



THYROGLOSSAL DUCT CYST WITH INCIDENTAL THYROID PAPILLARY CARCINOMA – A RARE CASE REPORT

Surekha Bhalekar ¹, Sonia Kundu ^{2*}, Hemant Bhalekar ³, Prakash Roplekar ⁴,
Manthan Patel ², Devarshi Mhatre ⁵

¹ Associate Professor, Department of Pathology, D. Y .Patil School of Medicine, Nerul, Navi Mumbai, Madhya Pradesh, India.

² Resident, Department of Pathology, D. Y. Patil University School of Medicine, Nerul, Navi Mumbai, Madhya Pradesh, India.

³ Pathologist, Dr Bhalekar Path Lab, New Panvel, Navi Mumbai, Madhya Pradesh, India.

⁴ Head of the Department, Department of Pathology, D. Y. Patil School of Medicine, Navi Mumbai, Madhya Pradesh, India.

⁵ MS, ENT, Ashirwad hospital, old Panvel, Navi Mumbai, Madhya Pradesh, India.

Corresponding Author:- **Sonia Kundu**

E-mail: sonia.kundu3@gmail.com

Article Info	ABSTRACT
<p>Received 23/01/2016 Revised 27/01/2016 Accepted 30/01/2016</p> <p>Key words: Thyroglossal duct cyst, Thyroid Papillary Carcinoma, Sistrunk operation.</p>	<p>Thyroglossal duct cysts (TDC) are midline congenital neck masses occurring from a developmental anomaly of thyroid gland. Majority of patients present within first two decades of life. Persistent TDC occurs rarely in adults (7%). TDC are usually treated by Sistrunk operation. Malignancy occurring in TDC is extremely uncommon (1-2%). Majority of Thyroid Papillary Carcinoma occurring in TDC are diagnosed as incidental findings on histopathology after surgical resection of TDC. We report a case of an incidental thyroid papillary carcinoma (TPC) in suprahyoidal TDC in a 47 yrs old male and is described with special regard to its rarity and to discuss the diagnostic and management dilemmas. The rare findings of incidental TPC in TDC highlights the issue regarding best practice guidelines for further management which still remains debatable and controversial.</p>

INTRODUCTION

Thyroglossal duct cysts (TDCs) are the thyroid gland development anomaly with frequency of 70% in childhood and 7% in adults. During thyroid gland descent in embryogenesis, the connection between the thyroid gland and floor of the pharynx (foramen caecum) may persist to form TDC. Only about 1% of TDC are malignant, most of which are papillary thyroid carcinoma having good prognosis. Thyroglossal duct carcinoma (TGDC) may be clinically indistinguishable from benign TDC, and the diagnosis in most cases is incidental after surgical resection. Exact origin and optimal management of incidental Thyroglossal duct carcinoma is still controversial.

CASE REPORT

45 yrs old male presented with painless, smooth

cystic midline neck swelling progressively increasing slowly over past one year. Swelling moved with swallowing and tongue protrusion. Thyroid function tests and other laboratory investigations were normal. Ultrasonography suggested Thyroglossal duct cyst (TGC) showing cystic lesion with eccentric calcification at midline neck (supra hyoid level) measuring 2.5 x 1 cm in size. FNAC showed thyroid follicular cells along with colloidophages in the background of thin colloid and areas of calcification suggesting the diagnosis of TGC.

Mass was resected by Sistrunk procedure. There was neither local signs of invasion of tissue surrounding the cyst or duct at surgery nor any cervical lymphadenopathy.

Histopathological examination of specimen showed 2.5 x 1 cm sized cystic swelling, cyst wall



thickness measured 0.3 cm, partly filled with solid brownish tissue measuring 1 x 1cm. Sections examined showed a small focus of Papillary Thyroid Carcinoma, arising in a thyroglossal cyst (Figure 1,4).Tumor showed mainly papillary architecture with characteristic nuclear features of papillary thyroid carcinoma. Tumor cells showed large and irregular nuclei with prominent nuclear grooving and at places intranuclear inclusions. Psammoma bodies were also noted. Cyst wall showed lympho plasmacytic infiltrate but lymphovascular and perineural

invasion were not seen (Figure 2). At the periphery of tumor, normal thyroid tissue was seen (Figure 3). So final diagnosis made was Papillary Thyroid Carcinoma arising in TGC. No further surgical treatment was undertaken as this was considered as an incidental thyroid papillary carcinoma. The patient follow-up was done which included thyroid function tests, head and neck examinations, Ultrasonography of the surgical region and thyroid, and total body scintigraphy. The patient had been followed up for one year with no further evidence of disease.

1

Fig 1. 10 X, H & E, Papillary thyroid carcinoma arising in a thyroglossal duct cyst

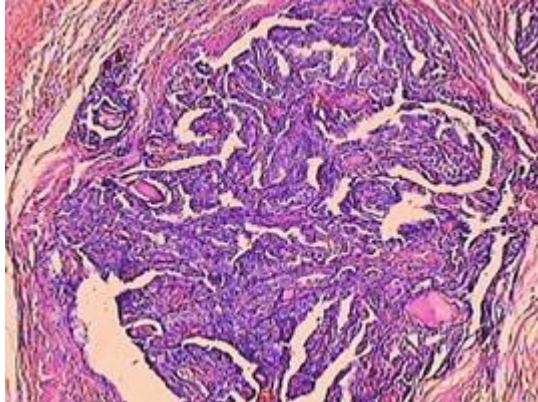


Fig 2. 10 X, H & E, Tumor showed mainly papillary architecture. Tumor cells showed large and irregular nuclei with prominent nuclear grooving and at places intranuclear inclusions

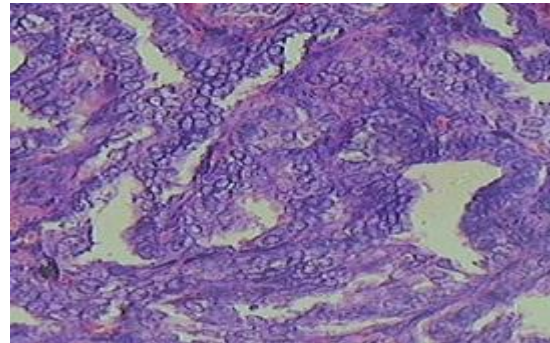


Fig 3. 10x, H&E, Ectopic thyroid tissue seen at the periphery of thyroglossal duct cyst carcinoma

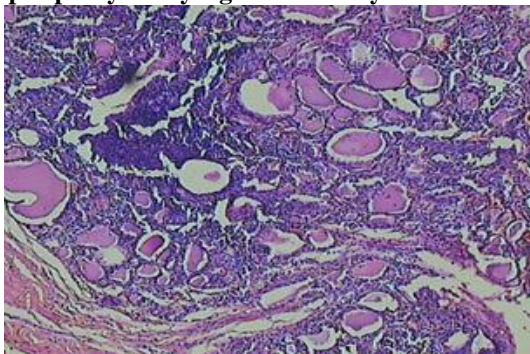
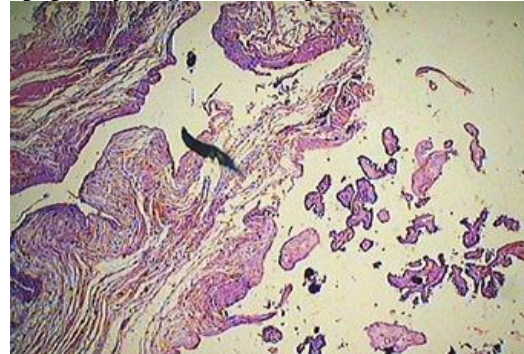


Fig 4. 10x, H&E, Thyroglossal duct cyst wall along with papillary fragments and psammoma bodies



DISCUSSION

In thyroid development, TDC is the most common anomaly. Thyroid gland descends from foramen caecum at the base of tongue to point below the thyroid cartilage via an epithelial tract known as thyroglossal duct. This thyroglossal duct disappears by 5th to 10th gestational week. Incomplete atrophy of duct leads to thyroglossal cyst formation which presents as midline neck mass at various levels viz thyrohyoidal (61%), suprathyroidal (24%), suprasternal (13%) and intra lingual region (2%) [1]. TDC is more common in women than in men (1.5 : 1) [2].

TDC clinically presents with soft to firm fluctuant mass, generally movable with swallowing and tongue protrusion. Majority of TDC are benign but 0.7 to 1 % may be malignant, 80% of these being Papillary Carcinoma, 7% Mixed Papillary and Follicular carcinoma, 5% Squamous

cell carcinoma, 1.7% Adenocarcinoma and Follicular carcinoma and 0.9% Anaplastic carcinoma [3]. Among all cancers, Squamous cell carcinoma has worst prognosis and papillary thyroglossal duct cyst carcinoma has best favorable prognosis with metastasis in less than 2% cases. Approximately 4% of thyroglossal duct carcinomas had local invasion while 11% were found to include cervical lymph node metastasis [4]. In 20% of thyroglossal duct carcinomas contemporary appearance of thyroid carcinoma was described.

There are two theories to explain the thyrogenic origin of TGD Adenocarcinoma. One, the denovo theory stating that ectopic thyroid tissue can be seen histopathologically in 62% of cases and this is supported by non occurrence of medullary carcinoma in TGD because it originates from parafollicular cells .Other theory



is metastatic theory suggesting that thyroglossal duct cyst carcinoma is metastatic from an occult primary thyroid gland, as papillary carcinoma is usually multifocal [5]. The presence of entrapped thyroid follicles in the cyst wall, as seen in our case and the persistent absence of medullary carcinoma occurring in a TDC in keeping with its embryology, strongly favours the de novo theory.

Imaging diagnostic techniques including ultrasound, CT scan are usually unable to preoperatively diagnose malignant disease in TDC. FNAC though simple, rapid, inexpensive with minimal risk complications is more reliable in diagnosing solid tumors rather than cystic lesions. Additionally, FNAC is not cost-effective due to the rarity of this malignancy and remains an inappropriate tool for routine use in children. FNAC gives correct result in only 66% of the cases [4].

Thus Joseph and Komorowski proposed strict criteria to diagnose primary TDC carcinoma which included a Thyroglossal remnant, ectopic thyroid nests within the cyst wall and a clinically normal thyroid gland [6].

Patel *et al* study of 57 cases of well-differentiated carcinomas in TDC concluded that significant predictor for overall survival was completeness of TDC excision. Patients treated with simple excision had a worse prognosis than those with Sistrunk procedure. (Resection of cyst along with body of hyoid bone and a cone of the bar of tongue muscle upto the foramen caecum It is hard to justify total thyroidectomy as the overall cure rate of a carcinoma arising in a TDC treated by Sistrunk's procedure is over 95% [7].

Recently, prognostic risk group assessments are used to identify patients who would benefit from additional total thyroidectomy. Plaza et al identified high risk factors to select patients who would benefit from additional total thyroidectomy with radio-iodine and suppressive hormonal therapy. a) Irradiation exposure b) Age more than 45 yrs c) Presence of tumor in thyroid gland radiologically. d) Tumor size more than 1.5cm in diameter. e) Presence of

clinical or radiological nodes f) Cyst wall invasion g) Histopathologically positive margins .When regional lymph node metastasis are detected preoperatively, in addition to Sistrunk procedure, total thyroidectomy and regional neck dissection are recommended.

In patients treated with Sistrunk procedure alone, suppressive thyroid hormone therapy is recommended to keep thyroid stimulating hormone levels between 0.1 and 0.5 mIU/L. In follow up management protocol of these patients, annual ultrasound scans of neck, TSH levels and total body scintigraphy are recommended [1]. For papillary carcinoma in TDC, long term follow up (more than 20 yrs) is recommended [8]. The overall 10 year survival rate for TPC in TDC is 95.6%.

CONCLUSION

Adult presenting with Thyroid Papillary Carcinoma (TPC) in Thyroglossal duct cyst (TDC) is a rarity, mostly being diagnosed post operatively on histopathology as an incidental finding as FNAC done before operation is usually inconclusive. So Pathologist should always consider this rare cancer in differential diagnosis of any midline neck mass because despite of its good prognosis, long term clinical follow up is recommended due to the prolonged natural clinical course of TPC.

ACKNOWLEDGEMENT: None

CONFLICT OF INTEREST:

The authors declare that they have no conflict of interest.

STATEMENT OF HUMAN AND ANIMAL RIGHTS

All procedures performed in human participants were in accordance with the ethical standards of the institutional research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. This article does not contain any studies with animals performed by any of the authors.

REFERENCES

1. Tharmabala M and Kanthan R. (2013). Incidental thyroid papillary carcinoma in thyroglossal duct cyst – management dilemmas. *Int. J. Surg Case Rep*, 4(1), 2013, 58-61.
2. Martins AS, De Melo GM, Tincani AJ, Lage HT, De Matos PS. (1999). Papillary carcinoma in thyroglossal duct : case report. *Sau Paulo Med. J.*, 117, 6.
3. Senthikumar JF, Neville R, Arvind R. (2013). Malignant thyroglossal duct cyst with synchronous occult thyroid gland carcinoma. *Indian J Endoa Metab*, 17, 9368.
4. Yang SI, Park KK, Kim JH. (2013). Papillary carcinoma arising from thyroglossal duct cyst with thyroid and lateral neck metastasis. *Int J surg Case Rep*, 4(8), 704-707.
5. Kazemi M, Assadi M, Kazami AA, Ghazvini LA. (2006). Primary papillary carcinoma in thyroglossal duct cyst. *J Nud Med*, 9, 39-40.
6. Joseph TJ, Komorowski RA. (1975). Thyroglossal duct carcinoma. *Hum pathol*, 6, 717-729.
7. Plaza CPR, Lopez ME, Carrasco CEG, Meseguer LM, Perucho AF. (2006). Management of well differentiated thyroglossal remnant thyroid carcinoma : time to close the debate> Report of five cases and proposal of definitive algorithm for treatment. *Annals of surgical oncology*, (13/5), 745-752.
8. Dan D, Rambally R, Narayan singh V, Maharaj R, Seetharaman H. (2012). A case of malignancy in a thyroglossal duct cyst recommendations for management. *Journal of the National Medical Association*, 104(3-4), 211-214.



