The US Appearance of Thyroglossal Duct Carcinoma

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Purpose

Thyroglossal duct cyst (TDCs) is the most common non-odontogenic cyst in the neck, representing approximately 70% of all congenital neck abnormalities. It occurs as a result of anomalous development and migration of the thyroid gland during the fourth through eighth weeks of gestation. It is a cystic remnant along the course of the thyroglossal duct, between the foramen cecum of the tongue base and the thyroid bed in the visceral space. [1,2] Ghaneim and Atkin indicated that 60% are located between the hyoid bone and the thyroid cartilage, 13% in the supra-sternal region, 24% above the hyoid bone including the submental site and 2% intra-lingual (fig. 1). [3]

When examined pathologically, TDCs have a variable number of histologic components, including columnar, cuboidal, and/or non-keratinized stratified squamous epithelium. Ectopic thyroid tissue is present in a proportion of TDCs, with estimates ranging widely, from 1.5 to 62%. The differential diagnosis of TDCs includes branchial cleft cyst, lipoma, metastatic thyroid carcinoma, dermoid cyst, sebaceous cyst and enlarged lymph node. [4]

The two most common complications of TDCs are infection (fig 2) and malignancy, the latter of which occurs in 1-4% of cases. Virtually every type of thyroid carcinoma has been identified within a TDCs, including, in descending order of frequency, papillary, mixed follicular-papillary, squamous, follicular, anaplastic and Hurthle cell carcinoma. There are no documented cases of medullary carcinoma arising in thyroglossal duct cyst. This is not surprising, because medullary carcinomas arise from parafollicular cells originating from the ultimobranchial bodies of the fourth branchial pouch rather than the thyroid anlage.

Thyroglossal duct carcinoma (TDCa), however, may be clinically indistinguishable from a benign thyroglossal duct cyst. A preoperative radiologic diagnosis can change the type of surgery that is performed and provide prognostic information. We evaluate the US appearance of thyroglossal duct carcinoma and identified the features that distinguish it from benign thyroglossal duct cysts.
Fig. 1: Course of the thyroglossal duct: 60% are located between the hyoid bone and the thyroid cartilage, 13% in the supra-sternal region, 24% above the hyoid bone including the submental site and 2% intra-lingual

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**Fig. 2:** TDCs with thick wall and hypoechoic content, suggesting infection.

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Methods and Materials

Retrospective analysis of 77 patients who underwent surgery to remove thyroglossal cyst, and had preoperative ultrasonography, from November 1998 to September 2010. To confirm the cyst nature, ultrasound findings were related to pathological. No attempt was made to distinguish between primary thyroglossal duct tumors and metastatic disease from tumor in the thyroid gland.

All diagnostic ultrasound examinations and fine-needle aspiration biopsy (FNAB) were performed with a top-level commercially available US machines: Logiq 500 / 700 (GE, Milwaukee, Wisconsin, USA), IU 22 (Philips Medical Systems; Bothell, WA, EUA) and Aplio 500 (Toshiba Medical Systems Corp.; Otawara, Japan) by a group of four cytologists under ultrasound guidance by one of five radiologists.

The patients were scanned supine with their necks hyperextended on a pillow. All scans were performed with a 7.5- or 10-MHz linear transducer, and images were obtained in the transverse and longitudinal planes. The sonograms were evaluated for the following features: site of the mass, size, walls, margins, loculation, internal echogenicity, posterior enhancement, internal septa, solid component and presence or absence of the thyroid.

The lesion to be biopsied was localized and a mark was placed on the patient's skin, the area was sterilized by using alcohol solution (70%). Local anesthesia was instilled, the transducer was placed in a sterile probe cover, and three passes were made with a freehand technique under direct US guidance by using a 25-gauge needle attached to a 10-mL syringe. The syringe was placed in a hand-operated aspiration biopsy syringe gun (Inrad, Kentwood, Mich) and suction was applied.

The specimen from each pass was extruded onto a glass slide and smeared with a second slide in the conventional manner. One slide of each pair was air dried; the second slice was placed immediately in 95% ethanol fixative. The needle and syringe from each pass were washed in 95% formol and combined to prepare paraffin-embedded cellblock sections for cytopathologic evaluation and immunostaining as needed.

Sensitivity, specificity, positive and negative predictive values of FNAB and US were obtained for characterization of TDCa; we considered as reference standard the pathological confirmation.

The study was approved by the Local Ethics Committee.
Results

Out of 77 cases of thyroglossal duct cyst, six (7.8%) were proven pathologically to be carcinoma (fig. 3). All malignant lesions were papillary carcinoma.

In the TDCa group, there were three female and three male patients. The average age of these patients was 46 years (age range, 18-66 years). The average age of the patients with benign TDC was 36 years (age range, 10-73 years; 26 male and 44 female patients). (Table 1)

All cases of carcinoma had a midline anterior cervical cystic mass with parietal solid component. The cystic area showed thick content with debris and septa in 83% and 17% of anechoic content. In the solid part, microcalcification was present in 33% and all showed vascularization (fig 4), being that in 50% the rate of resistivity was # 0.8 (fig 5). Benign cyst showed no vegetation in 97%, presenting cystic area ranging from thick content (52%), anechoic (32%) and debris-septa (16%).

Concerning the volume, malignant was larger and averaged 29.6 mL (volume range, 11.7-73.9 mL), while benign average was 4.8 mL (volume range, 0.1-302.2 mL). The same occurred to the parietal vegetation, which had an average of 12.1 mL in carcinoma (range 1.5-19.9 mL) and 0.001 mL in benign cyst, which shouldn't have been considered as a vegetation, but just a wall focal thickening or thin septa.

<table>
<thead>
<tr>
<th></th>
<th>TDCs</th>
<th>TDCa</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sex (male x female)</td>
<td>3 x 5</td>
<td>1 x 1</td>
</tr>
<tr>
<td>Age (average)</td>
<td>36 years</td>
<td>46 years</td>
</tr>
<tr>
<td>Volume (average)</td>
<td>4.8 mL</td>
<td>29.6 mL</td>
</tr>
<tr>
<td>Parietal vegetation</td>
<td>None</td>
<td>100 %</td>
</tr>
</tbody>
</table>

Table 1

Fine-needle aspiration biopsy (FNAB) of the thyroglossal duct cyst revealed papillary carcinoma in two of the six patients (33%). Thyroidectomy was performed in five patients, and in two (40%) of these patients, a focus of papillary carcinoma was discovered in the thyroid isthmus.

Long- term clinical follow-up was unavailable for all six patients.
**Fig. 3:** TDCs: 92,2 % TDCa: 7,8%

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**Fig. 4:** Color Doppler shows the presence of papillary projections in a TDCa with vascularization inside.

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Fig. 5: Spectral analysis shows pulsatile arterial flow in a papillary projection, characterized by high-resistance (RI # 0.8)

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Conclusion

A mass in the neck is a common clinical finding and differential diagnosis may be extremely broad. Although most masses are due to benign processes, malignant diseases must not be overlooked. Therefore, it is important to develop a systematic approach for the diagnosis and management of neck masses [5].

Benign thyroglossal duct cysts usually present as asymptomatic, soft, firm, or hard masses in the midline of the anterior neck and generally movable. Malignant thyroglossal duct cysts present in the same way. Carcinoma should be suspected in any thyroglossal duct cyst that is hard, fixed and irregular, or which has undergone recent change. A history of irradiation of the head and neck or mediastinum during childhood or adolescence should also arouse suspicion of carcinoma [5].

Malignant tumors developing from the thyroglossal duct have two origins: thy rogenic carcinoma arising from thyroembrionic remnants in the duct or a cyst, and squamous cell carcinoma arising from metaplastic columnar cells that line the duct. More than 200 cases of thyroglossal duct carcinomas have been reported in which papillary carcinoma accounts for 80% of cases and 20% squamous cell carcinoma. Only one case with both concomitant histologic findings has been reported [5]. We found just papillary carcinoma, probably due to the lower number of patients in our study.

Multicentricity and multifocal growth are common in papillary TDCa [6], the number of reported incidences of primary thyroid carcinoma, concomitant with TDCa is between 11 and 33% [7]. Conservative management of papillary thyroid cancer, is indicated only in low-risk patients (female, younger than 40 years with no invasion of the capsule) and a low-grade tumor of less than 1 cm in size [6].

In our study we had higher numbers in terms of carcinomas proportion between the cysts, as concomitant thyroid papillary carcinoma compared to other studies. Probably because our patients were selected retrospectively from surgical treatment, therefore they were more complicated cases.

The main difficulty encountered with a cancer evolving from a thyroglossal duct cyst is that the diagnosis is usually made during surgery or from definitive pathological samples. Because the frequency of cancer of the thyroglossal duct cyst is very low, the clinician often does not consider an oncologic diagnosis. A second difficulty lies in terms of what approach should be taken during and after surgery when dealing with a preoperatively diagnosed thyroglossal cyst; that is, how extensive should the surgery be and what type of adjuvant therapy should be used. [5]

Advances in imaging techniques such as ultrasonography are used in the confirmation of TDCa. Ultrasonography can also be carried out concurrently with FNAB [8,9] showing high sensitivity and specificity when performed by well-trained professionals. Our low
sensitivity value of FNAB was probably due to the wrong technic procedure. In case of TDCa suspicion based on our results (parietal vegetation, thick septa, high vascularity), we must guarantee that the needle will reach the solid part. For that reason, the cystic part must be drained before, thus becomes easier to achieve the parietal vegetation. Although we found only 33% of calcification (fig 6/7) in TDCa, we believe as other authors, that this is an important feature and should be taken into consideration.

In some occasions as infection and bleeding, the TDCs may present a pseudo-solid appearance due to the proteinaceous content of the fluid (fig 8). The uniform echogenicity may lead to an erroneous assumption that the lesion is solid, especially when posterior enhancement is absent. To prevent false positive findings, pressure with the transducer must be applied on the cyst, so the entire content shift, suggesting its cystic nature.

The surgical treatment for benign thyroglossal duct cysts is the Sistrunk procedure, in which the cyst, the central portion of the hyoid bone, and the entire thyroglossal tract are removed. The surgical treatment of thyroglossal duct carcinoma is controversial. Although some authors consider the Sistrunk procedure alone to be sufficient, recent reports recommend total thyroidectomy in addition to the Sistrunk procedure. In patients with abnormal cervical lymph nodes by clinical or radiographic criteria, the Sistrunk procedure combined with thyroidectomy and lymphadenectomy is widely accepted [10].

There are two goals for preoperative diagnostic testing: documenting a normal thyroid gland, and determining whether the Sistrunk procedure is sufficient surgical treatment. Documenting a normal thyroid gland is important because an ectopic thyroid gland may mimic a thyroglossal duct cyst clinically. If this tissue is removed, the patient will be rendered hypothyroid. Should the preoperative evaluation reveal thyroid abnormalities or cervical lymphadenopathy, the surgical approach may be altered [10]. High-resolution sonography remains the ideal initial investigation because it is easily available, inexpensive, and provides the surgeon with necessary preoperative information.

To conclude:

1. Carcinoma should be suspected in the cyst with thick septa and parietal vegetation.
2. Doppler can help find and characterize the parietal vegetation, and this generally associated with high rate of resistivity.
3. The main appearance of the benign cyst is a thick content or anechoic, without vegetation.
4. Preoperative diagnosis of carcinoma can have important implications for surgical planning and postoperative treatment.
Fig. 6: Conventional ultrasonography show the presence of papillary projections, solid components and calcification, suggesting TDCa

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Fig. 7: Conventional ultrasonography show the presence of papillary projections, solid components and calcification, suggesting TDCa

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Fig. 8: Pseudo-solid appearance due to the proteinaceous content of a bleeding fluid

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References


