



## Neoplastic Degeneration of a Thyroglossal Duct Cyst: Case Report

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### Abstract

The thyroglossal duct cyst is a congenital lesion of the neck, resulting from the incomplete obliteration of the thyroglossal canal; in the embryonic period, its purpose is to facilitate the migration to the final seat of the thyroid duct. Rarely, the retention of this cyst can cause a neoplastic degeneration. A 33-year-old female presented to our outpatient clinic complaining of the appearance, about a year ago, a swelling of the neck in the middle of the suprahyoid region. Clinical and medical history characteristics were compatible with a thyroglossal duct cyst. Computed Tomography (CT) and Magnetic Resonance Imaging (MRI) showed the presence of a thyroglossal duct cyst. The patient was subjected to surgery according Sistrunk procedure, and the diagnosis was papillary carcinoma based on histopathologic examination. After two years of follow-up, the patient is currently disease-free. This confirms the validity of the Sistrunk surgery for non-extended and well-differentiated tumors, with no associated thyroid lesions.

**Keywords:** Thyroglossal duct cyst; Papillary carcinoma; Sistrunk

### Introduction

The thyroglossal duct cyst (TGDC), together with gill cysts, falls within scope of cervical disease of embryonic origin. There are two theories about the pathogenesis of neoplastic degeneration: 1) the “*novo* theory”, the most accepted, which argues that the tumor would originate from thyroid gland ectopic tissue; 2) the metastatic theory, according to which the cancer arose in a cyst and represents tissue of a primitive metastatic thyroid carcinoma [1,2]. Also as is known, inflammatory episodes applicants help the tissue hyperplasia, which however can rarely lead to neoplastic degeneration. Clinically it is not possible to differentiate a benign cyst from a degenerate cyst in malignancy and the diagnosis is generally post-operative. Primary malignancy in TGDC is extremely rare and among all cases of TGDC, carcinoma is reported only in 0.7–1% of cases [3,4]. Papillary carcinoma (80–85 %) is the most common carcinoma reported in TGDC, followed by squamous cell carcinoma (5–6 %) [5].

Here, we report a case of 33-year-old-female presented with a midline neck mass, which was diagnosed and operated for TGDC. However, on final histopathology, a diagnosis of papillary carcinoma arising in a TGDC was made.

### Case Presentation

A 33-year-old female, smoker of 5 cigarettes per day, presented to our outpatient clinic complaining of the appearance, about a year ago, a swelling of the neck in the middle of the suprahyoid region. There was no history of pain, dysphagia, or hoarseness of voice. There was no history of cold or hot intolerance, tremor, palpitation, diarrhea, constipation, any menstrual irregularities. The patient had no family history of metabolic abnormalities and no comorbidities. She denied any radiation exposure or family history of thyroid cancer. The ear, nose and throat examination identified a neck mass, which moved during deglutition. The swelling appeared round with an approximate diameter of 2 cm, not skin hyperemia; not painful, superficial and deep mobile plans, with hard-elastic consistency, no-palpable cervical lymphnodes.

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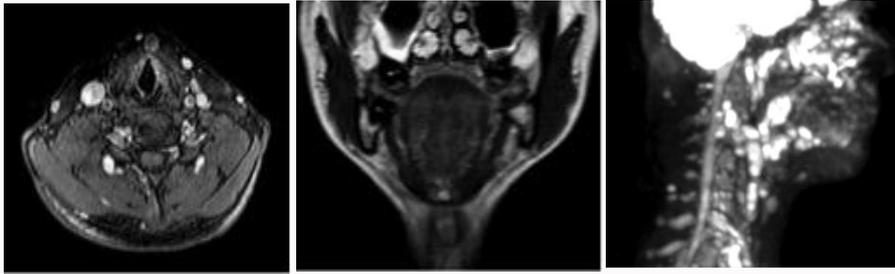


Figure 1: Preoperative MRI.

The routine laboratory investigations including complete blood count, liver function test, and kidney function test was within normal limit. Determination of blood levels of the thyroid hormones showed normal free triiodothyronine (FT3), free tetraiodothyronine (FT4), and TSH level.

The ultrasonography (USG) of the neck to confirm the presence of a lesion in the middle of suprahyoid region, cystic appearance, reactive lymph nodes and normal thyroid. Computed Tomography (CT) of the neck with contrast and Magnetic Resonance Imaging (MRI) showed the presence of a thyroglossal duct cyst (Figure 1 and 2).

The patient was subjected to surgery according to Sistrunk procedure [6]. The resected specimen sent for histopathological examination. Final pathology showed a papillary thyroid carcinoma in the context of a thyroglossal duct cyst (Figure 3). All of the margins were clear.

The patient was discharged three days after surgery. Based on the histopathology, it was decided to perform a close follow-up (every 3 months). After two years of follow-up, the patient is currently disease-free. Repeat USG of neck showed normal thyroid scan.

## Discussion

TGDCs are the most common congenital anomaly of the thyroid, usually manifested as enlarging, painless midline neck mass. This is the most common cause of midline neck mass in children (75%), but this is reported only in 7% of adult population. In the majority of the cases, TGDCs are benign [7]. However, malignancy is reported in around 1% of cases and is usually diagnosed as an incidental finding after histopathological evaluation of resected TGDC [8]. Thyrogenic carcinoma is the most common malignant tumor of TGDC, which originates from thyroglossal tract remnant followed by squamous cell carcinoma which originates from metaplastic columnar cells of the thyroglossal duct lining [9].

In literature, there are about 250 cases of cancer of the thyroglossal duct cyst degeneration, variously localized on the same route of the duct, from the cecum foramen to the intrathyroidal level; the main location (75%) is in subhyoid, of which only six cases are intrathyroidal, while those at the suprahyoid region, like in our case, are a rarity [10]. The average age of diagnosis of neoplastic degeneration of the thyroglossal duct cyst is about 50 years, with an M/F ratio of 1/2. In a percentage from 11.4 to 33% of cases, it coexists with cancer in the thyroid parenchyma. Histological variants of cancer of thyroglossal duct cysts are: 80% papillary carcinoma, 8% mixed papillary-follicular, 6% squamous cell carcinoma, and rarely follicular forms as well, at Hurtle cell, insular or anaplastic [11].

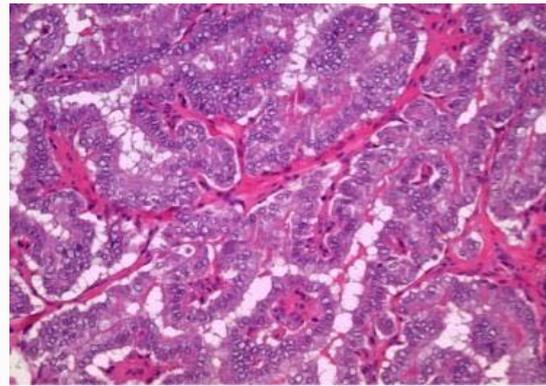


Figure 2: Histological relief: Microphotograph showing papillary projections arising from cyst wall (h and E, x 100).

In most cases, the presence of a swelling in the middle of the neck is the only clinical sign, and the combination with an infectious/inflammatory phenomenon causes skin hyperemia, spontaneous pain or cervical lymphadenopathy triggered by palpation and consensual. Symptomatology, both in cases of benign disease and malignant degeneration, is similar and it depends by the size of the cyst.

Symptoms may occur with dysphagia, dysphonia, cutaneous fistulas and rapid increase in volume.

Diagnostic methods are represented by ultrasound, computed tomography and MRI. There are no diagnostic and therapeutic protocols able to identify the cancer development, as in most of the cases diagnosis is casual [12]. The therapy is surgical and the technique for excision of the cyst is represented by Sistrunk procedure: originally described by Charles Sistrunk, the key to the procedure and prevention of recurrence of the cyst is to resect the central portion (body) of the hyoid bone and follow any tract up towards the base of the tongue, the embryological site of origin of the thyroid gland [13]. Remains debated the role of a partial or total thyroidectomy and radioiodine therapy in the cases with suprahyoid location.

Our case report has demonstrated the validity of Sistrunk procedure in follow-up, because we did not detect relapse. Furthermore, Patel and colleagues have revisited 62 clinical cases of well-differentiated thyroid cancer arising from thyroglossal duct cysts. These authors observed that total thyroidectomy and the radioiodine therapy did not further improve clinical outcomes [14]. Therefore, the Sistrunk procedure may be considered valid in the presence of a well-differentiated carcinoma confined inside of a thyroglossal duct cyst. Vigneri and coll, after analyzing studies of the last 25 years, have confirmed the validity of Sistrunk intervention in the cases without associated risk factors [15]. Therefore, a conservative surgical

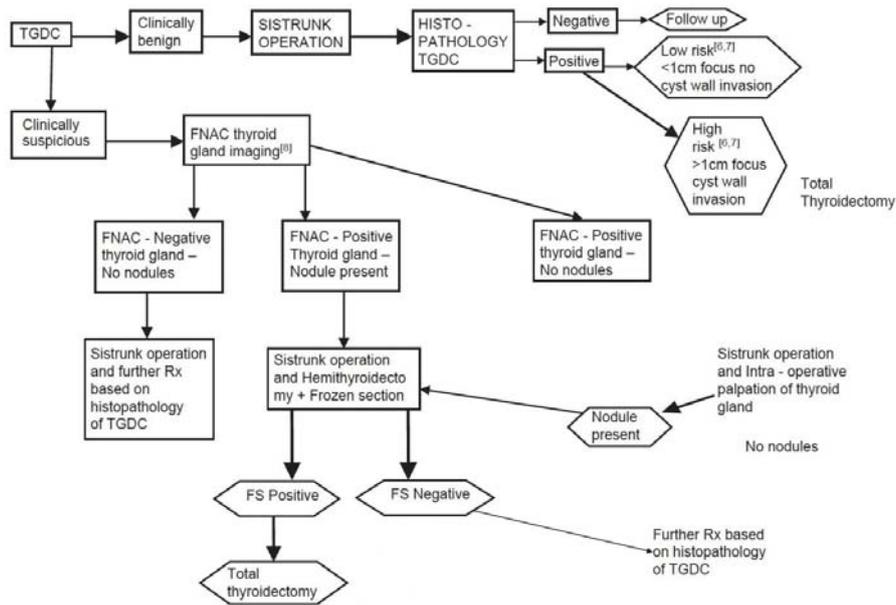


Figure 3: Treatment algorithm.

management is indicated in low-risk patients (female, age< 40 years, no capsular invasion, sizes of less than 1 cm in diameter tumor), while a more aggressive surgery is to be reserved for those patients with thyroid lesions associated, clinically obvious or apparent at follow-up.

Senthil kumar et al. [16] have proposed a treatment algorithm on the basis of the literature evidence in cases of cancer of thyroglossal cysts, which shows several therapeutic approaches for the clinically benign cysts with subsequent detection of malignancy on histologic examination (Figure 4) [16]. In the most suspicious cases, we carry out additional pre-operative investigations, such as fine needle aspiration cytology (FNAC), without differentiating between cysts or infrahoid.

Finally, from a careful analysis of the literature, we found that actually the phenomenon (cancer arising in a thyroglossal duct cyst) may be less rare than reported in the incidence statistics published until now: in the first 6 months of 2016 have already been published five case reports on the topic and probably there were other unpublished cases [17-21].

**Conclusion**

Based on our experience and in the absence of international guidelines about this disease, we believe that the Sistrunk procedure is more effective rather than others surgical interventions for primary tumors that occur in the context of thyroglossal cysts, without associated risk factors. Based on the increasing incidence of cancers arising from thyroglossal duct cyst, probably we should always take into account this possibility when it is diagnosed TGDC.

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