Case Report

AGENESIS OF ISTHMUS OF THYROID GLAND: EMBRYOLOGICAL BASIS–A CASE REPORT


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ABSTRACT

The thyroid gland is composed of two lateral lobes connected by a narrow median isthmus, thus giving an ‘H’ shaped appearance to the gland. A wide range of morphological variations and developmental anomalies of the thyroid gland have been reported in the literature such as hypoplasia, ectopic thyroid, hemi-agenesis and agenesis. Out of these, the incidence of agenesis of the isthmus of thyroid gland is rare. During routine dissection of the male cadaver, agenesis of isthmus of thyroid gland in the midline was noted. The arteries supplying the gland were confined only to the respective lobe with absence of anastomosis. Agenesis of isthmus of thyroid has clinical, phylogenetic and surgical significance. The knowledge of various developmental anomalies of the gland and variations in neurovascular relations will help the surgeon in better planning of a safe and effective surgery.

Keywords: agenesis, anomaly, thyroid, thyroglossal duct, thyroid isthmus.

INTRODUCTION:

The thyroid gland is a brownish-red and highly vascular endocrine gland, placed anteriorly in the neck, extending from the fifth cervical to the first thoracic vertebrae. It is ensheathed by the pre-tracheal layer of deep cervical fascia. The gland is composed of two lateral lobes connected by a narrow median isthmus. The normal size of each lobe of the thyroid gland has been described to be 5 cm long, its greatest transverse and anteroposterior extent being 3 cm and 2 cm respectively. The isthmus measures about 1.25 cm transversely as well as vertically and is usually placed anterior to the second and third tracheal cartilages.[1]

Thyroid gland is the first endocrine gland to start developing in the embryo. It is well known for its developmental anomalies ranging from common to rare. Common anomalies include persistence of pyramidal lobe and thyroglossal duct cyst. Rare anomalies are agenesis or hemiagenesis of thyroid gland, agenesis of isthmus alone or aberrant thyroid gland.[2]

The anomalies of the development of the thyroid gland distort the morphology of the gland and may cause clinical functional disorders and various thyroid illnesses. Incidence of agenesis of the thyroid isthmus has been reported to vary from 5% to 10% by Pastor et al (2006) and from 8% to 10% by Marshall (1895).[3,4] Ranade et al (2008) reported a 33% incidence of agenesis of the isthmus.[5] The present case report highlights about the agenesis of isthmus of thyroid gland and its developmental and clinical significance.

CASE REPORT

During routine dissection teaching to the 1st year MBBS students in the head and neck region of an elderly male cadaver, it was noticed that there was absence of isthmus of thyroid gland. The surrounding structures were cleaned and observation
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was made for any other associated anomalies and ectopic tissue. There was no scar on cervical region, suggesting that the patient has not undergone any surgery. The thyroid gland has two separate lobes with complete agenesis of isthmus. The two lobes were separate without any tissue intervening between them (Fig. 2). The individual lobes were supplied by branches of superior and inferior thyroid arteries. No accessory thyroid arteries were present. Anastomosis between the posterior and inferior branches of superior thyroid and inferior thyroid arteries was also noticed but there was no anastomosis between the arteries of right and left side.

![Diagram and Photograph of Thyroid Gland](image_url)

Fig. 1: (A) Diagram showing right and left lobe of thyroid gland along with thyroid isthmus (B) Photograph showing right and left lobe of thyroid gland along with thyroid isthmus

![Photograph of Thyroid Gland](image_url)

Fig. 2: Photograph showing right and left lobe of thyroid gland. Black star (*) represents absent thyroid isthmus
DISCUSSION

The agenesis of isthmus can be explained as an anomaly of embryological development. Phylogenetically, the thyroid follicles are structured to acquire a bilobed gland. The two lobes are joined together by an isthmus in the upper part of trachea (Fig.1 A & B). The isthmus may be missing in amphibians, birds and among mammals-Monotremes, certain Marsupials, Cetaceans, Carnivores and Rodents. In rhesus monkey (Marcus rhesus), the thyroid glands are normal in position but there is absence of isthmus.[2]

The thyroid gland begins to develop as a median thickening of endoderm on the floor of the pharynx between the first and second pharyngeal pouches. This area later invaginates to form the median diverticulum, which appears in the later half of the fourth week. This thyroid diverticulum grows in allometric proliferation, becoming a solid cellular cord called the thyroglossal duct. The duct grows caudally and bifurcates to give rise to the thyroid lobes and the isthmus. At the same time, when its caudal growth is taking place, the cephalic end of the thyroglossal duct degenerates.[6] This isolates it from the pharyngeal endoderm with the cessation of proliferation of the endodermic cells from which follicular cells of the gland are derived.

Rarely a high division of the thyroglossal duct can generate two independent thyroid lobes with the absence of isthmus. Morphologically difference in the evolutionary origin does not result in any changes in thyroid function. Usually agenesis of isthmus is difficult to determine unless the patients referred for other thyroid disease.[2] The absence of the isthmus can be associated with other types of dysorganogenesis, such as the absence of a lobe or the presence of ectopic thyroid tissue.[7] The reports in the literature suggest that chromosome 22 could play a role in the thyroid development.[8]

Agenesis of the thyroid isthmus is the complete and congenital absence of the thyroid isthmus as is defined by Pastor et al (2006).[3] In their study, they had reported agenesis of isthmus of thyroid gland with enlarged lobes in a Caucasian cadaver. Marshall (1895) documented the variations in the gross structure of the thyroid gland in 60 children, varying in age from a few weeks to 10 years and the absence of the isthmus was reported to be 10% in this group.[4] Ranade (2008) et al reported absence of isthmus in 35 out of 105 cases (33%), of which 8 were female cadavers.[5] According to Braun et al (2007), the isthmus was missing in 4 cases of the 58 cadavers they studied.[9] Won and Chung (2002) reported that in 3% of the cases studied, the isthmus was absent and the lateral lobes of the thyroid were separated.[10] The incidence in Northwest Indians is reported to be 7.9% in gross specimens.[11]

Allan (1952) reported 2.0-4.0% incidence of absence of isthmus in his study. He also observed that a band of connective tissue named levator glandulae thyroideae extended from the apex of right or left lobe or isthmus of thyroid gland to the hyoid bone.[8] Anson (1996) also reported the variations in size, shape and level of the thyroid gland. The isthmus was absent in 6.0 to 8.0% of cases in his study[12]. Devisankar et al (2009) reported a case of agenesis of isthmus of thyroid gland with bilateral levator glandulae thyroideae.[2] In another study conducted by Dixit et al (2009) who reported 14.6% incidence of agenesis of isthmus of thyroid in their study.[13] Kumar et al (2010) reported a case of absence of isthmus of thyroid gland with pyramidal lobe and levator glandulae thyroideae arising from right lobe.[14]

Clinically, the diagnosis of agenesis of the isthmus can be done with scintigraphy, which can also be performed with an overload of thyroid stimulating hormone (TSH). The diagnosis can also be done with the aid of ultrasonography, computerized tomography (CT), magnetic resonance imaging (MRI) or during a surgical procedure. In asymptomatic patients with nodular goiters, fine-needle aspiration biopsies and eventually immunohistochemistry tests are useful. When an image of the absence of isthmus is observed, a differential diagnosis against autonomous thyroid nodule, thyroiditis, primary carcinoma, neoplastic metastasis and infiltrative diseases such as amyloidosis should be considered.[2]

In the present case, the agenesis of thyroid isthmus was not associated with any other anomalies of the gland and it may be a congenital anomaly. Interestingly, the branches of superior thyroid and inferior thyroid arteries did not anastomose in the median plane. This type of variations should be kept
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in mind during transthyroid tracheostomy procedures.

CONCLUSION

Agenesis of isthmus of thyroid gland is rare in humans, the incidence varying from 5% to 10%. In present case report, there was absence of isthmus of thyroid gland in an elderly male cadaver. This agenesis can be explained as an anomaly of embryological development i.e. a high division of the thyroglossal duct giving rise to two independent thyroid lobes with absence of isthmus. Agenesis of isthmus can be associated with other types of dysorganogenesis, such as the absence of a lobe or the presence of ectopic thyroid tissue and hence in clinical practice when such a condition is diagnosed, it is necessary to perform a differential diagnosis against other pathologies such as autonomous thyroid nodule, thyroiditis, etc. The surgeon planning a thyroidectomy must be prepared to find variations like ectopic thyroid nodules around the normally located thyroid gland. Proper identification of vessels is very important in order to avoid major complications.

REFERENCES