BRIEF COMMUNICATION

Primary Papillary Carcinoma in Thyroglossal Cysts. Case Reports and Literature Review

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Abstract The thyroglossal cyst can exceptionally appear as a primary cyst carcinoma. We discuss a series of 6 adult patients, assessed for long-lasting asymptomatic suprahyoid or lateral-to-larynx mass. The images showed a heterogeneous mass invading adjacent soft tissues. Fine needle aspiration biopsy did not contribute to diagnosis. We performed a Sistrunk procedure in all cases, 3 combined with total thyroidectomy and 1 with neck dissection. The postoperative course was favourable. No additional treatment was required, without evidence of recurrence in follow-up. The management is controversial due to the limited number of cases reported. Some classifications based on size and extent have been proposed to define the surgical treatment of such cysts.

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Carcinoma papilar primario en el quiste tirogloso. Serie de casos y revisión de la literatura

Resumen El quiste del conducto tirogloso se puede manifestar excepcionalmente como un carcinoma primario. Se presenta una serie de 6 casos de pacientes adultos que consultaron por masa suprahioida o paralaringea asintomática de larga evolución. Las imágenes mostraron una masa heterogénea con extensión a tejidos blandos. La biopsia aspirativa de la lesión no contribuyó al diagnóstico. En todos los casos se realizó cirugía de Sistrunk, en 3 casos combinada con tiroidectomía total y en uno, con vaciamiento. El curso posquirúrgico fue favorable. No


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Requirieron tratamiento adicional y no presentaron recurrencias. El manejo es controvertido debido a los pocos casos reportados. Se han propuesto algunas clasificaciones basadas en el tamaño y la extensión para definir el tratamiento quirúrgico.

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Introduction

The thyroid gland begins to develop during the third week of pregnancy. It is openedly connected to the tongue by the thyroglossal duct, which normally involutes at the eighth week of life. The hyoid bone develops simultaneously from the second and third branchial arcs, enabling a close interconnection. If the obliteration procedure fails, due to secreting epithelial obstruction of this duct, a sinus or cyst may present. Thyroglossal duct cyst is the most common congenital anomaly of the cervical mid line in children. It presents from the base of the tongue to the lower part of the mid line of the neck. 60% is found between the hyoid bone and the thyroid cartilage, 13% is retro-terinal, 24% is above the hyoids, and 2% is submentonian. 70% is located in mid line, the rest are paramedian. Half of cases are diagnosed in adults over 20.

Up to 1.5% of thyroglossal duct cysts have been reported as hosting carcinomas, which usually present after 40 years of age. There are slightly less than 200 reported cases in literature. Due to its low incidence, management of this tumour is controversial and there is no consensus about how it should be treated. The aim of this article is to present 6 cases of patients with carcinoma in thyroglossal cysts, along with treatment administered, postoperative development and follow-up. In addition we will review outcomes reported in literature, with emphasis on surgical treatment offering the best oncological outcome.

Methods

We reviewed the clinical history of 6 patients treated by the Servicio de Cirugía de Cabeza y Cuello de la Facultad de Salud of the Universidad de Caldas and in the Clínica de la Policía de Bogotá between 1st January 2000 and 31st December 2013. The following were assessed: clinical presentation; symptoms; diagnostic imaging; fine needle aspiration biopsy (FNAB) outcome; surgery performed; laboratory reports on the disease, and evolution during follow-up based on clinical aspects and imaging. Recurrence was defined as when a mass was Identified in the surgical site or the appearance of lymph nodes with a confirmation of carcinoma following biopsy. Follow-up was calculated from date of surgery until the last clinical control. The decision regarding the surgical procedure and the follow-up schedule was determined by the surgeon treating the case. For follow-up the use of imaging tests was considered (scans or tomography) and for those patients who had undergone total thyroidectomy, measurement of specific serum markers (thyroglobulin and antithyroglobulin) was included.

Consultation was made every 4 months to assess local or regional tumour relapse, with the performance of an ultrasound scan of the neck and an annual chest X-ray. When concomitant thyroid nodules were identified, an FNAB was obtained to determine their histological characteristic.

Results

There were 6 patients, 5 women and one man, aged between 38 and 58 and with a mean age of 48.8 (Table 1). The main symptom was a painless slow-growing mass, with few signs of compression, with the exception of one patient who presented with dysphagia. One patient with spread to the laryngeal cartilage consulted due to local pain. Location was variable: 2 tumours were exclusively in mid line suprahypoid, one had spread to the submaxillar glands, one to the floor of the mouth, one paralaryngeal and one hypopharyngeal.

In all cases, the initial scan showed a heterogeneous (solid and cystic) tumour. Preoperative tomography in all of them showed a solid tumour with a cystic component, with extracapsular invasion almost always to adjacent soft tissues and with undefined sides (Fig. 1). Tumour size was between 3 and 5 cm. The FNAB of the cyst was unable to define malignancy and on 2 occasions when an associated thyroid nodule was found, its FNAB showed follicular neoplasia.

In all cases, the Sistrunk procedure was used, and in 2 this was combined with total thyroidectomy (Fig. 2). Pathological anatomy confirmed papillary carcinoma of a standard variety in 5 cases and a follicular variety in one (Fig. 3). The findings in the thyroid glands corresponded to the follicular adenoma. No additional treatment was necessary in 5 patients. During follow-up, 4 months after the operation, one patient presented with lateral cervical adenomegaly of 2.5 cm in diameter. On suspicion of local and regional relapse, FNAB was performed which indicated metastatic papillary carcinoma, and modified dissection in combination with total thyroidectomy was performed. Laboratory reports showed a goitre with 0–66 affected lymph nodes. There were no complications in the surgical procedures. To date there have been no reported tumour relapses.

Discussion

It has been reported in literature that up to 1.5% of thyroglossal cysts may be sites for tumours. Low incidence of cases has resulted in a scarcity of information on clinical findings regarding malignancy.

Up to 62% of patients with thyroglossal cyst tumours have ectopic thyroid tissue and it is believed that this is the site for de novo neoplastic transformation. Another theory
Table 1 Findings.

<table>
<thead>
<tr>
<th>Age</th>
<th>Gender</th>
<th>Dissemination</th>
<th>Symptoms</th>
<th>Size (cm)</th>
<th>Thyroid Nodules</th>
<th>Thyroidectomy</th>
<th>Variety</th>
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</thead>
<tbody>
<tr>
<td>42</td>
<td>Fem</td>
<td>Floor of mouth</td>
<td>Odynophagia</td>
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<td>Yes</td>
<td>Yes</td>
<td>Follicle papillary</td>
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<tr>
<td>38</td>
<td>Fem</td>
<td>Suprahyoid</td>
<td>Mass</td>
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<tr>
<td>53</td>
<td>Fem</td>
<td>Submaxillary</td>
<td>Mass</td>
<td>5</td>
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<td>No</td>
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</tr>
<tr>
<td>56</td>
<td>Male</td>
<td>Hypopharyngeal</td>
<td>Dysphagia</td>
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<td>No</td>
<td>No</td>
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</tr>
<tr>
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<td>Suprahyoid</td>
<td>Mass</td>
<td>4</td>
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<td>Yes</td>
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<tr>
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<td>Fem</td>
<td>Paralaryngeal</td>
<td>Pain</td>
<td>5</td>
<td>No</td>
<td>No</td>
<td>Standard papillary</td>
</tr>
</tbody>
</table>

Figure 1 Contrasted tomography of the neck, showing a heterogeneous (solid and liquid) tumour with undefined sides and right suprahyoid location with sub-maxillary compression and invasion of the floor of the mouth.

proposes that the carcinoma of the cyst actually corresponds to a metastasis, with primary focus on an occult thyroid cancer.3

A thyroglossal cyst is frequently diagnosed before 10 years of age, although in a third of cases it presents in adults over 20.1 1.5% may host a carcinoma, usually incidental, which is discovered in surgical specimen testing: 87% are papillary, 5% scaly cell, 1.7% follicular and 0.9% anaplastic.5 The patients of our series did not coincide with the epidemiological data found by others and the tumour occurred more frequently in women with a ratio of 6:1, but presentation mean age was 40. To date only 22 cases have been reported in the paediatric population.6

The carcinoma has a similar clinical presentation to that of benign cysts. Uncommonly, there are symptoms of dysphonia or dysphagia. In general an asymptomatic mass presents in the midline of the neck, although there are cases of cervical lateral location.7 In this series, the majority consulted with a mass in different locations to the classical one. In the majority of cases, patients were euthyroid status. Cancer diagnosis is normally made after surgical resection.

Figure 2 Block resection of a carcinoma of the thyroglossal cyst (hyoid bone, soft tissues, cyst and thyroid gland).

Figure 3 Pathological anatomy of a classical papillary carcinoma inside the thyroglossal cyst. Staining: Hematoxylin–eosin stain and enlargement ×4.
In 21% of cases the carcinoma was locally invasive and in 11% of cases there was metastasis of the cervical lymph nodes.8

Widström’s criteria were used for the histological diagnosis, which indicated that the carcinoma had to be within the wall of the remaining duct. It had to be different from a cystic nodal metastasis by demonstration of a scale epithelium layer and the presence of normal thyroid follicles in the remaining duct wall. No malignant thyroid gland tumour was to be found.

Imaging techniques such as scan, gammagrapy and computed tomography are usually not useful for preoperatively diagnosing malignancy. In the patients in this series several findings were suggestive of the malignancy described by others, such as the presence of a mural cell tumour in the cyst, sometimes with microcalcifications, invasion of the cyst wall, a solid image within the cyst or along the thyroglossal duct, or a complex invasive mass in mid line.10

Tomography is useful for determining the surgical approach limits. This should be performed on elderly patients, those with recent infection, or with laryngeal symptoms.

FNAB gives an accurate result in only solo 66% of cases, but may be used as an additional tool for diagnosis.11

Thyroglossal cyst carcinoma management is controversial. No consensus has been reached regarding the need or non need for additional treatments to the Sistrunk procedure, such as total thyroidectomy with or without total dissection of central nodes or modified nodes in the neck, treatment with radioactive iodine and hormonal suppression.12-14 Complete surgical resection of the cyst, including the thyroglossal duct and the central part of the hyoid bone (Sistrunk technique), may be sufficient for definitive management. In Patel13 analysis the only significant predictor of overall survival was the radicality of the cyst resection. The patients treated with simple resection had a worse prognostic than those treated with the Sistrunk procedure, with a general rate of cure above 95%. Others consider thyroidectomy in addition to Sistrunk to be routine in all patients due to the possibility of concomitant thyroid cancer in 1%–45%, the possible posterior use of radioactive iodine and to facilitate the use of thyroglobulin for follow-up.

Staging according to risk is one proposal for the identification of patients who would benefit from further surgical treatment.12 When the clinical thyroid gland tests normal under imaging scans, the Sistrunk procedure without thyroidectomy is valid in patients under 45 without a history of radio therapy in the neck, with a tumour under 1.5 cm, without any invasion of the cyst wall, low grade, free margins and with no lymph node or distant metastasis. Thyroidectomy is recommended in patients over 45 or with a history of cervical radiotherapy for any tumour found in the gland, or a tumour over 1.5 cm with invasion of the cyst wall. When the tumour has spread locally, with nodal compromise or metastasis, a similar approach to that of papillary cancer of the thyroid is recommended.

The challenge is whether to complete the procedure with thyroidectomy or not, when laboratory findings have shown details of carcinoma in the thyroglossal cyst. Tharmabala12 proposes staging risk into 3 classifications (Table 2). For low risk, observe and wait; for moderate risk, total thyroidectomy, hormonal suppressing treatment and radioactive iodine and for the high risk group, vertical lymph node dissection in addition to the other treatments. All patients must have follow up with a neck scan and be re-assessed every 6 months during the first year and annually after that. The prognosis of thyroglossal cyst duct carcinoma is similar to that of papillary thyroid cancer.

### Conclusions

Clinical presentation and several events such as masses which are irregular in appearance and which invade adjacent soft tissue, of variable location which may even be lateral, lead to suspicion of the presence of carcinoma. This enables an appropriate surgical approach to be taken prior to surgery. The Sistrunk procedure appears to be sufficient and only in selected cases is thyroidectomy performed or the lymphatic dissection of the neck. Tumour staging based on tumour size, histopathological findings, invasion of adjacent tissue, resection site edges, age, and the presence of nodal or metastatic compromise may be useful for treatment management. Follow up is similar to that of papillary thyroid carcinoma, generally based on clinical criteria and imaging.

### Conflict of Interests

The authors have no conflict of interests to declare.
References