Papillary Carcinoma of Thyroglossal Duct Cyst- A Diagnostic and Therapeutic Conundrum

Agrawal V,[1] Colbert KR,[2] Kumar S,[2] Dharanya,[2] Bhat RV[1]

[1]Department of Pathology,

[2]Department of ENT

Indira Gandhi Medical College and Research Institute,

Vazhudavur Road, Kathirkamam, Puducherry, India.

Corresponding Author

Satish Kumar

Department of ENT

Indira Gandhi Medical College and Research Institute,

Vazhudavur Road, Kathirkamam, Puducherry, India.

E-mail: docsatkumar@gmail.com

Citation

Agrawal V, Colbert KR, Kumar S, Dharanya, Bhat RV. Papillary Carcinoma of Thyroglossal Duct Cyst- A Diagnostic and Therapeutic Conundrum. *Kathmandu Univ Med J.* 2023;81(1):106-9.

ABSTRACT

Thyroglossal duct cysts (TGDC) are the most common congenital midline neck mass. It accounts for 70% of congenital neck masses in children and 7% in the general population. Untreated, Thyroglossal duct cyst may transform into a thyroid carcinoma in less than 1% of cases. A 47 year old female patient presented with complaints of swelling on the anterior aspect of neck for 1 year which was progressively increasing in size. Smears from needle aspiration revealed features suspicious of Papillary carcinoma of thyroid which was confirmed as on histopathology. Thyroid gland and the adjacent lymph nodes were free of the tumour on Computed Tomography (CT). Thyroglossal duct cysts undergoing neoplastic change is more common among women. Though sistrunk's procedure is widely accepted as a primary management, the role of total thyroidectomy with radioiodine therapy still remains a controversy due to contrasting opinions available in the literature.

KEY WORDS

Papillary carcinoma, Thyroglossal duct cyst, Thyroid

INTRODUCTION

Thyroglossal duct cyst contributes to 70% of congenital midline neck mass among children and 7% in the general population. A normal thyroid gland originates during the third week of foetal life from the midline endodermal invagination of the foregut at the level of foramen caecum and descends through the floor of mouth and passes anteriorly to inferior part of neck leaving behind an embryonic midline duct known as the thyroglossal duct. Thyroglossal duct cyst results when its regression fails.[1]

Untreated thyroglossal duct cysts may transform into a thyroid carcinoma in less than 1% of cases, mostly during third or fourth decade of life.[2,3] These carcinomas are generally non aggressive with rare lymphatic spread. Papillary carcinoma is the most Commonly observed variant (75-80%), followed by mixed papillary follicular carcinoma (7%), squamous cell carcinoma (5%), follicular carcinoma (1.7%), Hurthle cell carcinoma and anaplastic carcinoma (0.9%).[4]

CASE REPORT

A 47 year old female patient presented to ENT outpatient department with complaints of swelling on the anterior aspect of neck for 1 year which was progressively increasing in size. There was no history of pain, dysphagia, voice change, loss of appetite and weight. Patient did not have any systemic illnesses and her menstrual cycles were regular. There was no history suggestive of hyper or hypothyroidism. No history of thyroid disorders in the family as well.

On examination, a well-defined smooth surfaced swelling of approximately 2x2 cm was seen on the anterior aspect of neck in the midline just below the hyoid bone. The overlying skin was normal. On palpation, the swelling was non-tender and not warm, tense cystic in consistency, non-pulsatile, non-compressible and fluctuant. The swelling moved with deglutition and also with protrusion of tongue. The thyroid gland was normal on palpation and no neck

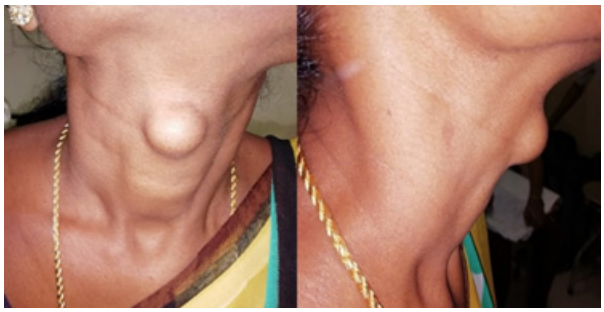

Figure 1. A well-defined globular swelling of size 1.5x1.5 cm in the midline just below the hyoid bone which moved with deglutition and protrusion of tongue.

nodes were palpable (Fig. 1).

With a provisional diagnosis of thyroglossal duct cyst and differential diagnosis of branchial cyst and dermoid cyst, the patient was subjected to laboratory investigations. Thyroid function test and the other routine blood and urine investigations performed were within normal range. Ultrasound scan of the neck confirmed a midline cystic swelling of size 2x1.4 cm in the subcutaneous plane with increased vascularity just below the hyoid bone. There was a calcified mass of size 08x0.5 cm withing the cystic swelling. Rest of the neck and thyroid gland were normal with no cervical lymphadenopathy.

Fine needle aspiration cytology (FNAC) was performed and 3 ml of straw coloured fluid was aspirated. The centrifuged smears and post-aspiration smears were moderately cellular and showed clusters of epithelial cells with occasional microfollicular and papillary pattern of arrangement. These epithelial cells had moderate anisonucleosis, nuclear membrane irregularity and occasional pseudonuclear inclusions with abundant calcific debris (Psammoma bodies) in haemorrhagic background (Fig. 2). A diagnosis of features suspicious of Papillary carcinoma of Thyroid was given. Ultrasound guided FNAC of the thyroid gland

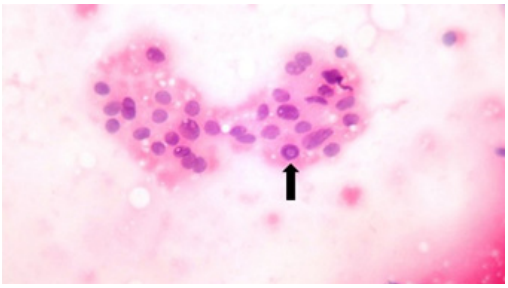

Figure 2. NAC of midline neck swelling showing Thyroid follicular epithelial cell clusters displaying moderate anisokaryosis with irregular nuclear membrane, powdery chromatin and rare pseudonuclear inclusions suggesting Papillary carcinoma (H&E stain 40x magnification)

exhibited benign follicular epithelial cell clusters.

A contrast enhanced CT of the neck was performed in view of suspected malignancy. It revealed a cystic lesion measuring 1.3x1.4 cm below the hyoid bone in midline with specs of calcifications within. Both lobes of thyroid gland, neck vessels, thyroid cartilage and hyoid bone were found

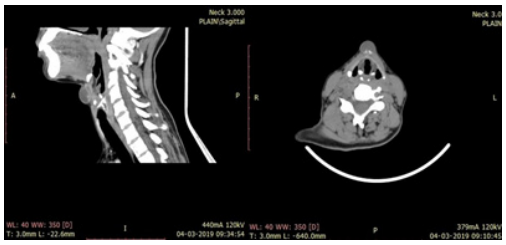

Figure 3. CECT neck demonstrated a midline cystic lesion 1.3x1.4 cm just below the hyoid bone in the subcutaneous plane and specks of calcifications noted within. The thyroid and neck.

to be normal. There were no cervical nodes either (Fig. 3).

Since there were no signs of local or nodal extension both clinically and radiologically, sistrunks operation was performed to excise the cyst. The cyst on excision measured 2x2x1 cm, with no extracapsular extension. The specimen consisting of the cyst, the cranial tract with the body of hyoid bone and the caudal tract was sent for histopathological examination.

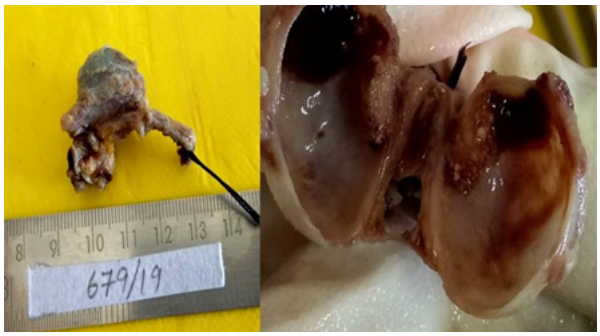

Figure 4. Gross specimen of Sistrunk specimen: Showing grey brown cystic mass (Thyroglossal cyst) measuring 2x1.5x1 cm with an attached tube like structure (Thyroglossal duct) measuring 2.3 cm in length. Dissected specimen revealed a Unilocular cyst filled with thin brownish fluid and a nodule projecting into it; Nodule measured 3 mm in diameter showing papillary excrescences with foci of calcification.

Grossly the cystic mass was grey brown in colour and, measured 2x1.5x1 cm with an attached tube like structure measuring 2.3 cm in length. The cut surface showed the cyst to be unilocular, thin walled, filled with a brownish fluid and, a nodule projecting within measuring 3 mm. The nodule had papillary excrescences with foci of calcification (Fig 4). On microscopy the cyst wall was lined by cuboidal epithelium with occasional fibrocollagenous tissues at some places suggestive of a thyroglossal cyst. The nodule attached to the cyst wall had a complex branching papillary structure with extensive areas of dystrophic calcifications on lower magnification. Higher magnifications revealed papillary structures with central fibrovascular core, lined cuboidal cells showing classical Orphan Annie eyed nucleus and, occasional intranuclear grooves (Fig. 5). Distal end of the duct had thyroid follicles interspersed with lymphoid aggregates. With this a final diagnosis of Papillary Carcinoma Thyroid arising from Thyroglossal Duct Cyst was made. All the surgical margins were free of any tumour.

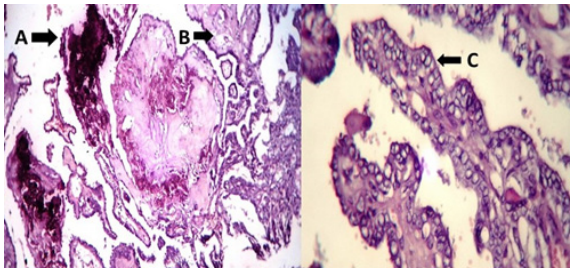

Figure 5. Histopathology section from Thyroglossal cyst showing complex branching papillary structures(B) with extensive areas of dystrophic calcifications (A) suggesting Psammomatous calcifications (H & E, 10x magnification). Higher magnification revealed papillary structures with central fibrovascular core and lining by classical Orphan Annie eyed nucleus (C) and occasional intranuclear grooves suggesting Papillary Carcinoma Thyroid arising from Thyroglossal Duct Cyst (H & E, 40x magnification)

DISCUSSION

Thyroid gland is the first endocrine gland to develop which begins by third week of gestation from the floor of pharyngeal gut (foramen caecum).[1] It descends down as a bilobed diverticulum but remains connected to the foregut by a narrow canal, the thyroglossal duct. It usually obliterates or disappears by tenth week of gestation but sometimes remains in the form of a cyst, tract or duct, occasionally associated with ectopic thyroid within the cyst or duct.[5]

TGD cysts are the most common congenital neck masses, with prevalence of 7% among the general population, whereas it contributes to 70% congenital neck mass in children.[1] Though most of the TGD cysts remain benign, a small percentage (0.7 to 1%) undergoes neoplastic change.[6] Women more frequently affected than men (2:1).[7]

This fascinating origin of cancerous lesions in TGDC has led to postulation of theories. One such theory suggests de novo origin from the thyroid cell rests. It is supported by the presence of thyroid tissue in 62% TGDC's operated and reinforced by the absence of documented medullary carcinoma in TGDC.[8] Another theory suggests that these are metastasis from the occult thyroid primaries of papillary carcinoma.[9] But theoretically the only true primary carcinoma of TGDC could be Squamous cell carcinoma originating from the ductal epithelium.[10]

The criteria for diagnosis of primary carcinoma of thyroglossal duct as given by Widstrom et al. which includes 1. Histological identification of thyroglossal duct carcinoma, 2. Demonstration of normal epithelial lining of the thyroglossal duct, 3. Normal thyroid follicles within walls of the cyst, 5. Normal thyroid tissue adjacent to the tumour and 6. No findings of primary thyroid carcinoma on histopathological examination of the thyroid gland.[11] All these criteria were demonstrated in our case, the last criteria being supported by a normal FNAC of the thyroid gland.

TGDC carcinoma mostly presents as an asymptomatic midline neck mass (95.1%) with an incidental diagnosis in majority of the cases (73.3%). Hence, the workup includes imaging studies with careful clinical evaluation followed by histopathological studies. The presence of microcalcifications along the wall in ultrasonography (USG) and pericystic calcifications with contrast enhancement on computerized tomography (CT) offers a clue towards the diagnosis.[12,13] The diagnosis is aided by a good pathologist as the sensitivity of Fine needle aspiration Biopsy for TGDC ranges from 56-62% with a positive predictive value of 69%, whereas it is 85% sensitive for primary thyroid carcinomas.[14]

Though sistrunk's procedure is accepted as a primary management, the role of total thyroidectomy with radioiodine therapy still remains a controversy due to contrasting opinions available in the literature. The presence of normal thyroid morphology on radiological studies leads to further dilemma, as with our case which demonstrated similar findings. The risk factors considered for poor prognosis are Male sex, Age > 45 years, Size > 4 cm, Extracystic invasion, Metastasis to lymph nodes, history of neck irradiation and nodules within thyroid on scan and thus indicators for performing a total thyroidectomy.[15]

In the presented case female sex, tumour size < 4 cm, absence of extracystic invasion and nodal metastasis, with no previous history of neck irradiation and a normal thyroid morphology on scan were favourable, the only unfavourable risk factor being age > 45 years. Since the 10 year survival rate of papillary carcinoma is > 90% and the locoregional recurrence rate is 2.16%, we performed only sistrunk's procedure necessitating follow up with USG every 6 months for 1st year and annually after that.[16-18]

Being a rare but clinically important lesion it's important to keep primary carcinoma of thyroglossal duct in mind when dealing with cystic neck lesion

The occurrence of carcinoma in TGDC offers a great challenge with dilemma in comparison to those arising primarily from the thyroid gland, with respect to diagnosis and treatment mandating careful surgical planning. The need for total thyroidectomy with sistrunk's procedure can be curtailed based on the presence of concurrent or suspicious lesions in the thyroid gland and associated - risk factors.

REFERENCES

1. Carter Y, Yeutter N, Mazeh H. Thyroglossal duct remnant carcinoma: beyond the Sistrunk procedure. *Surg Oncol.* 2014 Sep 1;23(3):161-6.
2. Torcivia A, Polliand C, Ziol M, Dufour F, Champault G, Barrat C. Papillary carcinoma of the thyroglossal duct cyst: report of two cases. *Rom J Morphol Embryol*. 2010 Jan 1;51(4):775-7.
3. Aghaghazvini L, Mazaher H, Sharifian H, Aghaghazvini S, Assadi M. Invasive thyroglossal duct cyst papillary carcinoma: a case report. *J Med Case Rep*. 2009 Dec;3(1):1-4.
4. Kalyani R, Hebbar A, Murthy S. Primary papillary carcinoma arising in thyroglossal duct cyst: A rare case report. *Nat J Lab Med*. 2015;4(3):27-9.
5. Kennedy TL, Whitaker M, Wadih G. Thyroglossal duct carcinoma: a rational approach to management. *Laryngoscope*. 1998 Aug;108(8):1154-8.
6. Martins AS, Melo GM, Tincani AJ, Lage HT, Matos PS. Papillary carcinoma in a thyroglossal duct: case report. *Sao Paulo Med J.* 1999;117:248-50.
7. Zivaljevic V, Bozic V. Papillary carcinoma in a thyroglossal duct cyst. *Acta Chir Belg.* 2006 Jan 1;106(2):252-3.
8. Hilger AW, Thompson SD, Smallman LA, Watkinson JC. Papillary carcinoma arising in a thyroglossal duct cyst: a case report and literature review. *J Laryngol Otol.* 1995 Nov;109(11):1124-7.
9. Tew S, Reeve TS, Poole AG, Delbridge L. Papillary thyroid carcinoma arising in thyroglossal duct cysts: incidence and management. *Aust N Z J Surg*. 1995 Oct;65(10):717-8.
10. Shah S, Kadakia S, Khorsandi A, Andersen A, Iacob C, Shin E. Squamous cell carcinoma in a thyroglossal duct cyst: a case report with review of the literature. *Am J Otolaryngol*. 2015 May 1;36(3):460-2.
11. Tharmabala M, Kanthan R. Incidental thyroid papillary carcinoma in a thyroglossal duct cyst–management dilemmas. *Int J Surg Case Rep*. 2013 Jan 1;4(1):58-61.
12. 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 Dec;25(11):947-54.
13. Branstetter BF, Weissman JL, Kennedy TL, Whitaker M. The CT appearance of thyroglossal duct carcinoma. *AJNR Am J Neuroradiol*. 2000 Sep 1;21(8):1547-50.
14. Shahin A, Burroughs FH, Kirby JP, Ali SZ. Thyroglossal duct cyst: a cytopathologic study of 26 cases. *Diagn Cytopathol.* 2005;33(6):365-9.
15. Pellegriti G, Lumera G, Malandrino P, Latina A, Masucci R, Scollo C, et al. Thyroid cancer in thyroglossal duct cysts requires a specific approach due to its unpredictable extension. *J Clin Endocrinol Metab.* 2013 Feb 1;98(2):458-65.
16. Pelizzo MR, Boschin M, Toniato A, Pagetta C, Mian C, Rubello D. Diagnosis, treatment, prognostic factors and long-term outcome in papillary thyroid carcinoma. *Minerva endocrinologica*. 2008 Dec 1;33(4):359-79.
17. Matsuzu K, Sugino K, Masudo K, Nagahama M, Kitagawa W, Shibuya H, et al. Thyroid lobectomy for papillary thyroid cancer: long-term follow-up study of 1,088 cases. *World J Surg*. 2014 Jan;38(1):68-79.
18. Lu Y, Jiang L, Chen C, Chen H, Yao Q. Clinicopathologic characteristics and outcomes of papillary thyroid carcinoma in younger patients. *Medicine.* 2020