



An enigmatic presentation of concurrent papillary carcinoma in thyroid and thyroglossal cyst: a dual challenge

Authors

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Abstract

In the realm of thyroid malignancies, the coexistence of concurrent papillary carcinoma in both the thyroid gland and thyroglossal cyst presents a unique and intriguing clinical scenario. We present an unusual case of 40 year old female with these dual papillary carcinoma who underwent total thyroidectomy with thyroglossal cyst excision with left selective neck dissection followed by I-131 radiation ablation therapy. On follow up, the patient was found to have papillary carcinoma cervical metastasis on the contralateral side 2 months later and was treated by right functional neck dissection. Through this case report, we aim to contribute to the complexities associated with such coexisting papillary carcinomas, emphasizing the need for comprehensive evaluation and tailored therapeutic approaches in managing these intricate cases.

Keywords: papillary carcinoma, thyroid, thyroglossal duct cyst.

Introduction

From the embryological point of view, the thyroid gland develops from an epithelial invagination which descends from foramen cecum of tongue towards first and second tracheal rings in the neck along midline during the third and fourth week of embryological development.¹ Thyroglossal tract is an epithelial tract left behind during this descend and it usually disappears during fifth to tenth gestational weeks.² It is the incomplete atrophy of this thyroglossal tract which gives origin to thyroglossal duct cyst.

The most common anomaly in the development of thyroid gland is thyroglossal duct cyst.³ Papillary carcinoma arising from a thyroglossal cyst is an infrequent finding with an incidence of <1%.⁴ Brentano in 1911 and Ucherman in 1915 are

acknowledged for being the first to document a neoplasm in a thyroglossal duct with median age of presentation being 40 years.⁵ Malignancy associated with thyroglossal duct cyst have been reported in approximately 278 cases only.⁶

When compared to primary carcinoma of thyroid, regional lymph node metastasis from papillary carcinoma of thyroglossal duct occur much less frequently with incidence of less than 8%.⁷ The concurrent thyroglossal duct cyst papillary carcinoma and thyroid papillary carcinoma is an even more uncommon presentation. We present a case of a 40 year old female who presented with neck mass with radiological and FNAC reports pointing towards right lobe of thyroid and thyroglossal cyst papillary carcinoma with suspicious left cervical nodal metastasis. But the

post operative histopathology report showed thyroglossal duct cyst papillary carcinoma alone. 2 months later, the same patient presented with right cervical nodal metastasis. This case report delves into the intricate details of these dual papillary carcinomas, shedding light on their diagnostic challenges, therapeutic considerations and the broader implications for patient management.

Case Report

A 40 year old female with no known comorbidities presented with the complaints of gradually progressing swelling in the neck since 6 months. There was no associated change in voice, dyspnoea or dysphagia. On local examination, there was a 2 x 2 cm firm swelling above the thyroid notch in the anterior aspect of neck which moved with deglutition and protrusion of tongue. No other swelling were palpable.

CT scan and MRI of neck revealed features consistent with thyroid malignancy with enlarged metastatic left level III node and possibility of neoplastic etiology of thyroglossal duct cyst. FNAC from right thyroid nodule was reported as papillary carcinoma. Left cervical FNAC showed cyst macrophage and follicular cell clusters.

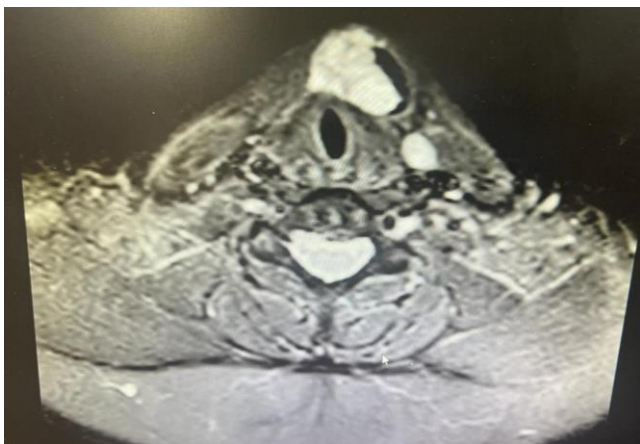


Image 1: MRI Neck with contrast showing well circumscribed midline mass lesion involving visceral space of infrahyoid neck.

The patient underwent total thyroidectomy with thyroglossal cyst excision with left selective neck

dissection. The histopathology was reported as papillary carcinoma of thyroglossal cyst with tumour deposits in left carotid triangle lymph nodes and no evidence of malignancy in thyroid gland and other cervical lymph nodes. 6 weeks post operatively, I-131 whole body scan was done for the patient on thyroid hormone withdrawal and it showed significant tracer uptake in the neck (thyroid bed) and there was no demonstrable tracer avid metastasis. Serum Thyroglobulin was 71.65 ng/ml and antithyroglobulin antibody was 3.7 IU/ml. In view of evidence of residual functioning thyroid tissue in neck and elevated tumour marker values, the patient was subjected to I-131 radiation ablation therapy.

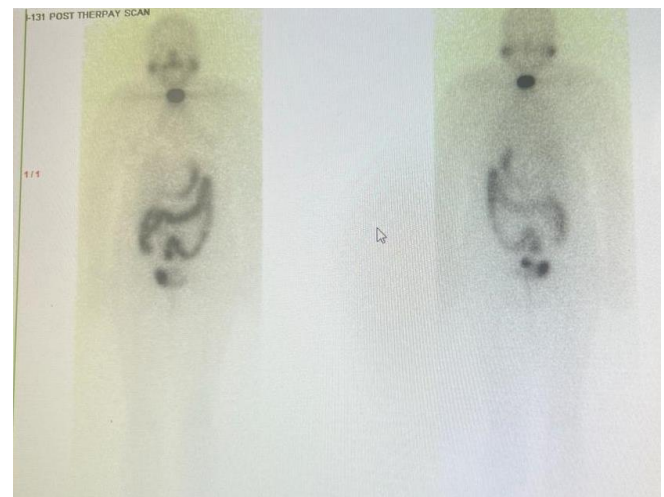


Image 2: I-131 scan showing significant tracer uptake in the neck

2 months later, the patient noticed swelling over right side of neck and ultrasound neck revealed enlarged right level III cervical lymph node. USG guided FNAC from the same showed atypical cells suggestive of metastasis. PET CT was done and it showed low grade metabolically active prominent right level III cervical lymph node and no abnormal metabolically active lesion in the surgical bed. There was also no evidence of metabolically active disease anywhere else in the body. The patient then underwent right functional neck dissection and the histopathology report came as positive for papillary carcinoma metastasis only in right level III cervical lymph

node. The patient is in regular follow up and is doing well.

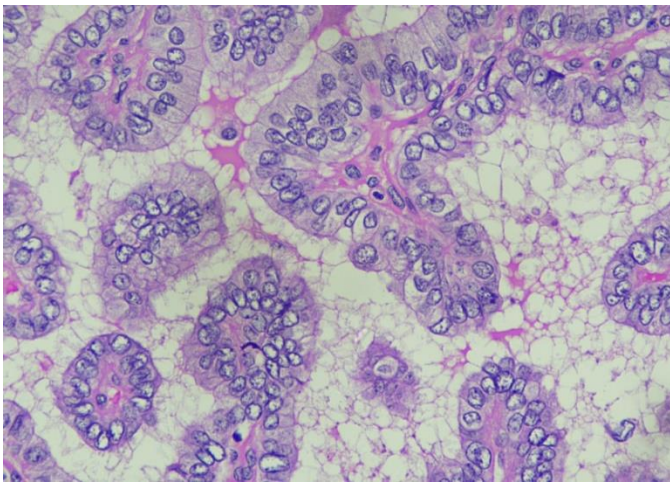


Image 3: Histopathology showing tumour cells arranged in branching papillae with nuclei showing chromatin clearing and occasional grooves.

Discussion

Though thyroglossal duct cyst is a common midline neck mass, malignant transformation is seen in <1%, of which the most common type is papillary carcinoma, as in our case. It usually presents in 4th to 6th decade with female to male ratio of 3:1 showing female preponderance.⁸ Karbek B et al reported a 40 year old male patient with coexistence of thyroglossal duct papillary carcinoma and thyroid papillary microcarcinoma who was treated with sistrunks procedure with total thyroidectomy and bilateral neck dissection.⁹ Our patient was a 40 year old female.

The cause of malignant transformation of thyroglossal cyst is unknown and there are no predisposing factors. Widely, two theories explain the thyrogenic origin of thyroglossal duct cyst carcinoma. First is the denovo theory based on the fact that ectopic thyroid tissue can be identified histopathologically in 62% of cases and the second being the metastatic theory. This theory suggests that thyroglossal duct cyst carcinoma is the metastasis from an occult primary thyroid gland supported by the fact that papillary carcinoma is multifocal in nature.⁵ Mobini et al.¹⁰

accepted that squamous cell carcinoma is probably the only true carcinoma of thyroglossal duct cyst as other malignancies develop in ectopic thyroid tissue.

Most of the thyroglossal duct papillary carcinoma patients are asymptomatic. They might present only with midline neck mass. Carcinoma should be suspected with any recent change in clinical behaviour. Proper clinical examination, radiological investigations like ultrasound and computed tomography or magnetic resonance imaging will guide in suspecting a malignant change. Fine needle aspiration cytology is widely used for diagnosis preoperatively. But in our case, FNAC showed features of papillary carcinoma of right lobe thyroid nodule whereas CT and MRI neck showed features consistent with thyroid malignancy with metastatic enlarged left level III node and possibility of neoplasm of thyroglossal duct cyst. It was based on these findings, the patient was subjected to total thyroidectomy with sistrunks procedure and left selective neck dissection. But to our surprise, the histopathology report came as papillary carcinoma of thyroglossal duct cyst alone with no evidence of malignancy in thyroid gland.

The management of thyroglossal duct cyst is not well defined in literature due to its rare nature. The crucial problem regarding the treatment strategy is the management of thyroid gland and cervical lymph nodes. LiVolsi VA et al stated that Sistrunks operation alone was adequate if papillary carcinoma was entirely within thyroglossal cyst.¹¹ O'Connell et al in his study suggests the use of imaging and pathological studies like FNAC preoperatively to decide whether to proceed with total thyroidectomy and or lymphnode dissection if significant doubt exists. They also explained the need for additional therapy of I-131 ablation in case of thyroid gland involvement, soft tissue extension and lymph node involvement.¹² So was in our case, the patient underwent I-131 ablation therapy 6 weeks post surgery on thyroid hormone suppression.

No standard treatment protocol have been reported regarding the management of regional lymph node metastasis in case of papillary carcinoma of thyroglossal cyst as case reports with neck metastasis is limited with incidence of less than 8%.⁷ The pivotal doubt is whether to undergo unilateral or bilateral neck dissection and whether one should perform modified neck dissection or selective neck dissection. In our case, the patient had to undergo a second surgery (right functional neck dissection) 3 months following the first surgery and radioiodine ablation as she developed right level III cervical lymph node metastasis later.

Conclusion

In conclusion, papillary carcinoma of thyroglossal cyst and also its coexistence with thyroid papillary carcinoma presents a unique diagnostic conundrum that demands proper approach. This case underscores the importance of thorough clinical evaluation, advanced imaging techniques and histopathological examinations in unraveling complex thyroid pathologies. Based on our experience, we recommend Sistrunks procedure and total thyroidectomy with bilateral neck dissection as the treatment for concurrent papillary carcinoma of thyroid and thyroglossal duct cyst.

References

1. Gomez-Alvarez LR, Trevino-Lozano MA et al. Papillary thyroid carcinoma from a thyroglossal cyst: case series. *J Surg Case Rep*. Feb 2022; 2022(2): 1-4.
2. Fernandez JF, Ordonez NG, Schultz PN, Samaan NA, Hickey RC. Thyroglossal duct carcinoma. *Surgery* 1991;110:928-34.
3. Vera-Sempere F, Tur J, Jaén J, Perolada JM, Morera C. Papillary thyroid carcinoma arising in the wall of a thyroglossal duct cyst. *Acta Otorhinolaryngol Belg*. 1998;52:49–54.
4. Liaw J, Cochran E, Wilson MN. Primary Papillary Thyroid cancer of a Thyroglossal duct cyst. *Ear, Nose & Throat Journal*. 2019;98(3):136-138.
5. Balalaa N, Megahed M, Ashari MA, Branicki F. Thyroglossal duct cyst papillary carcinoma. *Case Rep Oncol*. 2011 Jan 29;4(1):39-43.
6. Iftikhar H, Ikram M, Nathaniel KR, Muhammad AY. Papillary Thyroid Carcinoma within Thyroglossal Duct Cyst: Case series and Literature Review. *Introduction Arch Otorhinolaryngol*. 2018 Jul; 22(3): 253-255.
7. Gebbia, V., Di Gregorio, C. & Attard, M. Thyroglossal duct cyst carcinoma with concurrent thyroid carcinoma: a case report. *J Med Case Reports*. 2, 132 (2008).
8. Hakeem AH, Hakeem IH, Pradhan SA, Hathiram B et al. Papillary carcinoma in a thyroglossal duct cyst with concurrent thyroid microcarcinoma and neck nodal metastasis. *Indian J Surg Oncol*. Sep 2013; 4(3):229–232.
9. Karbek B et al. Coexistence of thyroglossal duct papillary carcinoma and thyroid papillary microcarcinoma. *Turk Jem* 2013; 17: 127-128.
10. Mobini J, Krouse TB, Klinghoffer JF. Squamous cell carcinoma arising in a thyroglossal duct cyst. *Am Surg* 1974;40:290–294.
11. LiVolsi VA, Perzin KH, Savetsky L. Carcinoma arising in median ectopic thyroid. *Cancer* 34:1974;1303–1315.
12. O'Connell M, Grixti M, Harmer C. Thyroglossal duct carcinoma: presentation and management including eight cases reports. *Clin Oncol*. 1998;10:186–190.