Agenesis of isthmus of thyroid gland - a case report

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Abstract

A wide range of morphological varieties and developmental anomalies of the thyroid gland have been reported in 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 and very few cases have been reported. During routine dissection of the male cadaver, agenesis of isthmus of thyroid gland in the midline was noted. It resulted in two lateral lobes and 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.

Keywords: agenesis, anomaly, thyroglossal duct, thyroid.

Introduction

The thyroid gland, brownish red and highly vascular endocrine gland is placed anteriorly in the neck, extending from 5th cervical to the 1th thoracic vertebrae. It is ensheathed by the pretracheal 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 5cm×3cm×2cms and its weight being 25gms. The isthmus measures about 1.25cm in width positioned anterior to the second and third tracheal rings.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

Incidence of agenesis of the thyroid isthmus has been reported to vary from 5% to 10% by Pastor et al and from 8% to 10% by Marshall3,4. Ranade et al in their study on anatomical variations of the thyroid gland reported a 33% incidence of agenesis of the isthmus5. The knowledge of agenesis of thyroid isthmus is of help to the surgeon on better planning of a safe and effective surgery.

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 there was absence of isthmus of thyroid gland. The surrounding structures were cleaned and observation was made for any other associated anomalies and ectopic thyroid tissue.

There were no scars in the cervical region, suggesting that the patient has not under gone any surgery. The thyroid gland had two separate lobes, with complete agenesis of isthmus. The two lobes were separate without any tissue intervening between them. (Fig. 1)

The individual lobes were supplied by branches of superior and inferior thyroid arteries. No accessory thyroid arteries were present. Anastomoses between the posterior and inferior branches of superior thyroid and inferior thyroid arteries was also noticed. But there were no anastomoses between the arteries of right and left side. (Fig. 2).

Discussion

Allan in 1952 reported 2.0 - 4.0% incidence of absence of isthmus of thyroid in his study. He also observed that a band of connective tissue named levator
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Figure-1 showing absence of isthmus of thyroid gland.

Figure-2 showing two separate lobes with absence of isthmus of thyroid gland.

glandulae thyroideae extended from the apex of right or left lobe or isthmus of thyroid gland to the hyoid bone.4

The variations in the size, shape and level of the thyroid gland was reported by Anson in 1996. The isthmus was absent in 6.0 to 8.0% of cases in his study7.

Agenesis of the thyroid isthmus is the complete and congenital absence of the thyroid isthmus as is defined by Pastor et al5. In their study, they had reported agenesis of isthmus of thyroid gland with enlarged lobes in a Caucasian cadaver. According to Gruber (quoted by Testus and Latarjet) the incidence of agenesis of isthmus is about 5%.4 Marshall documented the variations in the gross structure of the thyroid gland in 60 children, varying in age from few weeks to 10 years and the absence of the isthmus was reported to be 10% in his group5. Kanade et al reported absence of isthmus in 35 out of 105 cases (33%) of which eight were in females5.

According to the study by Braun et al, the isthmus was missing in four cases of the 58 cadavers they studied9. Won and Chung have reported that in 3% of the cases studied, the isthmus was absent and the lateral lobes of the thyroid were separated10. The incidence in Northwest Indians is reported to be 7.9% in gross specimens11. Devisankar et al reported a case of agenesis of isthmus of thyroid gland with bilateral levator glandulae thyroideae in the year 200912. In another study conducted by Dixit et al reported 14.6% incidence of agenesis of isthmus of thyroid in their study in 200912. In 2010, Kumar et al reported a case of absence of isthmus of thyroid gland with pyramidal lobe and levator glandulae thyroideae arising from right lobe13.

Agenesis of thyroid isthmus can be explained as an anomaly of embryological development. Phylogenetically, the thyroid follicles are structured to acquire a bilobed shaped gland. The two lobes were joined together by an isthmus in the upper part of trachea.

The isthmus may be missing in amphibians, birds and among mammals- Monotremes, certain Marsupials, Cetaceans, Carnivores and Rodents. In rhesus monkey (Macacus rhesus), the thyroid glands are normal in position but there is absence of isthmus2.

ThyroGLOSSAL duct arises from the endodermal epithelium of primitive pharynx at the level of 2nd and 3rd pharyngeal arch, when it descends downward; its caudal end bifurcates and gives origin to the thyroid lobes and the isthmus. At the same time the cephalic end of the thyroglossal duct degenerates14. 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 separation of thyroglossal duct can engender two independent thyroid lobes and pyramidal lobes with the absence of isthmus5.

The morphological difference in the evolutionary origin does not result in any changes in thyroid function.
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Usually agenesis of isthmus is difficult to determine unless the patients referred for other thyroid diseases. Agenesis of isthmus can be associated with dysorganogenesis related to developmental anomalies of thyroid gland such as absence of either lobe or presence of ectopic thyroid tissue in the nearest cervical region.

Reports in the literature suggest that chromosome 22 could play a role in the thyroid development.

Agenesis of isthmus can be diagnosed via scintigraphy, ultrasonography, CT and MRI. When suspected, the individual may be directed for a differential pathological diagnosis such as autonomous thyroid nodule, thyroiditis, primary carcinoma, neoplastic metastases, and infiltrative diseases such as amyloidosis.

In the present case, the agenesis of thyroid isthmus was not associated with other anomalies of the gland and it may be a congenital anomaly. Interestingly, the glandular branches of Superior thyroid and Inferior thyroid arteries did not anastomose in the median plane, which is a variant from other studies regarding agenesis of thyroid isthmus. This type of variations should be kept 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 the 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 knowledge of this variation is important for surgeons especially for surgical intervention, physicians and radiologists for further course of diagnosis and therapeutic use.

References


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