

Agenesis of the thyroid isthmus

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SUMMARY

We describe a case of thyroid isthmus agenesis (the incidence varies from 5% to 10%). This absence can be explained as an anomaly of embryological development and can be associated with other types of disorganogenesis, such as the absence of a lobe or the presence of ectopic thyroid tissue.

In human clinical practice, when an image of the absence of isthmus is found it is necessary to perform a differential diagnosis against other pathologies such as the autonomous thyroid nodule, thyroiditis, etc.

Key words: Thyroid gland – Endocrinology – Malformations

INTRODUCTION

Agenesis of the thyroid isthmus is the complete and congenital absence of the thyroid isthmus.

Philogenetically, in some species in which the thyroid follicles are organized in a gland, this gland can acquire a bilobate shape, in which the lobes join together in front of the upper part of the trachea by an isthmus or bridge of thyroid tissue. The isthmus may be missing, as happens in amphibians, birds and, among the mammals, in the monotreme, certain marsupials, cetaceans, carnivores and rodents. It seems that these differences do not

have an evolutionary origin, because the morphology does not result in any changes in thyroid function and, additionally, the follicular structure is similar in species that are distant from one another in taxonomic classifications. In all the higher primates, including human beings, the gland is composed of two lateral lobes joined by an isthmus located in the anterior cervical region, at the level of the first tracheal rings and protected by the infrahyoid muscles.

It is difficult to determine the incidence of agenesis of the thyroid isthmus because the diagnosis is usually done in populations of individuals presenting with other thyroid diseases.

According to Grüber (quoted by Testut et al., 1978) the incidence is about 5% while according to Marshall (1895) it is about 10%.

CASE REPORT

After anatomical dissection of the thyroid gland of an adult (65 years old) caucasian man (Fig. 1), we observed that it was composed of two enlarged lobes (5.2 x 3 cm the right one and 5.5 x 3.2 the left one) with no type of nexus between them. In the patient's clinical history there was no reference to any surgical intervention to the neck and neither were any scars found in the cervical region. No ectopic thyroid tissue in the migration site of the thyroid cells from the base of the tongue was observed.

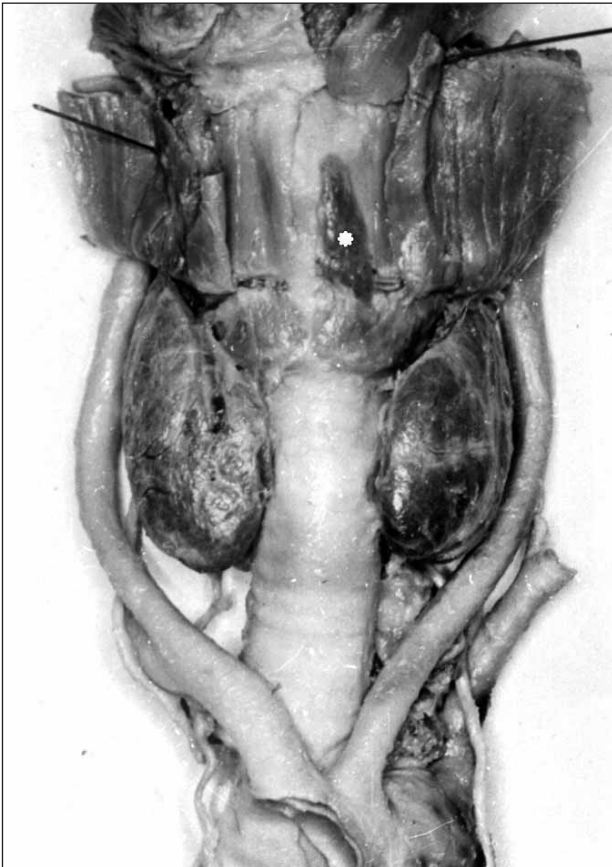


Figure 1. Anterior view of the thyroid area, in which the isthmus agenesis can be observed. * = Lymphatic node.

DISCUSSION

The absence of the isthmus of the thyroid gland can be explained as an anomaly of embryological development.

The adult thyroid gland has two kinds of endocrine cells, the follicular and the parafollicular cells, or "C" cells, which are originated from two different embryological cell families. The follicular cells come from the endodermic cells of the primitive pharynx and the parafollicular cells come from the neural crest (Le Douarin et al., 1974).

The thyroglossal duct is an odd formation which originates from the endodermic epithelium of the primitive pharyngeal base, at the level of the second and third brachial arches. Thereafter, it becomes lobe-shaped. The so-called lateral thyroid glands merge with these lobes and contribute with the cells of the neural crest, which will produce "C" cells.

The pharyngeal endodermic epithelium begins its differentiation by transforming itself into a slightly depressed plate (Sgalitzer, 1941), which proliferates and invaginates to form a spherical diverticulum with multi-stratified walls that stands out in the aortic pouch.

The thyroid diverticulum grows in allometric proliferation, becoming a solid cellular

cord called the thyroglossal duct. Then, its caudal end bifurcates and gives origin to the thyroid lobes and the isthmus.

At the same time that its caudal growth is taking place, the cephalic end of the thyroglossal duct degenerates (Sgalitzer, 1941). This isolates it from the pharyngeal endoderm and eliminates the endodermic cells forming it. The persistence of islets of endodermic cells from the thyroglossal duct due to the absence of cell death explains the nodular aspect of the aberrant thyroid glands.

A high division of the thyroglossal duct can generate two independent thyroid lobes with the absence of isthmus.

The absence of an isthmus can be associated with other types of dysorganogenesis, such as the absence of a lobe or the presence of ectopic thyroid tissue (Duh et al., 1994).

Clinically, the diagnosis of agenesis of the isthmus can be done with scintigraphy, which can also be performed with an overload of TSH, in the event of there being non-functioning thyroid tissue in the isthmus. The diagnosis can also be done with the aid of ultrasonography, computerized tomography (C.T.), magnetic resonance imaging (M.R.I.) or during a surgical procedure. When the condition is suspected it is necessary to perform an in-depth interview addressing previous surgical procedures in the cervical region (isthmectomies due to neoplasms, decompressive techniques due to thyroiditis or due to transthyroid tracheotomies).

When an image of the absence of isthmus is observed, a differential diagnosis against the following pathologies should be carried out: a) Autonomous thyroid nodule; b) Thyroiditis; c) Primary carcinoma; d) Neoplastic metastases; and e) Infiltrative diseases such as amyloidosis.

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