the Korean medical literature and added 19 cases from their institution. In this review, most cases of ectopic thyroid were diagnosed in patients aged between 1 and 29 years; it was also found to be more common in females (43 patients).⁴ In the present series, most of the patients were under 30 years of age with definite female preponderance.

Lingual thyroid tissue in the region of the tongue base adjoining the foramen caecum is the most common site in all the case series. Okstad *et al.* reviewed cases of thyroid ectopia over a period of 10 years. There were five cases of total ectopia; three of these were lingual thyroid, one was situated in the perihyoid region which had been clinically diagnosed as a thyroglossal cyst, while the fifth presented as a tumor in the lateral neck.¹¹ In the Korean study, lingual thyroid was found in 23 patients, a sublingual thyroid in 17 patients, combined type in seven patients, a prelaryngeal thyroid in one patient, and an intratracheal thyroid in one patient. Only in four cases was the thyroid gland in the normal position.⁴ In the KEM Hospital study, 17 patients (47%) presented with lingual thyroid and 19 patients (53%) had sublingual thyroid, which mainly presented as an anterior neck swelling.⁶ In this study, most of the cases were lingual thyroid followed by subhyoid midline neck mass.

Ectopic thyroid may present with symptoms from infancy to adulthood. Depending on the patient’s age, the symptoms may be drastic: infants and young children, whose lingual thyroid is detected



via routine screening may suffer from failure to thrive and mental retardation or may present with respiratory or feeding difficulty.¹² Growth retardation was the most common clinical finding at the time of diagnosis in Kaplan's series.⁵ In the neck, it may present as a midline neck mass mimicking thyroglossal cyst in early childhood. In the KEM series 53% of patients presented with an anterior neck swelling.⁶ Some cases may present with slowly progressing dysphagia and other symptoms of oropharyngeal obstruction before or during puberty, or even during pregnancy. This occurs as a response to the increased demand for thyroid hormone in these hypermetabolic states. In the Korean study, chief complaints at presentation were palpable mass in 20 patients, growth retardation in 10 patients and a sensation of lump in the throat in six patients.⁴ Carcinomatous changes may occur in median ectopic thyroid and in lingual thyroid.^{13,14} Nodal metastasis in lingual thyroid is also reported.¹⁴ In our series, dysphagia, sensation of lump in the throat, midline neck mass and bleeding per mouth were the presenting symptoms.

Clinical examination of ectopic thyroid includes a thorough head and neck examination with palpation of the thyroid gland to detect its location in the normal site. Indirect laryngoscopy usually detects lingual thyroid and flexible laryngoscopy is required to document the size of the mass.

Functionally 30-70% of the reported cases were hypothyroid and 70% had cervical atrophy.¹⁵ In the Korean study, 26 of 42 patients (61.9%) had hypothyroidism, and 16 patients (38.1%) were euthyroid.⁴ In the KEM study, 83% had hypothyroidism.⁶ In this series, all cases were biochemically hypothyroid. Functioning at marginal levels, they are subject to excessive thyroid stimulating hormone (TSH) stimulation, with resulting hyperplasia and compensatory enlargement. Occasionally, ectopic thyroid may present with features of hyperthyroidism like Graves' disease and Graves' ophthalmopathy.¹⁶

Regarding investigations, I¹³¹ and Tc⁹⁹ scanning are useful to detect the presence of ectopic thyroid tissue in the neck or in the base of the tongue. I¹³¹ scanning also alleviates the need for fine needle aspiration biopsy in lingual thyroid. However FNAB is still used in ectopic thyroid in the neck. On CT scans, ectopic thyroid tissue appears to be hyperdense and shows contrast enhancement.⁷

Jaromir *et al.* classified heterotopic thyroid tissue into three groups. Group 1 is named the ectopy dystopy group. Arrest of descent presenting as lingual, sublingual or subhyoid thyroid hypertrophy belongs to the ectopic subgroup. Simultaneous descent with the foetal basis of heart results in intrathoracic or primary retrosternal goiter (dystopy). Group 2 consists of accessory thyroid tissue & aberrant thyroid tissue. Accessory thyroid tissue develops in the midline in the presence of normal thyroid tissue in its physiological position. Descent

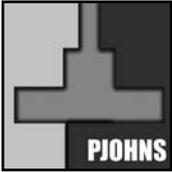
of thyroid outside the midline results in aberrant thyroid; however it has to be differentiated from metastasis of well differentiated thyroid cancer. Group 3 or heterotopic thyroid tissue consists of teratogenic development of thyroid tissue. *Struma ovarii* or abdominal thyroid tissue are examples of teratogenic development of thyroid.¹⁷

The management of ectopic thyroid is still a controversial issue. The primary factor to be considered in management planning is the high incidence of absence of normal thyroid gland. Long *et al.* reviewed first 10 described cases of ectopic thyroid, where five of the cases underwent excision. All the five cases developed myxedema. The rest of the five cases that underwent auto transplantation of excised tissue remained clinically euthyroid.¹⁸ Surgical excision and auto-transplantation of thyroid tissue followed by lifelong thyroid substitution therapy provides results similar to excision of the tissue alone followed by thyroid replacement therapy. However auto-transplantation is associated with more surgical time and auto-transplantation without a vascular pedicle may lead to failure. Moreover, malignant changes may occur in auto-transplanted ectopic thyroid tissue.^{13,14}

In the KEM series, 86% were treated medically, and surgery was performed in only 14% of cases with either recurrent bleeding or dysphagia.⁶ Rahbar *et al.* reviewed four cases of lingual thyroid from the Children's Hospital Boston that underwent surgical excision of the mass followed by lifelong hormonal replacement. They observed that majority of patients with lingual thyroid require surgical excision of the symptomatic mass, and in case of absence of orthotopic thyroid tissue, long-term thyroid hormone replacement is needed.¹² In the Korean series, 22 patients received thyroid supplement whereas 11 patients underwent surgical excision.⁴ In the present series, symptomatic lingual thyroid and midline neck mass needed surgical excision with or without auto-transplantation. The rest of the patients were treated with thyroxin suppression therapy.

While many authors advocate surgical excision of all ectopic thyroid tissue for fear of malignant changes, the risk of malignant changes in ectopic thyroid tissue may not be significantly greater than in colloid goitre or thyroid nodule – therefore vigilant follow up of the patient may be an acceptable management of choice rather than aggressive surgical removal.^{18,19}

In summary, ectopic thyroid is an uncommon entity that is more common in females. The age ranges and symptoms of presentation of ectopic thyroid vary from series to series and depend on two factors: 1) presence of normal thyroid gland along with ectopic thyroid; and 2) thyroid hormone secreting capacity of the ectopic thyroid tissue. Thyroid scintigraphy and CT scans remain the radiological investigations of choice. Thyroid status must be assessed in all cases of midline neck swelling and in base of tongue swelling, as the majority of these



patients are hypothyroid at presentation. FNAC should be avoided in lingual thyroid for risk of brisk haemorrhage but maybe performed in suspected ectopic thyroid in neck. Surgical excision may be avoided where this is the only functioning thyroid, provided vigilant follow up is observed except in cases where there is severe dysphagia, bleeding from the mass or respiratory obstruction. In the presence of normal thyroid gland surgical excision may be attempted. Hypothyroidism must be corrected first before undertaking any surgical intervention as auto-transplantation of ectopic thyroid does not provide any added advantage. The chance of malignancy in ectopic thyroid is extremely rare and if present it is usually papillary carcinoma or its follicular variant. Any suspicion of malignant change in the ectopic thyroid should be dealt with by complete excision of the mass based on oncological principles supported by radio-iodine therapy if required and lifelong follow up.

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