

THYROGLOSSAL DUCT CYST

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THYROGLOSSAL DUCT CYST (Abstract): The thyroglossal duct cyst (TDC) results from a failure in obliterating the embryogenic duct produced during thyroid migration and it represents the most common type of developmental cyst encountered in the neck region. Ectopic thyroid tissue neoplasias are rare, and even rarer when associated with the TDC. Methods: During the period 1998-2008, in the First Surgical Clinic, University Hospital “St. Spiridon” Iasi, 14 patients with thyroglossal duct cyst were diagnosed and treated. All records were reviewed for age and sex, diagnostic methods, sizes, surgical management and recurrences. Results: All patients with thyroglossal duct cysts are described as midline cysts of the neck. The ratio females/males was 6/1 with mean age 37.6 years (13-60 years). One case was with an external fistula. The treatment performed was a variant of Sistrunk’s procedure in which the thyroglossal tract was excised to a variable extent, but in all cases with central hyoidectomy. The size of the cyst ranged from 1.2 to 4 cm (mean 2.6 cm). Postoperative course was uneventful in all cases. No recurrence was recorded in this series. We describe a case, a 19 years old female with thyroid papillary carcinoma evolving from a TDC. The literature is reviewed. Conclusion: The standard surgical approach to TDC is Sistrunk’s operation with low recurrence rates. Malignancy within a thyroglossal duct cyst is very rare but should be included in the differential diagnosis of a neck mass. In such cases total thyroidectomy with removal of the tumour of thyroglossal duct and the body of the hyoid bone are recommended – because the carcinoma may be multifocal and because a lymphatic invasion of the thyroid may take place – in order to ensure a correct follow-up.

KEY WORDS: THYROGLOSSAL DUCT CYST, SISTRUNK'S OPERATION, THYROID PAPILLARY CARCINOMA

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INTRODUCTION

The thyroglossal duct cyst (TDC) is a well recognized developmental abnormality which arises in some 7% of the population. Consequently, it represents the most common type of developmental cyst encountered in the neck region. TDC results from a failure in obliterating the embryogenic duct produced during thyroid migration. TDCs are most often diagnosed in the pediatric age group and a minority of patients with TDCs are over 20 years of age at the time of diagnosis. A cyst may develop from the secretory residual epithelium. Ectopic thyroid tissue neoplasias are rare, and even rarer when associated with the TDC. It has been reported that over 62% of them may have some ectopic thyroid tissue. It typically presents itself as a mobile, painless mass in the anterior midline of the neck, usually in close proximity to the hyoid bone.

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METHODS

During the period 1998-2008, in First Surgical Clinic, „St. Spiridon” Hospital of Iași, 14 patients with thyroglossal duct cyst were diagnosed and treated. 1902 thyroidectomies for thyroid disease were performed during this period. All records were reviewed for age and sex, diagnostic methods, sizes, surgical management and recurrences. All patients underwent clinical examination and screening laboratory tests, cervical and chest radiography, ultrasound scan, scintigraphy with ¹³¹I and thyroid function tests.

RESULTS

This TDC group consisted of 11 females (78.6%) and 3 males (21.4%) with mean age 37.6 years (13-60 years). The mean age of onset of symptoms was 24 years. The history of the disease varied from 1 month to 35 years. 13 patients with thyroglossal duct cysts are described as midline cysts of the neck. In this series 1 cyst was situated laterally.

The level of the cysts in the neck was as follows: two in the suprahyoid region, 9 in the prehyoid region, three at the level of the thyroid cartilage. In this series no cysts were seen in the chest.

A case with an external fistula with recurrence of 10 years after cyst removal by a variant of Sistrunk procedure is reported.



Fig.1 Thyroglossal duct cyst in the midline of neck

The commonest reason for seeking treatment was the discovery of an asymptomatic lump in the anterior part of the neck in 10 cases. Thyroid pathology (goiter) was associated in two cases. A tender inflamed lump and external fistula were noticed in another case. The clinical diagnosis was suggested by the presence of a cystic mass situated towards the front of the midline neck, which moved with protrusion of the tongue and could be transilluminated.

In this series all patients were operated on. One case was initially treated with antibiotics for infected thyroglossal cysts. The treatment performed was a variant of Sistrunk's procedure in which the thyroglossal tract was excised to a variable extent, but in all cases with central hyoidectomy.

The original 1920 monograph by an american surgeon, Walter Ellis Sistrunk from the Mayo Clinic, described the surgical management of this congenital abnormality and remains the classic basic reference. The „Sistrunk operation” consists

en bloc cystectomy and central hyoidectomy, with tract excision up to the foramen cecum.

The size of the cyst ranged from 1.5 to 6 cm (mean 2.8 cm). Postoperative course was uneventful in all cases. In this series no recurrence was recorded. Three of the patients were not followed up.

In one case the histopathologic diagnosis was a papillary carcinoma evolving from a TDC. We describe this case in view of its extreme rarity and in order to highlight the therapeutic options in the management of malignancies arising in thyroglossal cysts. A 19 years old female noticed a painless mass in the midline of her neck, one month prior to her first visit to our clinic (Fig. 1).

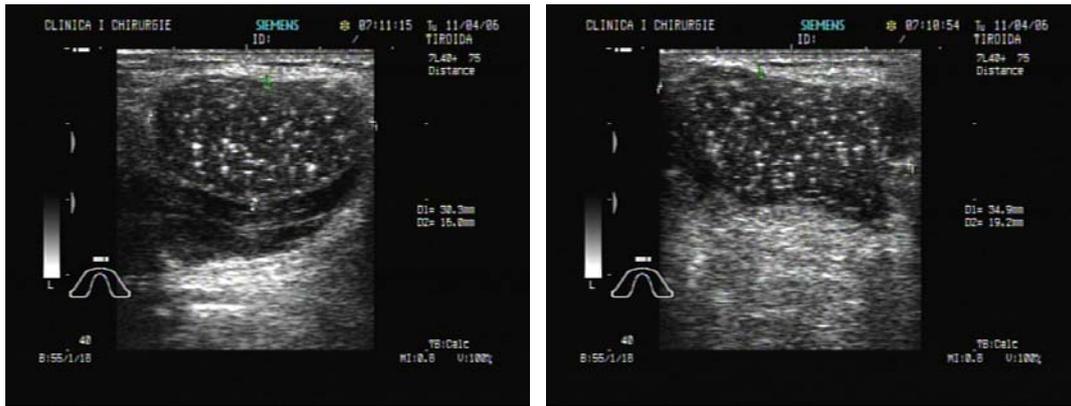


Fig.2 Ultrasound scan revealed a cystic mass, with a thick content

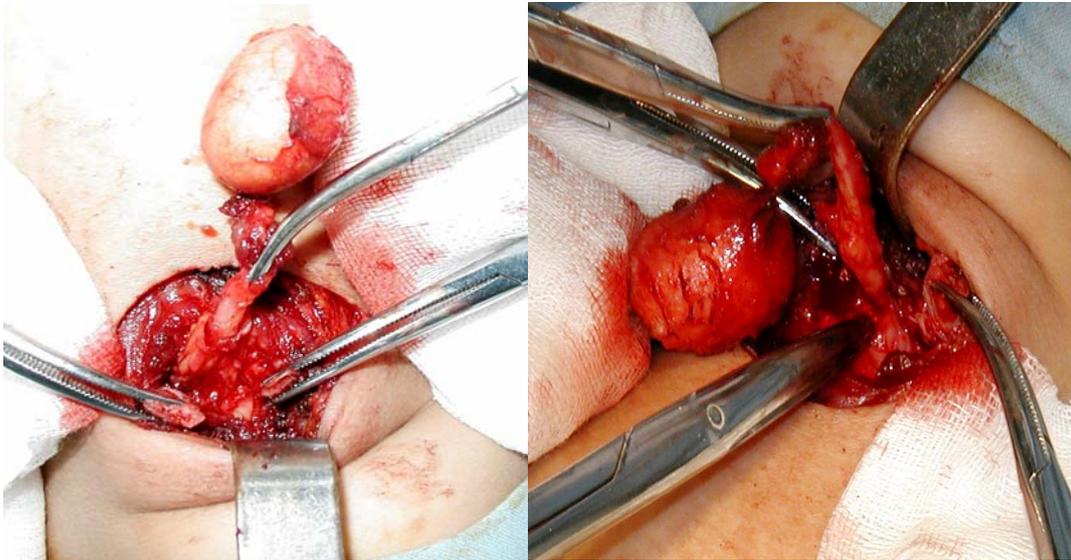


Fig. 3 Excision of thyroglossal duct cyst with a well defined smooth surface accompanied by the suprahyoid tract.

She had no past history of neckinjury. She did not complain of dysphagia or odynophagia, or of phlogistic signs. During the clinical examination, we noticed a deep nodular lesion, of about 3x2 cm. It was fibroelastic, mobile during swallowing and at tongue protrusion. There were no clinically significant nodes. The thyroid gland was

normal at palpation. Ultrasound scan showed a cystic mass, with a thick content; the thyroid was within normal limits (Fig. 2).

The thyroid scintigraphy was normal. We performed a Sistrunk's procedure (removal of the mid-portion of the hyoid bone in continuity with the TDC and excision at the core of tissue between the hyoid bone and the foramen cecum (Fig. 3).

The histopathologic exam revealed a papillary carcinoma *in situ* evolving from a TDC (Fig. 4). We didn't performed any other thyroidian procedure because the clinical exam as well as thyroidian ultrasonography and scintigraphy didn't revealed any lesion. The patient has been followed for seven years. No metastasis occurred.

DISCUSSION

Embryologically, by the seventh or eighth week of development, the thyroid reaches its normal position, the area below the thyroid cartilage, descending through the thyroglossal duct (TDC). During the 10th week of fetal life, the TDC is usually obliterated. Failure of obliteration may result in the development of a cystic dilatation at any time in life. The body of the hyoid bone subsequently develops in the mesoderm joining the ventral ends of the second and third branchial arches and may incorporate the thyroglossal tract into its substance. TDCs are localized towards the midline, between the base of the tongue and the pyramidal lobe of the thyroid gland.

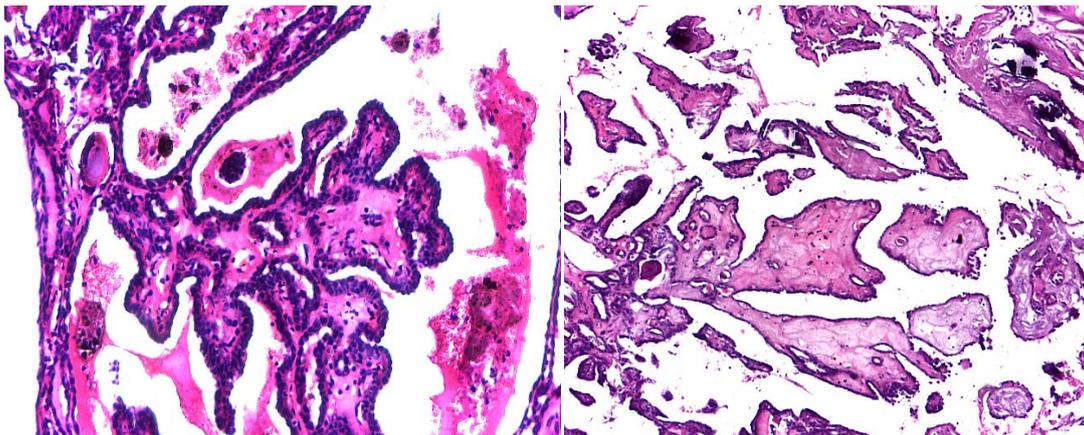


Fig.4 Histopathologic exam – papillary carcinoma in TDC (hematoxylin-eosine ob. 10x)

TDCs are present in approximately 7% of the general population; up to 62% of these may contain ectopic and functional thyroid tissue, thereby enabling the development of thyroid-related tumours. 70% are diagnosed in childhood and 7% are diagnosed in adulthood. Malignant neoplasms rarely arise in thyroglossal cysts, in less than 1% of cases [1]. They usually take the form of either papillary carcinoma of thyroid origin (arising from thyroembryonic remnants in the duct or a cyst: 85-95% of cases), squamous carcinoma (arising from metaplastic columnar cells that line the duct: 5% of cases), or anaplastic, and Hurthle cell carcinoma. Criteria for the diagnosis of primary papillary carcinoma arising in a TDC after Widstrom are: 1) histologic identification of TDC demonstrating that the cyst or duct has an epithelial lining with normal thyroid follicles in the cyst wall; 2) there is normal thyroid tissue adjacent to the tumor; and 3) histopathologic examination of the thyroid gland reveals no signs of

primary carcinoma [2]. In this paper we describe a case with primary papillary carcinoma.

As TDCs are most often diagnosed in the pediatric age group, only a minority of cases with TDCs, are operated on at an adult age.

The most common clinical sign was a non-tender, mobile neck mass, which was painful at swallowing in the anterior midline of the neck, usually in close proximity to the hyoid bone. Less often, TDCs may show signs and symptoms of secondary infection or present evidence of a fistula. The cystic mass is situated anteriorly towards the midline of the neck, which moved with tongue protrusion and could be transilluminated. In rare cases the cysts are situated laterally [3]. Hatada T et al report a case with intrathyroid thyroglossal duct cyst [4].

Preoperative evaluation of thyroglossal duct cyst includes: cervical and chest radiography, ultrasound scan, scintigraphy with ^{131}I and thyroid function tests. An ultrasound exam is useful in malignancy by demonstrating a mural nodule, calcification or lymph node metastases. Computerized tomography has also been used in case of malignancy and ectopic cysts. Clinical confirmation of aberrant thyroid tissue is proved by a radioactive iodine scan. The ultrasound-guided fine-needle aspiration (FNA) is only moderately sensitive for a preoperative evaluation of TDCs. Cytomorphologic features are not always specific, but associated with clinical and radiological signs, they may be helpful for an accurate diagnosis. Thyroid epithelium is rarely identified [5].



Fig. 5 Autoportret Piero della Francesca in Polyptych of the Misericordia

The differential diagnosis involves: dermoid cyst, epidermoid cyst, branchial cleft cyst, lymph nodes, lymphangioma, thyroid pathology. The thyroglossal duct cyst raised many differential diagnostic problems obvious even in the Renaissance paintings (Fig. 5). Thus H.E. Emson contradicts L. Bondenson stating that Piero della Francesca's self portrait from the Polyptych of the Misericordia betrays a thyroglossal duct cyst and not goiter [6].

The standard surgical approach to TDCs dating back to early 20th century is Sistrunk's operation encompassing removal of the mid-portion of the hyoid bone in continuity with the TDC and excision of a core of tissue between the hyoid bone and the

foramen cecum [7]. The Sistrunk procedure is recommended as the main operation of choice, especially in adults in whom a more extended tract resection should be performed [8].

The risk of recurrence is high in case of inadequate tissue resection from the tongue base with such multiple tracts. In our series we performed a complete thyroglossal tract in one case with recurrence, 10 years after the simple cyst removal.

The alternative solution for surgical treatment is percutaneous ethanol injection. If the presence of a malignant lesion can be excluded, percutaneous ethanol injection may be considered a secondary treatment in patients with thyroglossal duct cysts in selected cases [9].

Primary carcinoma is rare and is seen in less than 1% of cases [10,11]. Carcinoma arises slightly more often in females in the fourth decade of life. The malignancy should be suspected by rapid increase in size, dysphagia, hoarseness of voice and pain. The thyroglossal duct cyst is hard, fixed and irregular. The diagnosis is usually based on pathological examination of the cyst. Approximately 200 cases have been reported in the literature worldwide, diagnosed with papillary carcinoma arising from the thyroglossal duct. The tall cell variant is a rare papillary carcinoma and has a poor prognosis [12].

Following the Sistrunk's procedure, when a histopathological examination reveals a malignancy, the thyroid gland must be studied radiologically and scintigraphically. Sistrunk's procedure would suffice if the thyroid gland was found to be normal. Total thyroidectomy is recommended in selected cases, as a papillary carcinoma, may be present [13]. Cervical bilateral node dissection is not necessary in all cases, only in 8% of cases a significant involvement of regional lymph nodes was present [14]. In our case, we used a radical surgical method with total thyroidectomy. For all patients with papillary carcinoma of thyroglossal cyst, radioactive iodine and thyroid suppression are recommended, if the patient had or not a thyroidectomy or the thyroid scan is normal [1,15].

Patel et al made a retrospective study and analysed the prognostic factors predictive of overall survival in patients with TDCs, and revealed that the only significant predictor of outcome was the extent of surgery for TDCs. Patients who had simple excision have had a 10-year overall survival rates of 75%, in comparison with 100% in the patient who suffered Sistrunk's procedure [16]. The patients with squamous carcinoma arising in a thyroglossal cyst have a poorer prognosis and should be treated with postoperative external beam radiotherapy [10].

CONCLUSION

TDC results from a failure in obliterating the embryogenic duct produced during thyroid migration. The cyst usually appears as an asymptomatic swelling in the prehyoid region of the neck. The standard surgical approach to TDC is Sistrunk's operation with low recurrence rates.

Malignancy within a thyroglossal duct cyst is very rare but should be included in the differential diagnosis of a neck mass. The therapy includes surgery, radioactive iodine and thyroid suppression, as is the case with differentiated thyroid cancers. In such cases total thyroidectomy with removal of the tumour of thyroglossal duct and the body of the hyoid bone are recommended because the carcinoma may be multifocal and because a lymphatic invasion of the thyroid may take place in order to ensure a correct follow-up.

REFERENCES

1. Kandogan T, Erkan N, Vardar E. Papillary carcinoma arising in a thyroglossal duct cyst with associated microcarcinoma of the thyroid and without cervical lymph node metastasis: a case report. *J Med Case Reports*. 2008; 2: 42.
2. Widstrom A, Magnusson P, Hallberg O, Hellqvist H, Riiber H. Adenocarcinoma originating in the thyroglossal duct. *Ann Otol*. 1976; 85: 286–290.
3. Mohan PS, Chokshi RA, Moser RL, Razvi SA Thyroglossal duct cysts: a consideration in adults. *Am Surg*. 2005; 71(6): 508-511.
4. Hatada T, Ichii S, Sagayama K, Ishii H, Sugihara A, Terada N, Yamamura T. Intrathyroid thyroglossal duct cyst simulating a thyroid nodule. *Tumori*. 2000; 86(3): 250-252.
5. Cignarelli M, Ambrosi A, Marino A, Lamacchia O, Cincione R, Neri V. Three cases of papillary carcinoma and three of adenoma in thyroglossal duct cysts: clinical – diagnostic comparison with benign thyroglossal duct cysts. *J Endocrinol Invest*. 2002; 25: 947-954.
6. Emson HE. Thyroid swellings in Renaissance art. *J R Soc Med*. 2004; 97: 311.
7. Sistrunk WE. The surgical treatment of cysts of the thyroglossal tract. *Ann Surg*. 1920; 71: 121-124.
8. Lin ST, Tseng FY, Hsu CJ, Yeh TH, Chen YS. Thyroglossal duct cyst: a comparison between children and adults. *Am J Otolaryngol*. 2008; 29(2): 83-87.
9. Baskin HJ. Percutaneous ethanol injection of thyroglossal duct cysts. *Endocr Pract*. 2006; 12(4): 355-357.
10. Vijay R, Rajan KK, Feroze M. Inapparent twin malignancy in thyroglossal cyst: case report. *World J Surg Oncol*. 2003; 1(1): 15.
11. Yang YJ, Haghiri S, Wanamaker JR, Powers CN. Diagnosis of papillary carcinoma in a thyroglossal duct cyst by fine-needle aspiration biopsy. *Arch Pathol Lab Med*. 2000; 124(1): 139-142.
12. Köybaşıoğlu F, Simşek GG, Onal BU. Tall cell variant of papillary carcinoma arising from a thyroglossal cyst: report of a case with diagnosis by fine needle aspiration cytology. *Acta Cytol*. 2006; 50(2): 221-224.
13. Plaza CP, López ME, Carrasco CE, Meseguer LM, Perucho Ade L. Management of well-differentiated thyroglossal remnant thyroid carcinoma: time to close the debate? Report of five new cases and proposal of a definitive algorithm for treatment. *Ann Surg Oncol*. 2006; 13(5): 745-752.
14. Renard TH, Choucair RJ, Stevenson WD, Brooks WC and Poulos E: Carcinoma of the thyroglossal duct. *Surg Gynecol Obstet*. 1990; 171: 305-308.
15. Maziak D, Borowy ZJ, Deitel M, Jaksic T and Ralph-Edwards A. Management of papillary carcinoma arising in thyroglossal duct anlage. *Can J Surg*. 1992, 35: 522-525.
16. Patel SG, Escrig M, Shaha AR, Singh B, Shah JP. Management of well-differentiated thyroid carcinoma presenting within a thyroglossal duct cyst. *J Surg Oncol* 2002; 79: 134–139.