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Colloid goitre of ectopic thyroid in an adolescent female- A therapeutic dilemma

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Abstract: Ectopic thyroid tissue refers to presence of thyroid tissue at locations away from the normal site in the pretracheal region in the lower neck. Few of such cases are associated with a functioning thyroid gland in its normal anatomical location. However in many cases, the ectopic thyroid gland is the only functioning thyroid. Radionuclide scan is mandatory when dealing with an ectopic thyroid lesion. A colloid goiter in such a case may be large enough to cause cosmetic disfigurement, but still may be functioning. Patient will be deprived of a functioning thyroid tissue, if such a case is treated surgically, necessitating thyroid hormone supplementation for the rest of patient's life.

Key words: Ectopic thyroid; Colloid goitre; Radionuclide scan; Surgical excision.

Introduction:

Ectopic thyroid tissue (ETT) refers to the presence of thyroid tissue in locations other than the normal pretracheal region in the lower neck between the 2nd and 4th tracheal cartilages. It is a rare developmental anomaly, with the prevalence being 1 per 100,000-300,000, and one in 4,000-8,000 patients with thyroid disease. Post mortem studies show that 7-10% of adults have asymptomatic ectopic thyroid tissue in the neck. More than 500 cases have been reported to date.[1,2] It commonly occurs in females with the female to male ratio of 3-4:1. [3,4,5] ETT can occur at any age ranging from 5months to 40 years, but it is usually seen in adolescence, pregnancy and menopause due to increased physiological demand of thyroid hormones and marked endocrine activity. ETT is seen as a result of failure or incomplete descent of thyroid gland from its origin to its normal position.[5] Lingual thyroid is the most common type of ETT accounting for 90% of the cases. [1] Most of the patients of ectopic thyroid are asymptomatic. Symptoms if exists are usually related to size, location and thyroid hormonal variations. Thyroid function tests, Ultrasound, computerized tomography(CT), magnetic resonance imaging(MRI), fine needle aspiration cytology(FNAC) followed by radionuclide thyroid scan can be used as a diagnostic tool. [3] Asymptomatic and euthyroid patients can be advised regular follow up and do not require any intervention. Surgery is reserved for symptomatic cases, suspecting malignancy and failure to respond to thyroxin. Any disease involving normal thyroid gland can also affect ETT. Incidence of malignant transformation is approximately 1%.[5] We report a case of colloid goitre of ectopic thyroid in a 17 year old female.

Case report:

A 17 year old female presented with a painless swelling in the anterior aspect of the neck just below the chin. She noticed the swelling before one year when it was very small like a pea nut, but the swelling started growing later. However she did not have any disturbance of respiration and deglutition. On

examination, she was in good general health state. There was a firm, non tender swelling measuring 3cm X 3cm in the anterior aspect of the neck, just above the level of the thyroid prominence. (Figure 1) The swelling was moving with deglutition, and also with protrusion of the tongue. A clinical diagnosis of thyroglossal cyst was made. She was advised Ultrasonography of the neck for confirmation of diagnosis. Ultrasonography revealed absence of normal thyroid glands in its anatomical location. Sonological features were suggestive of ectopic thyroid gland with nodule formation. Fine needle aspiration cytology was suggestive of colloid goitre. (Figure 3) Her thyroid function tests were within normal limits. Thyroid scan was performed with 99m-sodium pertechnetate, which revealed absence of uptake in the normal location of the thyroid gland. Normal uptake of technetium (1.6%) was seen in the ectopic thyroid tissue in the sub hyoid region. (Figure 2) Her parents were very much concerned about the cosmetic deformity caused due to the swelling. They were explained about the consequence of surgical excision and the need for thyroid hormone replacement lifelong. They wanted to get the swelling excised. Surgical excision of the mass was performed under general anesthesia. (Figure 4, 5) Patient withstood the procedure well. Postoperative period was uneventful. Histopathological features were suggestive of Colloid goitre. She is advised to take thyroxin tablets 100 µg daily, the dose being titrated according to the thyroid hormone levels.

Discussion:

Thyroid gland is the first endocrine organ to develop at 24th day of gestation. It develops as a median endodermal diverticulum in the pharyngeal wall between the first and second branchial arches at the foramen caecum which is the junction between anterior two third and posterior one third of tongue. Thyroid diverticulum becomes bilobed and descends in the neck and fuses with the 2 lateral diverticulae that are derived from the fourth pharyngeal pouch, which forms the parafollicular cells. During the process of descent, thyroid gland remains connected to the floor of the pharyngeal gut by thyroglossal duct. It descends in front of the hyoid bone and laryngeal cartilages to reach the final position in front of the trachea at 7th week of gestation. [1,3] ETT is the result of failure or arrest in migration of thyroid along the route of thyroglossal duct. It was first described by Hickman in 1869 in a newborn who was suffocated 16 hours after birth due to a lingual thyroid causing upper airway obstruction.[3] ETT can be lingual,

sublingual, sub hyoid, laryngeal and lateral. Lingual ETT is the most common site accounting for more than 90% of cases. [5] They usually present with dysphagia, dysphonia, stomatolalia, foreign body sensation, sleep apnea and hemorrhage. Lateral or submandibular ectopic occurs when the cells of lateral anlage do not join those of the median. These patients will have a palpable mobile painless mass in the submandibular region. Other less common sites are intratracheal and intrathoracic. Intratracheal ETT may be due to division of developing thyroid by trachea or ingrowth of thyroid tissue into the tracheal lumen. These patients may complain of dyspnea, cough, dysphagia, hemoptysis and stridor. Intrathoracic ETT accounts for 1% of mediastinal tumors and is found in mediastinum, lungs and heart.[2] An overdescent of thyroglossal duct remnants has been suggested as the cause in mediastinal location. The presence of ETT in genital tract is due to parthenogenetic development of germ cells into thyroid tissue after failure of all germ cells to migrate to the genital crest. [1] Intracardiac thyroid involves mainly the right ventricle. There is a close anatomic relationship between thyroid primordium and the developing myocardium and hence cardiac malformations are more frequently observed birth defects related to thyroid dysgenesis. Extremely rare locations of ETT are subdiaphragmatic involving ovaries, adrenal, gallbladder, esophagus, iris, pancreas, duodenum and mesentery. Unique cases of ETT in pituitary fossa, sphenoid sinus and uterus have been reported. In all these locations, the possible explanation may be aberrant migration or heterotopic differentiation of uncommitted endodermal cells. [2] It is very uncommon for two ectopic foci to be present simultaneously. In most cases, the first lesion is lingual or sublingual and the second is sub hyoid or supra hyoid or infra hyoid. [3]

Mean age at presentation is 18.7 years with equal distribution between sexes. Most of these patients are asymptomatic and euthyroid while remaining are hypothyroid with no radioiodine uptake in the region of normal TG. [3] ETT is common in females than males and can be seen at any age but it is usually seen in adolescence or puerperium due to increased physiological demand of thyroid hormones.

Differential diagnosis for a sub hyoid ectopic thyroid include thyroglossal cyst, Subhyoid bursitis and lymphadenopahy. Ultrasound scanning should be done as initial assessment in patients presenting with anterior neck masses. It also helps in conveying cystic or solid nature of the lesion. The sensitivity is increased by using color Doppler technique to demonstrate hypervascularity. [1,2] CT and MRI are useful in

detecting the location and extension of ETT when ultrasound fails. MRI is particularly useful in lingual thyroid to differentiate ETT from tongue muscle. [1,2] Chest radiograph is useful in intrathoracic goitre.

FNAC can be done in palpable ETT in the neck and is diagnostic in 95%-97% of cases. Endoscopic biopsy can be done in lingual and intratracheal ETT. [1] Scintigraphy using Tc-99m, I-131, or I-123, is useful in detecting ETT and shows the presence of thyroid tissue in other sites as well. It also tells whether ETT is the only functioning thyroid tissue and helps in planning treatment. [2] Thyroid function tests that assess the serum levels of T_3, T_4, TSH and thyroglobulin should be carried out in suspected cases of ETT. [1] Subclinical or clinical hypothyroidism is seen in 24%-60% of ETT patients, while the rest are often euthyroid. Very rarely hyperthyroidism is seen. [5] ETT can be affected with same diseases as normally located thyroid gland such as nodular goitre, hyperplasia, thyroiditis, adenoma and malignancy. The incidence of malignancy arising in ETT is approximately 1%. [1,6]

Asymptomatic and euthyroid patients do not require any treatment but can be kept under follow up. For those with obstructive symptoms and hypothyroidism, suppressive therapy with levothyroxine is administered which helps to achieve euthyroid state and reduce the size of the TG. [3] Surgical intervention is reserved in cases which fail to respond to thyroxine and if there is any pathology. [6] Most of the times ETT is the only functioning thyroid and excision is followed by lifelong hormone replacement. As the patients presenting with this disease are mainly young adolescent females, who are concerned about the cosmetic deformity caused by the swelling, they need to be counseled about the consequences of surgical excision. Transposition of ETT with the vascular pedicle flap into the muscle space at the floor of mouth, lateral pharyngeal wall, anterior rectus sheath or under strap muscles are recommended to retain some amount of thyroid function. [1,7]

Radioactive iodine 131 therapy is an alternative in patients refusing and not fit for surgery and in cases where surgery is not feasible due to anatomic difficulties. However, outcome is unpredictable as it may destroy any normally located thyroid gland as well. [1,5]

Conclusion:

Ectopic thyroid is a rare developmental anomaly. It commonly occurs in females at ages when physiological demand of thyroid hormones are high. Lingual thyroid is the commonest location and patients usually have hypothyroidism. An ultrasound and FNAC will help in confirming thyroid tissue and diagnosing any existing pathology. Thyroid scintigraphy has to be done to see if it is the only functioning thyroid tissue and if dual ectopic thyroid is present. Surgical excision should be done followed by life long thyroid hormone replacement. ETT though rare, should be kept in differential diagnosis of midline neck swelling as it is often misdiagnosed as thyroglossal cyst.

References:

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Legend of Illustrations:

Figure 1. Clinical photograph showing a midline cervical swelling above the level of thyroid notch

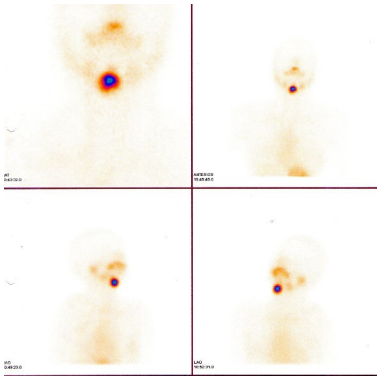

Figure 2. 99m pertechnetate scan showing uptake of technetium in the sub hyoid region, with absence of uptake ate normal anatomical site of thyroid

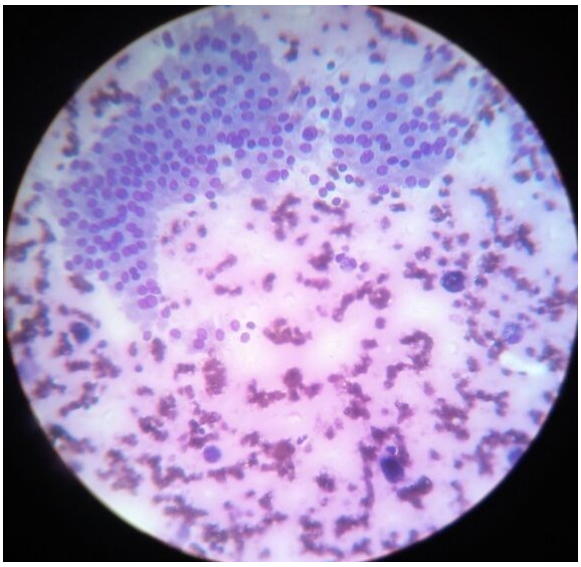

Figure 3: FNAC showing follicular epithelial cells in monolayer sheets and macrophages in a background of colloid. (40x MGG stain)

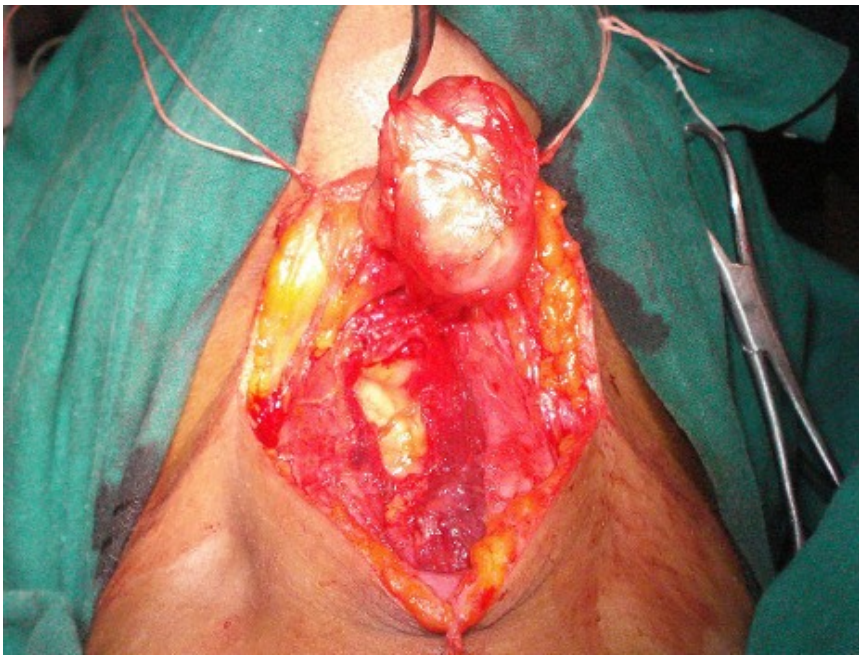

Figure 4: Intraoperative photograph showing excision of mass

Figure 5: clinical photograph of specimen