

CASE REPORT

# Papillary carcinoma arising in a submental-intralingual thyroglossal duct cyst

## *Carcinoma papillare a sede sottomentoniera-intralinguale insorto da cisti del dotto tiroglossa*

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### SUMMARY

An extremely rare case of papillary carcinoma is described arising in a submental-intralingual thyroglossal duct cyst. A 27-year-old male presented with a painless swelling in the submental region extending to the tongue which had gradually been increasing in size over the last three months. Upon examination of the neck, a tender right paramedian mass was found in the submental region just above the hyoid bone, it was mobile upon swallowing without clinical evidence of cervical nodes. The Sistrunk surgical technique, extended to the submental and intra lingual region was used to radically remove the mass. The occurrence of carcinoma of the thyroglossal duct cyst, even though rare and unexpected, should always be considered an option, and histologically ruled out mainly on account of the atypical localization, such as the submental-intralingual reported herein. The two different surgical approaches reported in the literature, one more conservative and the other more aggressive, apparently alternatives, are, instead, complementary and adequate when strict diagnostic criteria and adequate follow-up, are observed.

KEY WORDS: Head and neck neoplasm • Papillary carcinoma • Thyroglossal cyst • Sistrunk procedure

### RIASSUNTO

*Gli Autori riportano un caso estremamente raro di un carcinoma papillare a sede sottomentoniera-sottolinguale insorto da cisti del dotto tiroglossa. Un giovane di 27 anni si è presentato alla nostra osservazione riferendo dolore in regione sottomentoniera e linguale con difficoltà alla deglutizione da circa tre mesi. Alla palpazione del collo si evidenziò la presenza di una tumefazione in regione sottomentoniera, a sede paramediana destra, al di sopra dell'osso ioide, mobile alla deglutizione, senza linfonodi clinicamente palpabili. La massa fu radicalmente asportata effettuando l'exeresi mediante tecnica descritta da Sistrunk allargata alla regione sottomentoniera. L'insorgenza di un carcinoma papillare da una cisti del dotto tiroglossa è un evento raro e inaspettato. Tuttavia deve essere sempre sospettato e istologicamente escluso specie nelle tumefazioni a localizzazione rara quale quella a sede sottomentoniera descritta. In letteratura sono riportati due differenti approcci chirurgici per la relativa exeresi, uno più demolitivo, l'altro più conservativo, con risultati sovrapponibili; in ogni caso è necessario, in questi casi, seguire adeguati criteri diagnostici e stretti controlli post-operatori.*

PAROLE CHIAVE: Tumori della testa e del collo • Carcinoma papillare • Cisti del dotto tiroglossa • Intervento di Sistrunk

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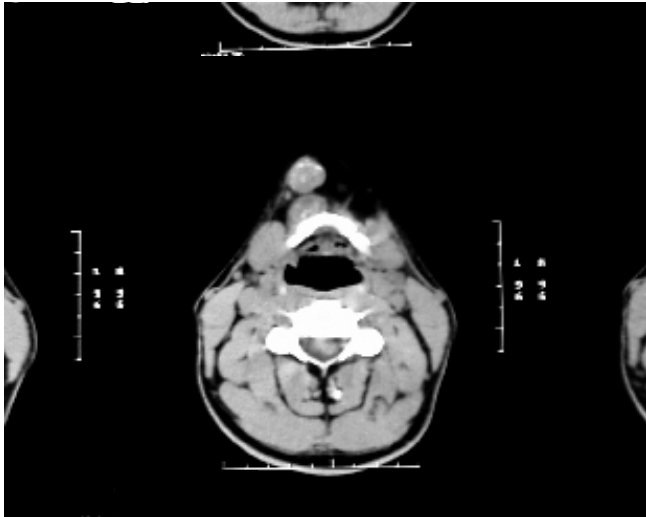
## Introduction

Thyroglossal duct cysts (TDC) are cystic expansions, arising from the abortive obliteration of the duct during embryogenesis, representing the most common congenital abnormality of thyroid development, involving around 7% of the population<sup>1</sup>. The presence of thyroid tissue, in a TDC, is due to its embryologic origin and ranges from 1.5% to 62% of cases<sup>2,3</sup>. Thyroglossal duct carcinomas are very uncommon findings, occurring in approximately 1% of all the thyroglossal duct cysts, often incidentally discovered following surgical excision<sup>4</sup>. There is predilection for females and the mean age of patients is within the fourth decade, with age at presentation, ranging from 1 to

82 years. The case presented here refers to a 27-year-old male with the unexpected finding of a papillary carcinoma arising in a submental-intralingual TDC and is described with special regard to the rarity of the localization and the different options in the management strategy when carcinoma is found incidentally following surgery.

## Case report

A 27-year-old male presented in our Department with a 3-month history of a gradually enlarging painless swelling in the submental region extending to the tongue. He had no evidence of other diseases. Examination of the neck showed a right paramedian tender mass in the submen-



**Fig. 1.** Axial section of a CT of the neck showing a well-defined soft tissue lesion, low-density with a smooth, thin wall.

tal region above the hyoid bone, mobile with swallowing, measuring 3 x 2 cm, without clinical evidence of cervical nodes; the thyroid gland was found to be normal.

Computed Tomography (CT) scan with contrast of the neck, showed a right paramedian submental and intralingual multi-loculated cystic structure with rounded solid components inside (Fig. 1). Laboratory investigations revealed normal T3, T4 and thyroid-stimulating hormone (TSH) levels. The patient was not given any other treatment. The Sistrunk surgical technique, extended to the submental and intralingual regions, was performed to radically remove, with a cuff of healthy surrounding tissue, the mass, that at intra-operative examination appeared to extend through the body of the tongue, keeping, however, a good cleavage plane from the intrinsic lingual muscles (Fig. 2). Post-operative histological examination demonstrated a 3 x 2 cm cystic mass surrounded by fibro-muscular tissue containing a papillary carcinoma about 1 cm in diameter within the cyst wall (Fig. 3).

The post-operative course was regular; 2 months after surgery T3, T4 and TSH levels, both thyroid scintigraphy and cervical magnetic resonance imaging (MRI), were normal. The patient accepted to begin hormone suppression and strict long-term follow-up.

For this case, all procedures were approved by the Institutional Review Board.

## Discussion

TDCs are common congenital neck masses which have a comparable sex distribution, presenting in several locations along the cervical midline. Of these, 60% are located between the hyoid bone and the thyroid cartilage, 13% in the substernal region, 24% above the hyoid bone including the submental site, 2% intra-lingual<sup>1,2</sup>. As far as concerns the distribution according to age, 31% occur



**Fig. 2.** Surgical view: resection of the cyst and the tract in continuity with mid portion of the hyoid bone.



**Fig. 3.** Papillary Carcinoma arose in the wall of the thyroglossal duct cyst (hematoxylin-eosin stain, original magnification x 100).

before the age of 10 years, 20% in the second decade, 14% in the third decade and 35% after 40 years of age. Although they are usually benign lesions, in about 1% of cases, TDC may develop neoplastic changes<sup>3,4</sup>. Most TDCs are located in the thyrohyoid region and 94% of these originate from the thyroid, being primarily papillary (including mixed papillary/follicular forms) while 6% are squamous cell carcinomas. Follicular and anaplastic carcinomas are very rare, while medullary carcinomas, instead, have not been reported, probably on account of the ultimobranchial origin of the C-cells thus supporting the theory of primary de novo origin of this rare tumour by the ectopic thyroid nests of the cyst wall rather than the metastatic spread hypothesis through the duct from the thyroid carcinoma. Furthermore, a third hypothesis of synchronous papillary carcinoma, in a TDC and the thyroid gland, can be explained as representative of multifocal tumour and not as metastatic foci<sup>5,6</sup>.

The incidence of cervical and distant metastasis is low, with, respectively, a rate of about 8% and 1.3%<sup>4</sup>.

Due to these aetiopathogenetic controversies, strict criteria are required to diagnose a primary TDC carcinoma, as proposed by Joseph and Komorowski<sup>7</sup>. These are a thyroglossal remnant, ectopic thyroid nests within the cyst wall and a clinically normal thyroid gland.

The case reported here, was characterised by the presence

of a neoplastic focus within the cyst wall and of a clinically and radiologically normal thyroid gland, supporting the suspicion of a primary papillary carcinoma arising in a TDC. Furthermore, its localization in the submental intra-lingual region, even less common for benign TDC, makes it a very distinctly rare case. Most thyroglossal carcinomas are located in the thyro-hyoid region, and the closer the cyst is to the lingual area, the greater the percent of salivary (mucous or seromucous), rather than thyroid tissue within its walls<sup>4</sup>. Regarding the pre-operative diagnosis, finding a TDC carcinoma is unusual. As observed in the present case, patients are generally euthyroid, and the mass is often asymptomatic and not distinguishable from other more common, benign, histological types, in terms of growth, location, size and consistency<sup>3</sup>. Imaging (ultrasound, scintigraphy, CT scan) are unable to preoperatively diagnose malignant diseases<sup>8</sup>, even though they are mandatory to determine the characteristics of the lesion (cystic, solid, capsulated, vascularised), to detect suspicious enlarged neck lymph nodes, and to check that the thyroid gland is in its usual position, to avoid removing an ectopic gland<sup>1</sup>. Regarding the fine needle aspiration biopsy (FNAB), it is not so easy to obtain sufficient material for diagnosis, and if positive cells for carcinoma are found, it could be helpful for the pre-operative planning. MRI with gadolinium was performed to complete the evaluation of the neck for local spread, cervical metastasis and thyroid gland, to exclude here the presence of lesions<sup>9</sup>.

Surgical treatment of these carcinomas is still debated. The small number of cases reported in the literature, the lack of long-term follow-up required for this kind of tumour, and the young age of the patients, contribute to maintaining these different treatment schedules. Some Authors<sup>15710</sup> believe that well-differentiated incidentally discovered thyroid carcinomas arising in a TDC and limited within their walls, without positive histological margins and cervical metastasis spread, in the presence of a clinically and radiologically normal thyroid gland, can be managed adequately by the Sistrunk operation<sup>11</sup>, thyroid suppression and strict long-term follow-up. The same approach was followed in our case, having those clinical and pathological criteria. Resection of the cyst and the tract, was performed which extended to the *foramen caecum*, at

the base of the tongue, in continuity with the mid portion of the hyoid bone. On the other hand, other Authors<sup>12-14</sup> have suggested a more aggressive approach characterised by the Sistrunk procedure, total thyroidectomy, post-operative radioactive iodine therapy and thyroid hormone replacement, based upon the observation that papillary thyroid carcinoma may metastasize through the thyroglossal duct remnant without a lesion being clinically detected in the gland itself. There is a 11-27% incidence of occult thyroid gland carcinoma, reported in patients with TDC carcinoma, who underwent thyroidectomy<sup>12-15</sup> and the long natural history of papillary thyroid carcinoma requires long-term surveillance, justifying this approach. Some Authors<sup>11</sup>, in the past, considered that papillary carcinoma in TDC was the expression of a multifocal disease also present in the thyroid gland or that primary occult thyroid tumour had metastasised to the duct and developed a cystic form. The evidence now available in the literature indicates that these tumours are generally primary, arising in ectopic thyroid tissue associated with TDC and the incidence of cancer in the thyroid gland removed prophylactically is much lower than expected if the ductal lesion was always metastatic<sup>14</sup>.

In conclusion, the occurrence of carcinoma of the TDC, although rare and unexpected, has always to be taken into consideration and later histologically excluded, also on account of atypical localization, such as the submental-intralingual, reported here. The two different treatment approaches, one more conservative and one more aggressive, proposed in the literature, apparently alternative, become, instead, complementary and both adequate if strict diagnostic criteria are observed. The Sistrunk operation is adequate for most patients presenting with a clinically and radiologically normal gland<sup>15</sup>. The more aggressive one, including total thyroidectomy, with or without neck dissection, radioactive iodine ablation therapy, and thyroid-stimulating hormone suppression, are necessary in clinically or radiologically suspicious synchronous neoplastic lesions in the thyroid gland<sup>15</sup>. However, due to the long natural history of papillary thyroid carcinoma, conservative management has to be chosen only if the patient shows good compliance to undergoing long-term surveillance by means of scrupulous follow-up.

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