Case Series

Thyroid ectopia: a case series

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ABSTRACT

Ectopic thyroid is a form of thyroid dysgenesis and is defined as the presence of thyroid tissue in an abnormal location. Its prevalence is about 1 per 1,00,000-3,00,000 people, rising to 1 per 4,000-8,000 patients with thyroid diseases. The most common sites of ectopic thyroid are lingual (90%) and anterior neck (10%). They are usually diagnosed with hypothyroidism with a mass with, or without pressure symptoms in the 2nd or 3rd decade of life when there is increased demand of thyroid hormone. Presence of two ectopic foci of thyroid tissue simultaneously is rare, and very few such cases of dual thyroid ectopia have been reported. We present a case series of 3 ectopic thyroid with different clinical presentation. The first case is a 19-year-old boy who presented with clinical and biochemical overt hypothyroidism and was discovered to have only a sublingual ectopic thyroid gland. The second case is a 16-year-old girl who presented with dysphagia and on evaluation was found to have a dual ectopic thyroid gland in the lingual and sublingual region. The third case is a 28-year-old women who presented with a sub hyoid swelling which on evaluation was diagnosed as a dual ectopic thyroid in the lingual and sub lingual region.

Keywords: Ectopic thyroid, Dual ectopic thyroid, Subclinical hypothyroidism, Thyroid dysgenesis

INTRODUCTION

Thyroid ectopic is a form of thyroid dysgeneis and is commonly found anywhere along the obliterated thyroglossal cyst. Its prevalence is about 1/1,00,000-3,00,000 people increasing to about 1/4,000-8,000 patients with thyroid diseases. It is 4 times more common in females especially in the Asian population. Thyroid gland migration to the normal pre-tracheal position is controlled mainly by transcription factors like TITF1/NKX2-1, FOXE1, PAX8 and HHEX. They play a very important role as a precursor in thyroid development, migration and morphogenesis. The most common location of ectopic thyroid gland is lingual (90%) which usually present with dysphagia, choking, dyspnea and hemorrhage. The other less frequent sites in the head and neck are sublingual, trachea, sub-mandibular gland, lateral cervical regions, maxilla, palatine tonsils, carotid bifurcation, iris of the eye, and the pituitary gland. Thyroid ectopic in the ovaries is known as 'struma ovarii' and is considered as a thyroid tissue teratoma.

The indications for surgical removal of the ectopic thyroid includes malignancy, bleeding or ulceration of the gland, uncontrolled hyperthyroidism and severe local or respiratory symptoms. Some ectopic thyroid gland can be visible in the neck and females prefer surgical removal for cosmetic reasons.

The estimated incidence of carcinoma arising in an ectopic thyroid is less than 1%. In our series we present 3 different and unique cases of thyroid ectopia.
CASE SERIES

Case 1

A 19-year-old boy presented to us with a history of hypothyroidism for the past 3 years and was on thyroxine 250 mcg daily. A detailed history and clinical examination revealed a swelling in the sublingual region of size 3.5×3 cm. Oral examination showed no evidence of any ligual thyroid gland. Thyroid function test was in the normal range with elevated levels of Anti TPO and Anti thyroglobulin antibodies, suggestive of Hashimotos thyroiditis.

Ultrasound neck revealed the absence of thyroid in the normal pretracheal position and the presence of thyroid like tissue in the sublingual region of size 3.5×3 cm, isoechoic, well encapsulated with vascularity.

A Tc-99m pertechnetate thyroid scan was done and showed ectopic thyroid in the sublingual region with no tracer uptake in the normal pretracheal thyroid region, since the patient had no compressive symptoms and no suspicious features of malignancy in ultrasound neck he was treated conservatively with thyroxine and kept on close follow up.

Case 2

A 16-year-old girl was referred to us with complaints of dysphagia since the past 5 months and on detailed clinical and physical examination she was found to have a thyroid like swelling in the base of tongue and was diagnosed as case of ectopic lingual thyroid. A Thyroid function test was done which was suggestive of subclinical hypothyroidism.

Ultrasound neck showed absence of thyroid gland in the normal pretracheal region and the presence of a mass in the submental region of size 2×1.5 cm isoechoic and well encapsulated. She was then planned for a Tc-99m pertechnetate thyroid scan which revealed dual ectopic thyroid in the lingual and the submental region. Since the ectopic lingual thyroid only caused mild dysphagia she was started on thyroxine 50 mcg daily for the treatment of subclinical hypothyroidism as well as for gland size reduction. She was counselled regarding complications like bleeding, compressive symptoms and malignant transformation and was kept on close follow up.
Case 3

A 28-year-old lady presented with a midline swelling in front of neck since the past two years with history of pressure symptoms of dysphagia and dyspnea. On examination, the patient had a well-defined, firm swelling of 3x2.5 cm in the sub-hyoid region which moved with deglutition. Ultrasound revealed the absence of thyroid gland in its normal site and the presence of a mixed solid and cystic swelling in the sub hyoid region. Video direct laryngoscope examination showed the presence of a lingual ectopic thyroid. The fine needle aspiration of the sub-hyoid swelling was done and was reported as hyperplastic nodule with cystic degeneration (Bethesda 2). A Tc-99m pertechnetate thyroid scan was done and showed tracer uptake in the lingual and the sub-hyoid regions, which was suggestive of a dual ectopic thyroid gland. Patient was initially treated with thyroxine replacement therapy and subsequently underwent surgical excision of the sub hyoid thyroid tissue in view of her pressure symptoms. The histopathology report was consistent with that of ectopic thyroid tissue.

DISCUSSION

Thyroid ectopia is a congenital disease due to an abnormal migration of thyroid gland in the embryonic stage, which usually manifests later in life especially during increased physiological demand of thyroid hormones. Ectopic thyroid tissue most commonly is seen in females (65 to 80%). First ectopic thyroid case was reported by Hickman as a lingual thyroid in 1869 where a newborn presented with an upper airway obstruction 16 hours after birth. When thyroid tissue is present at sites other than its normal anatomical position it is referred to as ectopic thyroid. This happens when there is an abnormality in the normal descent of the median thyroid diverticulum from the posterior third of the tongue to its final pretracheal location. This normal descent usually occurs on the 3rd to 7th week of embryonic life. A number of transcription factors play a key role in morphogenesis of the thyroid, of which FOXE1 is required for thyroid migration. Failure of this migration results in the formation of ectopic thyroid tissue.

Most of the patients with ectopic thyroid are asymptomatic. Patients with ectopic thyroid usually have insufficient hormone production resulting in a hypothyroid state. When there is an increased demand for thyroid hormone as in during adolescence or pregnancy, ectopic thyroid tissue can enlarge in size and present as a neck swelling. Symptoms can arise in view of the size and location of the ectopic thyroid tissue as well as due to the associated thyroid dysfunction.

Ectopic thyroid tissue is subject to the same stimuli as a normally placed thyroid gland. The ectopic gland produces qualitatively normal hormones, although usually lesser in quantity. All diseases that affect the normal thyroid gland can also affect the ectopic thyroid tissue. Ectopic thyroid is a common association with congenital hypothyroidism with a prevalence rate of 24% to 60%. Hypothyroidism also might remain subclinical till puberty. Graves disease is very rarely detected in a patient with an ectopic thyroid.

Scintigraphy, using Tc-99 m, 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 ultrasound also unmasks additional sites of thyroid tissue. It is both sensitive and
specific for differentiation of an ectopic thyroid from other causes of midline neck masses. Radiological imaging modalities, such as grayscale or 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 presurgical evaluation of these cases. Last but not least, fine needle aspiration cytology (FNAC) provides considerable assistance in confirming the diagnosis of an ectopic thyroid. It is the only modality to differentiate between a benign and a malignant lesion. The most useful immunohistological marker is thyroglobulin.

There is no consensus about the optimal therapeutic strategy, perhaps 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). Some recommend complete surgical resection, considering the potential of malignant transformation. For cases completely asymptomatic and euthyroid, regular follow-up is recommended in order to detect mass enlargement or development of complications. For mild symptoms and hypothyroid states, levothyroxine replacement therapy may be effective, leading to considerable mass reduction.29

CONCLUSION

Ectopic thyroid is a rare and an important differential diagnosis in a case of neck swelling which should be ruled out by the surgeon as inadvertent removal of it will result in permanent hypothyroidism. A detailed history with thorough clinical evaluation along with adequate investigations like, video laryngoscopy, ultrasound neck and TC-99m pertechnetate thyroid scan will help in the diagnosis of a ectopic thyroid gland. Dual ectopic thyroid is a very rare finding. Conservative management is the usual mode of treatment. Thyroxine supplementation is given lifelong to suppress the TSH levels and decrease the size of the thyroid swelling. Surgery is only indicated in patients with obstructive symptoms, bleeding or with any suspicious features of malignancy.

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