Peertechz

Medical Group

Global Journal of Rare Diseases

DOI CC By

Sadia Ashraf*, Prashant Nadkarni, Roberto E Izquierdo and Nidhi Bansal

Department of Endocrinology, SUNY Upstate Medical University, Syracuse, NY, USA

Dates: Received: 22 November, 2016; **Accepted:** 10 December, 2016; **Published:** 12 December, 2016

***Corresponding author:** Sadia Ashraf, MBBS, SUNY Upstate Medical University, Syracuse, NY, 13210, USA, E mail: fakhar_ud_din@yahoo.com.

Keywords: Thyroid; Cancer; Papillary thyroid cancer; Thyroglossal duct cyst

https://www.peertechz.com

Case Report

Dilemma in Managing Thyroglossal Duct Cyst Carcinoma

Abstract

The thyroglossal duct cyst (TGDC) is a well-known developmental abnormality encountered in the neck. It represents over 75% of childhood midline neck masses and 7% of the adult population. It typically presents as a mobile, painless mass in the anterior midline of the neck, usually in close junction to the hyoid bone. TGDCs infrequently present with signs and symptoms of secondary infection, or with evidence of a fistula. Malignancy is rarely encountered in TGDCs approximately in 1.5% of patients. We present an interesting and rare case of elderly female who presented with an asymptomatic neck mass. CT scan of neck with contrast showed lobulated vascular enhancing mass 3.1 cm in size with no significant lymphadenopathy. Fine needle aspiration of neck mass revealed papillary carcinoma. She underwent Sistrunk procedure with total thyroidectomy and central neck dissection followed by radioactive iodine (RAI) and thyroid hormone suppression. Since the incidence of TGDC carcinoma is low, controversy exists in its management in relation to type and the extent of surgery and use of RAI.

Introduction

The thyroglossal duct cyst (TGDC) is the most common congenital anomaly of the thyroid. It is usually manifested as painless mobile midline neck mass. TGDC carcinoma was first described by Brentano in 1911 and Uchermann in 1915[1]. Malignancy is very rare and is reported in around 1.5% of cases as an incidental finding [2]. Papillary carcinoma represents almost 80% of the cases and usually characterized by non-aggressive behavior with good prognosis [3]. Most cases of TGDC carcinoma are diagnosed during the third and fourth decade of life and rarely in children under 14 years of age [4]. TGDC is very rare in elderly population above age 60 with incidence of approximately 0.6% [5]. Ducic mentioned there is higher risk of malignancy in these cysts in the elderly [6]. We present a case of a TGDC carcinoma because of its rarity and unusual manifestation.

Case Report

73 year old female was seen in our office for evaluation of an anterior neck mass after her primary physician felt a midline upper neck mass. She had subtotal thyroidectomy done about 30 years ago for an unknown pathology. She also had received radioactive iodine (RAI) with 31 millicuries (mCi) of Iodine-131 (I-131) for hyperthyroidism secondary to toxic multinodular goiter two years prior to current presentation. She was asymptomatic with no hypo-hyperthyroid or compressive symptoms. Neck examination revealed 3 cm firm mass in the middle of neck at the level of hyoid bone and well healed lower neck surgical scar of thyroidectomy. Thyroid function tests (TFTs) were in normal range. Further work up with thyroid ultrasound and CT scan of neck with contrast showed 3 cm heterogonous mass with moderate vascularity and heterogeneous thyroid gland with multiple calcifications. Fine needle aspiration (FNA) of the neck mass was consistent with papillary thyroid carcinoma (PTC). She underwent Sistrunk procedure along with total thyroidectomy and central neck dissection. Pathology of neck mass revealed PTC 3.5 x 2.5 x 2 cm with no vascular invasion with squamous lined areas suggesting being from thyroglossal duct cyst and BRAF positive. Tumor was almost abutting the overlying bone tissue. Both thyroid lobes had normal thyroid tissue and no pathologic lymph glands in central compartment. Patient developed postoperative transient hypoparathyroidism and was treated with calcium and calcitriol along with thyroid hormone suppressive therapy. She received 52 mCi of I-131. On follow up thyroglobulin level was undetectable. She had no evidence of distant metastasis on post treatment whole body scan.

Discussion

The thyroid gland develops at approximately 4th week of gestation and descends from foramen cecum to its permanent location below the hyoid bone and anterior to trachea and thyroid cartilage by the 7th week of embryonic development. It remains connected to the base of the tongue via thyroglossal duct. If this duct subsequently failed to atrophy, it may give

Citation: Ashraf S, Nadkarni P, Izquierdo RE, Bansal N (2016) Dilemma in Managing Thyroglossal Duct Cyst Carcinoma. Glob J Rare Dis 1(1): 001-003.

rise to thyroglossal duct cyst anywhere along the course from foramen cecum at the base of the tongue to the thyroid gland [7].

Fernandez et al. [8], found that 55% of patients with TGDC carcinoma were younger than 40 years of age and approximately 30% were aged between 20 and 30 years. There is slightly more female: male ratio of 1.6: 1 [9]. The size of the mass is variable, but most are in the 1.2–5 cm range [10].

Histologically, papillary adenocarcinoma is the most common form of TGDC carcinomas (80%) with mixed follicular and papillary carcinoma the next common (7%) followed by squamous cell carcinoma (5%) [11]. Different hypothesis have been proposed regarding the origin of the tumor. Carcinoma arising de novo from the thyroid remnant in the cyst as normal thyroid tissue is known to exist in the TGDC in more than half of cases [3,12]. Other theory is metastatic from the occult malignancy in the thyroid gland [12].

TGDC carcinoma may be clinically indistinguishable from a benign TGDC. TFTs and thyroid scans are usually normal. Preoperative radiological imaging like ultrasonography and CT scan along with FNA can detect malignancy and help plan type and extent of surgery [13]. In our patient size and increased vascularity was suggestive of malignancy which was confirmed on FNA of the mass. FNA may confirm the TGDC carcinoma; however, negative FNA does not exclude it. Sensitivity rate of approximately 60% and positive predictive value of 69% have been reported [10].

Management of TGDC carcinoma varies and included Sistrunk procedure (involving excision of the cyst, center part of hyoid bone and a core of tissue around the thyroglossal tract up to the foramen cecum) with thyroid hormone suppression along with strict long term follow up or Sistrunk procedure along with total thyroidectomy followed by RAI and thyroid hormone suppression. Patel et al [14] in their retrospective review of 62 cases concluded that the only significant predictor of overall survival in TGDC carcinoma was the extent of surgery and not the size of tumor. They [14] also mentioned that addition of total thyroidectomy to Sistrunk procedure did not have significant impact on survival especially young population below age 45 who constitutes low risk group. Plaza et al [15] recommended an algorithm for TGDC carcinoma to be treated with Sistrunk procedure without total thyroidectomy in low risk patients. They stratified low risk patients as younger than 45 years, tumor less than 1.5 cm that is confined to the cyst, without distant metastasis and with normal thyroid gland on ultrasonography and no suspicious lymph nodes [15]. Because of the rarity of this disease, and lack of specific guidelines, some authors agreed with following the current guidelines for management of papillary thyroid carcinoma concluding that Sistrunk procedure is sufficient for tumor less than 1 cm [13,16]. For tumor between 1-4 cm, one can consider Sistrunk procedure only or add total thyroidectomy with or without central compartment neck dissection depending on other risk factors like age above 45, radiation exposure, soft tissue invasion, presence of distant metastasis, clinically or radiologically abnormal lymph nodes and thyroid gland [13,16,17]. Recent article by Bakkar et al [18] recommended addition of total thyroidectomy regardless of the size of TGDC carcinoma as selecting a subset of patients free of concomitant thyroid cancer is a difficult task. Newer surgical techniques like robot assisted neck dissection via a transaxillary and retroauricular (TARA) approach for TGDC carcinoma treatment has been reported as a novel surgical approach with benefits of smaller incision, wider operating view and lesser complications [19]. This technique includes excision of TGDC carcinoma through retroauricular incision followed by total thyroidectomy with central and lateral neck dissection through transaxillary approach [19]. Use of RAI in patients with total thyroidectomy is also controversial and should be used according to the established guidelines for differentiated thyroid cancers [17]. Further studies are needed to help better guide clinicians and surgeons for management of TGDC carcinoma. Our patient received total thyroidectomy followed by low dose RAI remnant ablation as she was older and tumor was almost abutting the bone.

Overall prognosis for papillary TGDC carcinoma like well differentiated papillary thyroid carcinoma is excellent, with occurrence of metastatic lesions occurring in less than 2% of cases [8].

Conclusion

Proper workup including clinical and radiologic evaluation of cyst, thyroid gland and cervical lymph nodes along with FNA is needed to guide type and extent of surgery and overall prognosis in TGDC carcinoma.

References

1. Balalaa N, Megahed M, Ashari MA, Branicki F (2011) Thyroglossal duct cyst papillary carcinoma. Case Rep Oncol 4: 39-43. Link: https://goo.gl/ekskgb
2. LiVolsi VA, Perzin KH, Savetsky L (1974) Carcinoma arising in median ectopic thyroid (including thyroglossal duct tissue). Cancer 34: 1303-1315. Link: https://goo.gl/xLWDaH
3. Kazemi M, Assadi M, Kazemi AA, Ghazvini LA (2006) Primary papillary carcinoma in a thyroglossal duct cyst. Hell J Nucl Med 9: 39-40. Link: https://goo.gl/N0MzMC
4. Martin-Perez E, Larranaga E, Marron C, Monje F (1997) Primary papillary carcinoma arising in a thyroglossal duct cyst. Eur J Surg 163: 143-145. Link: https://goo.gl/2vD79e
5. Murphy JP, Budd DC (1977) Thyroglossal duct cysts in the elderly. South Med J 70: 1247-1248. Link: https://goo.gl/Jn0XkW
6. Ducic Y (2002) Thyroglossal duct cysts in the elderly population. Am J Otolaryngol 23: 17-19. Link: https://goo.gl/WQ2hi4
7. Gross E, Sichel JY (2006) Congenital neck lesions. Surg Clin North Am 86: 383-392. Link: https://goo.gl/ky5g88
8. Fernandez JF, Ordonez NG, Schultz PN, Samaan NA, Hickey RC (1991) Thyroglossal duct carcinoma. Surgery 110: 928-934; discussion 934-935.
9. Van Vuuren PA, Balm AJ, Gregor RT, Hilgers FJ, Loftus BM , et al. (1994) Carcinoma arising in thyroglossal remnants. Clin Otolaryngol Allied Sci 9: 509-515. Link: https://goo.gl/PHKRvB
10. Shahin A, Burroughs FH, Kirby JP, Ali SZ (2005) Thyroglossal duct cyst: A cytopathologic study of 26 cases. Diagn Cytopathol 33: 365-369. Link: https://goo.gl/2gX4w6

Citation: Ashraf S, Nadkarni P, Izquierdo RE, Bansal N (2016) Dilemma in Managing Thyroglossal Duct Cyst Carcinoma. Glob J Rare Dis 1(1): 001-003.

11. Choi YM, Kim TY, Song DE, Hong SJ, Jang EK, et al. (2013) Papillary thyroid carcinoma arising from a thyroglossal duct cyst: A single institution experience. Endocr J 60: 665-670. Link: https://goo.gl/7AZ4lI

12. Hilger AW, Thompson SD, Smallman LA, Watkinson JC (1995) Papillary carcinoma arising in a thyroglossal duct cyst: A case report and literature review. J Laryngol Otol 109: 1124-1127. Link: https://goo.gl/Xv7OQZ

13. Chrisoulidou A, Iliadou P, Doumala E, et al. (2013) Thyroglossal duct cyst carcinomas: Is there a need for thyroidectomy? Hormones (Athens) 12: 522-528. Link: https://goo.gl/DYRrP2

14. Patel SG, Escrig M, Shaha AR, Singh B, Shah JP (2002) Management of well-differentiated thyroid carcinoma presenting within a thyroglossal duct cyst. J Surg Oncol 79: 134-139; discussion 140-141. Link: https://goo.gl/q1g3Kv

15. Plaza CP, Lopez ME, Carrasco CE, Meseguer LM, Perucho Ade L (2006) 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 13: 745-752. Link: https://goo.gl/Gux8Nr

16. Hartl DM, Al Ghuzlan A, Chami L, Leboulleux S, Schlumberger M, et al. (2009) High rate of multifocality and occult lymph node metastases in papillary thyroid carcinoma arising in thyroglossal duct cysts. Ann Surg Oncol 16: 2595-2601. Link: https://goo.gl/Fzcdt5

17. Haugen BR, Alexander EK, Bible KC, Doherty GM, Mandel SJ, et al. (2016) 2015 American Thyroid Association Management Guidelines for Adult Patients with Thyroid Nodules and Differentiated Thyroid Cancer: The American Thyroid Association Guidelines Task Force on Thyroid Nodules and Differentiated Thyroid Cancer. Thyroid 26: 1-133. Link: https://goo.gl/sgAioP

18. Bakkar S, Biricotti M, Stefanini G, Ambrosini CE, Materazzi G, et al. (2016) The extent of surgery in thyroglossal cyst carcinoma. Langenbecks Arch Surg [Epub ahead of print]. Link: https://goo.gl/frlljB

19. Byeon HK1, Ban MJ, Lee JM, Ha JG, Kim ES, et al. (2012) Robot-assisted sistrunk's operation, total thyroidectomy, and neck dissection via a transaxillary and retroauricular (TARA) approach in papillary carcinoma arising in thyroglossal duct cyst and thyroid gland. Ann Surg Oncol 19: 4259-4261. Link: https://goo.gl/XPJMFh

Citation: Ashraf S, Nadkarni P, Izquierdo RE, Bansal N (2016) Dilemma in Managing Thyroglossal Duct Cyst Carcinoma. Glob J Rare Dis 1(1): 001-003.