Case report

Papillary carcinoma on a thyroglossal duct cyst: diagnostic problems and therapeutic dilemma. A case report

Un raro caso di carcinoma papillare insorto su cisti del dotto tireoglosso: problematiche diagnostiche e dilemmi terapeutici

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SUMMARY

Thyroglossal duct cysts are one of the most common congenital abnormalities of the cervical region. Complications of these swellings are rare, and among these, appearance of a carcinoma has also been noted. We present a case of papillary carcinoma arising in a thyroglossal duct cyst in 20-year-old woman with a swelling of about 4 cm, located at the middle region of the neck over the hyoid bone. Our patient was treated using a modified Sistrunk operation, in which thyroidectomy proved crucial for the correct diagnosis and continuation of appropriate treatment. Our case confirms the difficulty in distinguishing a primitive thyroglossal duct carcinoma from a synchronous metastatic papillary carcinoma of the thyroid. This dilemma often remains unresolved.

KEY WORDS: Thyroglossal duct cysts • Papillary carcinoma • Sistrunk operation

RIASSUNTO

Le cisti del dotto tireoglosso sono fra le più comuni anomalie congenite della regione cervicale. Le complicanze di queste tumefazioni sono rare e, fra queste, è stata descritta la comparsa di un carcinoma. Presentiamo un nuovo caso di carcinoma papillare insorto in una cisti del dotto tireoglosso in una giovane donna di 20 anni portatrice di una tumefazione di circa 4 cm, localizzata nella regione media del collo al di sopra dell'osso joide. La nostra paziente è stata trattata mediante l'operazione di Sistrunk, nella quale la tiroidectomia ha rappresentato uno step avanzato risultato cruciale per il raggiungimento di una corretta diagnosi e la continuazione di un appropriato protocollo terapeutico. Il nostro caso conferma la difficoltà nel distinguere un carcinoma del dotto tireoglosso primitivo da una metastasi sincrona di carcinoma papillare della tiroide. Questo dilemma spesso rimane irrisolto.

PAROLE CHIAVE: *Cisti del dotto tireoglosso • Carcinoma papillare • Intervento di Sistrunk*

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Introduction

Thyroglossal duct cysts are one of the most common congenital abnormalities of the cervical region [1 2]. They originate from the persistence of the thyroglossal duct epithelium in the route of the descent of the thyroid gland from the base of the tongue to the anterior lower neck region [2]. Complications of these swellings are rare, and among these, the appearance of a tumour has also been noted [3]. We present the case of 20-year-old woman suffering from papillary carcinoma on the thyroglossal duct cysts. The case is interesting for its clinical-pathological findings, and especially for its controversial diagnostic aspects.

Case report

A 20-year-old woman came to our observation for a swelling of about 2.5 cm, located at the middle region of the neck over the hyoid. The swelling had a tense-elastic consistency, and was mobile and nontender, which had formed about a year before. Ultrasound examination was compatible with thyroglossal duct cyst, with no abnormalities in the thyroid, which was in site and size, and the absence of suspicious adenopathy. Thus, there was a clear indication for surgical excision of the lesion using a modified Sistrunk technique, which involved removal of the cyst en bloc from the soft tissue surrounding the central portion of the front bone hyoid.

At macroscopic examination, the sample showed a cystic area with a gelatinous content and a firm mass in the wall.

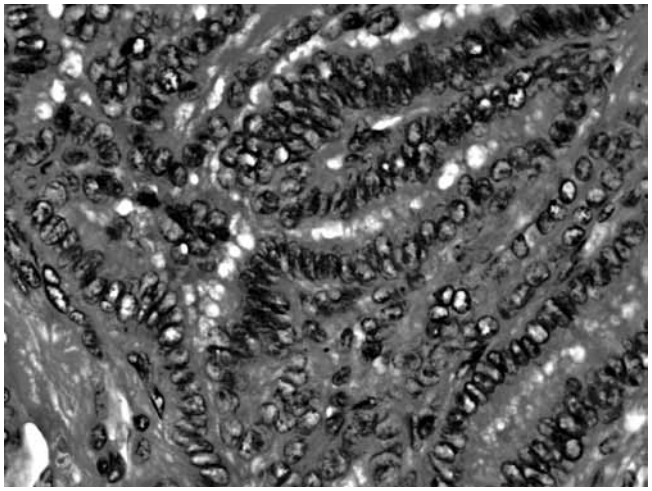

Fig. 1. Tumour tissue with a classic papillary appearance (HE 40X).

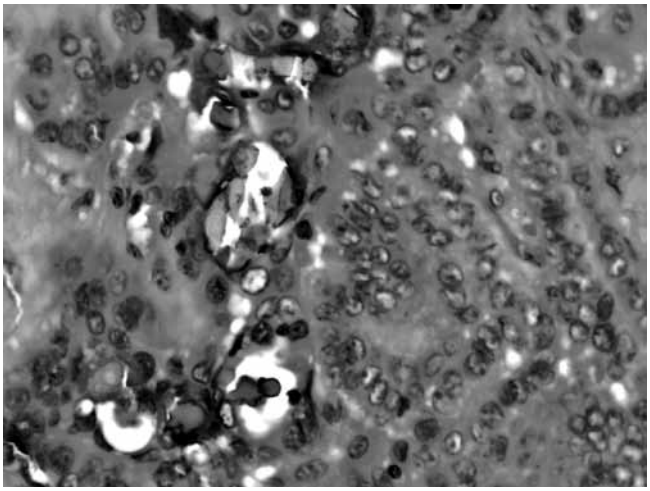

Fig. 3. Tumour tissue with typical microcalcifications (HE 40X).

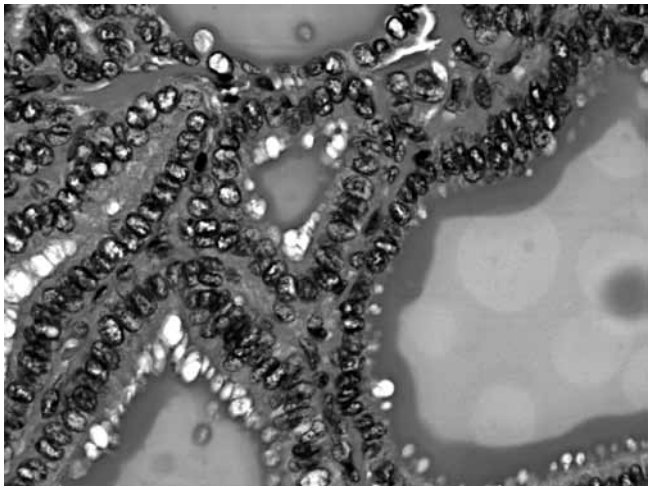

Fig. 2. Tumour tissue with voluminous and overlapping cells, with clear, irregular nucleus, with characteristic notches or grooves (HE 40X).

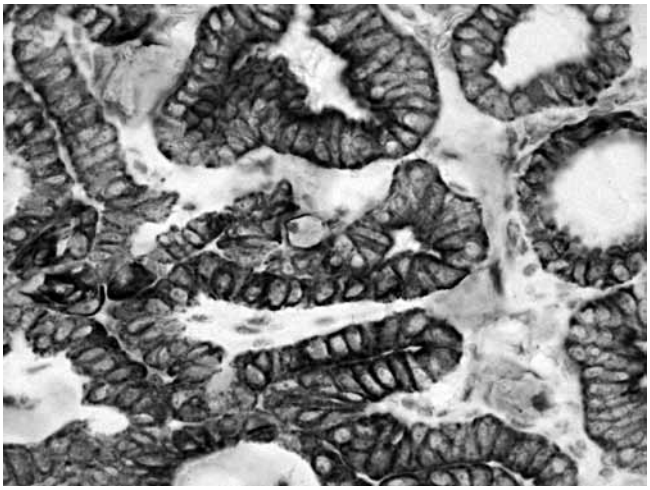

Fig. 4. Immunohistochemistry: clear positivity for CK 19, a marker of papillary carcinoma showing squamous differentiation, is observed (CK19 40X).

Histology revealed a papillary lesion (Fig. 1) with complex architecture in conjunction with follicles of varying sizes. The cellular component showed nuclear clearing or ground-glass appearance. The nuclear contour was irregular with grooves and rarely with any pseudoinclusion (Fig. 3). Occasionally, psammoma bodies were seen. Immunohistochemical staining revealed reactivity for high-molecular weight, cytokeratin (CK19, (Fig. 4) and galectin-3, while HBME was not expressed. Furthermore, proliferation index assessed with Mib1 was moderate. Thus, a diagnosis of papillary carcinoma arising in the thyroglossal duct cyst was made.

After diagnosis, the patient was subjected to further investigation, and as recommended by the consultant endocrinologist, underwent total thyroidectomy. Final histological examination of the surgical specimen showed the presence of foci of papillary carcinoma, with CK19 and galectin-3 expression. The patient was then subjected to two rounds of radioiodine therapy. No recurrence has been observed over one year of follow-up.

Discussion

Although thyroglossal duct cysts represent the most frequent congenital cervical abnormalities encountered in both adults (7% of the population [4]) and children, neoplastic lesions, either benign or malignant, appear to be particularly unusual and quantifiable in only 1–2% of the cases. The clinical presentation of a neoplasm of thyroglossal duct is similar to that of median cysts of the neck, and therefore, diagnosis is almost always made at the time of histological examination.

Papillary carcinoma, as noted in the thyroid gland itself, is the most common histological type (80%), followed by mixed papillary-follicular (8%) and squamous cell carcinoma (6%). The surgical procedure, reported by Sistrunk in 1920, is considered to be the treatment of choice for radical excision of the thyroglossal duct cyst. The original procedure included resection of the cyst along with the body of the hyoid, extending to the foramen cecum at the floor of the mouth. Later, the technique was modified, and

today, not all surgeons extend the resection further than the body of the hyoid.

Our case, although unusual considering the average age of presentation of this type of lesion, does not differ from the clinical presentation, treatment protocol and timing for diagnosis reported in the literature [1 5], However, the finding of foci of cancer cells after thyroidectomy presented interesting insights regarding an issue that is still controversial. Indeed, despite the fact that more than 50% of cases of papillary carcinoma of the thyroglossal duct have not been identified as thyroid cancer [6], it is often difficult to distinguish a carcinoma on thyroglossal duct cysts from metastatic thyroid cancer.

The histological criteria for diagnosis of primary cancer of the thyroglossal duct provide the need to distinguish the lesion from cystic lymph node metastases and to observe a normal thyroid gland, preferably by microscopic observation [7]. However, these diagnostic criteria are often disregarded as 30% of cases show synchronous neoplastic lesions. Moreover, in many cases presented in the literature, there are no histological thyroid specimens.

Our case confirms the difficulty in distinguishing a primary thyroglossal duct carcinoma from a metastatic papillary carcinoma of the synchronous thyroid [3], which often remains unresolved. In our patient, despite the fact that the preoperative blood chemistry and instrumental thyroid tests were all negative, the presence of papillary carcinoma in the lining of the cyst, the higher rate described in young patients and the capsular invasion of tumour prompted us to perform total thyroidectomy [8].

In conclusion, we present a new case of thyroglossal duct carcinoma, diagnosed and treated using modified Sistrunk operation, in which thyroidectomy proved crucial for correct diagnosis and continuation of appropriate treatment.

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