



Median ectopic thyroid - a rare cause of midline neck lump

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Abstract :

Median ectopic thyroid is a rare entity. Its description is limited to a few case reports in literature. The clinical importance is related to its misdiagnosis as thyroglossal cyst resulting in unnecessary excision and permanent hypothyroidism as this is the only thyroid tissue in these patients. We describe a patient with median ectopic thyroid whom we managed. We present this case to reiterate the need for precise diagnosis in these patients.

Keyword : Ectopic thyroid, Thyroglossal cyst, Midline neck swelling

Case report

A 14 year old girl presented with a swelling in front of the neck of 10 years duration. This has been gradually increasing in size. She had no pressure symptoms secondary to this swelling. Examination revealed a 3x3 cm, firm subhyoid swelling which moved up with deglutition and protrusion of the tongue (Fig 1). There was no palpable goitre. On evaluation she was found to be

hypothyroid with a TSH of 45.65mIU/ml (normal: 0.3 - 4.5mIU/ml). An ultrasonogram of the neck showed a 3 x 1.7 cm heterogeneous lesion in left paramedian position with internal vascularity. The thyroid gland was not seen in the normal location. A functional assessment of this swelling with Tc99m thyroid scan

revealed activity only in the region of the swelling and no uptake in the thyroid bed (fig 2). This was diagnostic of median ectopic thyroid. As she was hypothyroid she was started on thyroxin and reviewed a year later. A repeat ultrasound of the neck showed that the lesion had increased in size to 3.7 x 2.3 x 1.2 cm. TSH had decreased to 5.46mIU/ml. In view of the documented increase in size of the swelling and cosmetic concerns surgical management was planned. She underwent a cervicalexploration through a Kocher's incision. Intraoperatively a 4cm reddish brown lesion was seen in the subhyoid region on the left side of the thyroid cartilage which was excised (Fig. 3). The thyroid gland was not present in the normal position

The histopathology was reported as nodular hyperplasia of thyroid.

Discussion:

The thyroid gland originates as an endodermal diverticulum from the floor of the pharynx at the 4th week of gestation. It descends from the foramen cecum to reach its final position by the 7th week of gestation, anterior and lateral to the 2nd, 3rd and 4th tracheal rings. As a result a tract is formed from the foramen cecum to the thyroid called the thyroglossal duct. This normally atrophies prior to definitive thyroid formation.¹ Ectopic thyroid is defined as thyroid cells located at any site other than its normal position.² This entity was first reported by Hickman in 1869 in a neonate.³ Its prevalence is approximately 1 in 1,00,000 to 300,000 persons and is more commonly seen in females.⁴ The most common location is at the base of the tongue or lingual thyroid, accounting for about 90% of ectopic thyroids. Other locations include any site in its descent from foramen cecum to anterior to trachea, intratracheal, esophageal, mediastinal, aortic, intracardiac and abdominal.⁵

Transcription factors appear to play an important role in the organogenesis of the thyroid gland. These include transcription factor TTF 1/ NKX 2-1 (responsible for thyroid specific expression of thyroglobulin and thyroperoxidase), PAX 8, HHEX and FOXE 1 (essential for early stages of thyroid morphogenesis). It is speculated that thyroid dysgenesis may be due to mutations in the genes regulated by these transcription factors.⁶

Median ectopic thyroid results from incomplete descent of the thyroid. This usually presents as a midline neck swelling. They can be associated with pressure symptoms due to its size and location. The clinical implication of this entity lies in the fact that it is often misdiagnosed as a thyroglossal cyst leading to its excision. In majority of patients this is the only thyroid tissue and excision will result in permanent hypothyroidism.

Median ectopic thyroid is associated with hypothyroidism in one third of patients.⁵ A thyroglossal cyst containing ectopic thyroid tissue should also be considered. In this situation the ectopic thyroid tissue may be harboring a malignancy as has been reported in literature.⁷ The differential diagnosis to be considered include angiomas, fibromas, lymphangiomas, epidermal cyst and midline branchial cyst.

Scintigraphy with ^{99m}Tc, ¹³¹I or ¹²³I is the most important diagnostic tool. This can detect ectopic thyroid tissue and also show the absence or presence of thyroid in its normal location. Radiological imaging modalities such as ultrasonogram, computerized tomography and magnetic resonance imaging can provide information on the exact location and extent of the ectopic tissue. This will help in better pre-surgical evaluation. Fine needle cytology may also help differentiate between a benign and a malignant lesion.

Treatment generally depends on size of swelling and local symptoms. In the era prior to availability of imaging diagnosis, surgical exploration of midline neck lumps were standard. These patients were diagnosed with median ectopic thyroid during surgery, the gland divided in half and each half placed on side of the trachea behind the sternothyroid muscles. This is not advocated because of the poor function of implanted thyroid and the ease of thyroxine replacement. Small swellings with subclinical hypothyroidism may be observed or given a trial of levothyroxine.

Indications for excision include large swellings that are cosmetically unsightly, presence of pressure symptoms (airway compromise, dysphagia, voice change or cough) or a proved malignancy.

Conclusion:

Developmental defects occurring at an early stage of embryogenesis result in median ectopic thyroid. The clinician must account for this rare entity and differentiate it from other midline neck swellings, especially a thyroglossal cyst. Majority are asymptomatic. Thyroid scintigraphy plays an important role in establishing the diagnosis, although other radiological modalities may contribute. Surgery is the treatment of choice in symptomatic cases.

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Fig:1 Fig:2

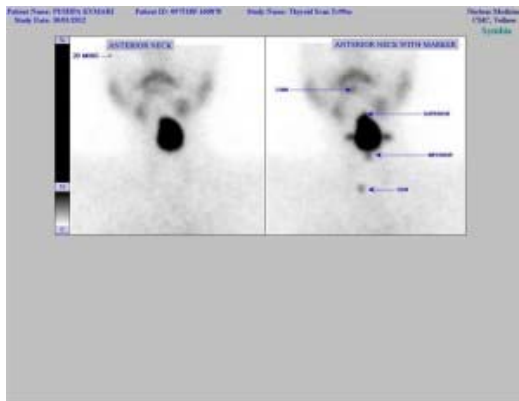


Fig:3