

Synchronous squamous cell carcinoma and papillary thyroid carcinoma arising from the thyroglossal duct remnant: Case report and a review of the literature

SAGE Open Medical Case Reports
Volume 8: 1–5
© The Author(s) 2020
Article reuse guidelines:
sagepub.com/journals-permissions
DOI: 10.1177/2050313X20917846
journals.sagepub.com/home/sco



Marco Puccini^{1,2} , Nicolò Roffi¹, Valentina Pucci¹,
Giacomo Fiacchini³ , Clara Ugolini⁴ and Piero Bucciatti²

Abstract

Squamous cell carcinoma and papillary thyroid carcinoma simultaneously spreading from the thyroglossal duct remnant (TGDR) is a very rare event. The recognition of this condition allows a correct management and treatment, offering the best chances of cure to the patient. We describe the case of a 42-year-old woman who noticed a right-sided lump in her neck. An ultrasound scan confirmed multiple clusters of enlarged lymph nodes on the right side associated to a pre-hyoidal solid nodule. The thyroid gland was normal. Fine-needle aspiration cytology on two nodes revealed distinct metastases from squamous cell carcinoma and from papillary thyroid carcinoma. A careful screening for other head and neck tumors was negative. She underwent a Sistrunk procedure, total thyroidectomy and right lateral lymphadenectomy with en bloc jugular vein resection. On histology, a 2 cm papillary and a small squamous cell carcinoma of the TGDR were documented, with nodal metastases from both primaries. We report the overall management strategy, treatment and outcome at 26-month follow-up, and a review of the literature.

Keywords

Thyroglossal duct neoplasms, thyroglossal duct remnant, squamous cell carcinoma, papillary thyroid carcinoma, head and neck cancer

Date received: 6 June 2019; accepted: 26 February 2020

Introduction

Thyroglossal duct remnant (TGDR) derived lesions are relatively common¹ but their malignant transformation is not. The most frequent tumor of the TGDR is papillary thyroid carcinoma,² derived from the follicular cells, while tumors arising from the squamous epithelial lining are much rarer.³ Squamous cell carcinoma (SCC) and papillary thyroid carcinoma (PTC) simultaneously arising from TGDR are exceedingly rare (only three cases reported in the literature). As a consequence, their origin is disputed and treatment and prognosis are uncertain.

We report the case of a woman affected by SCC and PTC of the TGDR with metastases to the cervical lymph nodes, with details of her multidisciplinary management, and we review the literature.

Case

A 42-year-old Caucasian woman was referred to us for a cervical midline mass of 15 mm associated to lymph nodal

metastases from SCC and PTC in her right neck. The history began with occasional auto palpation of right neck lumps. The initial work-up included a sonography of the neck that confirmed the presence of enlarged lymph nodes on the jugular (max diameter 16 mm with calcifications) and accessory lymphatic chains, a normal thyroid gland and a 15 mm median

¹Department of Surgical, Medical, Molecular Pathology and Critical Area, University of Pisa, Pisa, Italy

²General Surgery Unit, Azienda Ospedaliero-Universitaria Pisana, Pisa, Italy

³Otolaryngology, Audiology and Phoniatic Operative Unit, Department of Surgical, Medical, Molecular Pathology and Critical Care Medicine, Azienda Ospedaliero-Universitaria Pisana (AOUP), University of Pisa, Italy

⁴Section of Pathology, Department of Laboratory Medicine, Azienda Ospedaliero-Universitaria Pisana, Pisa, Italy

Corresponding Author:

Marco Puccini, Department of Surgical, Medical, Molecular Pathology and Critical Area, University of Pisa, via Paradisa 2, 56124 Pisa, Italy.

Email: marco.puccini@med.unipi.it



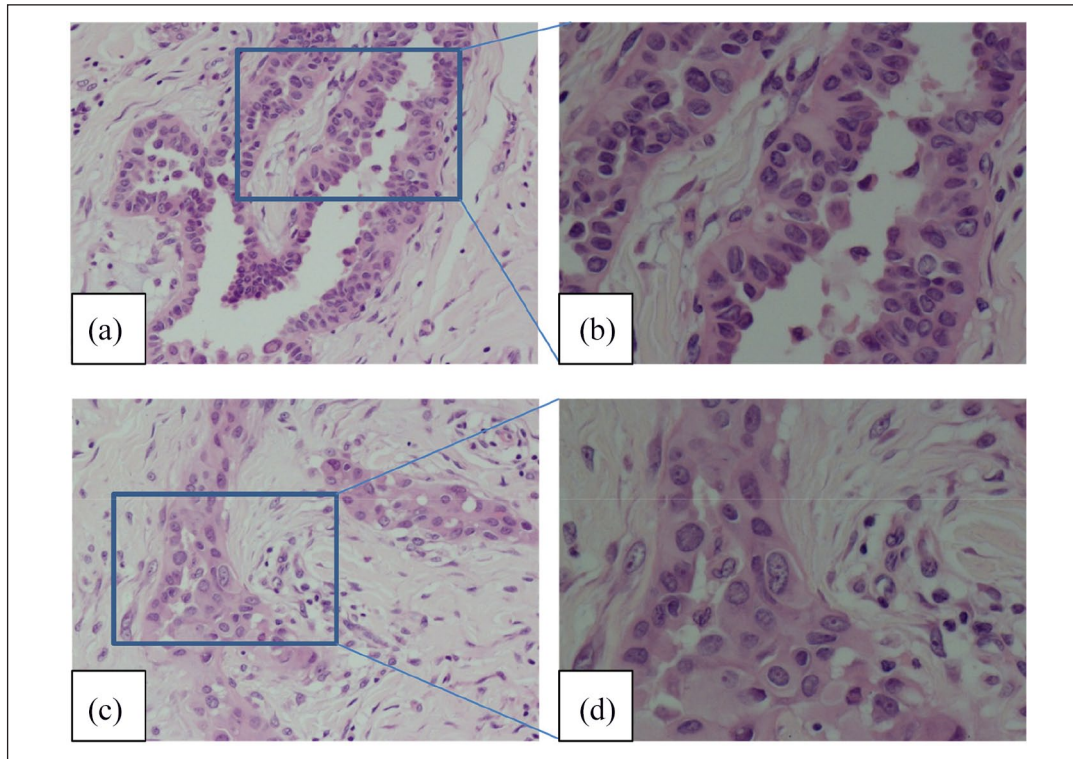


Figure 1. (a) Papillary structure (20× magnification, hematoxylin-eosin staining). (b) Papillary thyroid cancer nuclear features (pleomorphism, elongation, grooves) (40× magnification, hematoxylin-eosin staining). (c) Islands of moderately differentiated squamous cell carcinoma (20× magnification, hematoxylin-eosin staining). (d) In detail large squamous cells with polygonal shape and large cytoplasm (40× magnification, hematoxylin-eosin staining).

lump at the level of the hyoid bone. She had a previous history of cytomegalovirus (CMV) and Epstein-Barr virus (EBV) infection. The fine-needle aspiration cytology (FNAC) on a calcified lymph node at level II revealed a PTC metastasis (immuno-phenotype TTF1 +, TG + and CK7 +), while the sample on a lymph node at level III revealed a scarcely differentiated SCC metastasis (immuno-phenotype p63+, p16- and EBV/LMP-). Ear, nose and throat (ENT) evaluation with fiberoptic transnasal laryngoscopy was negative for other head and neck primary tumors. She also performed a computed tomographic scan of neck, thorax and abdomen that confirmed a 15 × 8 mm lymph node without apparent cleavage from the common jugular vein and the deep surface of the sterno-cleido-mastoid muscle, other enlarged lymph nodes at levels IIA, III, IV, VA, VB and VII (maximum diameters from 10 to 18 mm) and a 15 mm pseudo-nodular lump attached to the central portion of the hyoid bone. No abnormalities were detected in her thorax and abdomen.

Our diagnostic hypothesis was SCC and PTC simultaneously arising from TGDR. She underwent a Sistrunk operation, total thyroidectomy, and right lateral neck dissection (levels II-VII) with an en-block resection of the jugular vein. Histology confirmed a 2 cm PTC of the TGDR, classical type, infiltrating the surrounding soft tissues and the hyoid bone (Figure 1(a) and (b)) and adjacent

1 cm moderately differentiated SCC (Figure 1(c) and (d)) infiltrating the surrounding muscles, with diffuse lymphatic embolism. The thyroid gland was normal. The accessory chain harbored 5 lymph nodes with metastasis from SCC; the right lateral compartment also contained 25 lymph nodes with massive metastasis from SCC (Figure 2(b) and (d)) and 2 lymph nodes with mixed metastases from SCC and PTC. The 18 mm lymph node on the jugular vein was a metastasis from PTC (Figure 2(a) and (c)) infiltrating the soft tissues.

Postoperative course was uneventful, and she was discharged without apparent nervous deficits or fluid collections. She started suppressive thyroid hormone treatment. A multidisciplinary discussion of the case indicated an evaluation with a magnetic resonance imaging (MRI) of the head and neck and a PET-FDG of the whole body (both negative for suspicious findings), followed by adjuvant chemo-radiation treatment. She underwent Cisplatin 40 mg/m² qw (7 cycles) and 60 Gy external beam radiation in 30 seats with tolerable side effects. After 8 months, a new PET-FDG scan did not reveal suspicious hypermetabolic spots. One year after the operation, she complained of dry mouth and neck stiffness. The MRI at 18 months from surgery was negative and the multidisciplinary team recommended surveillance. She is currently alive with no evidence of disease

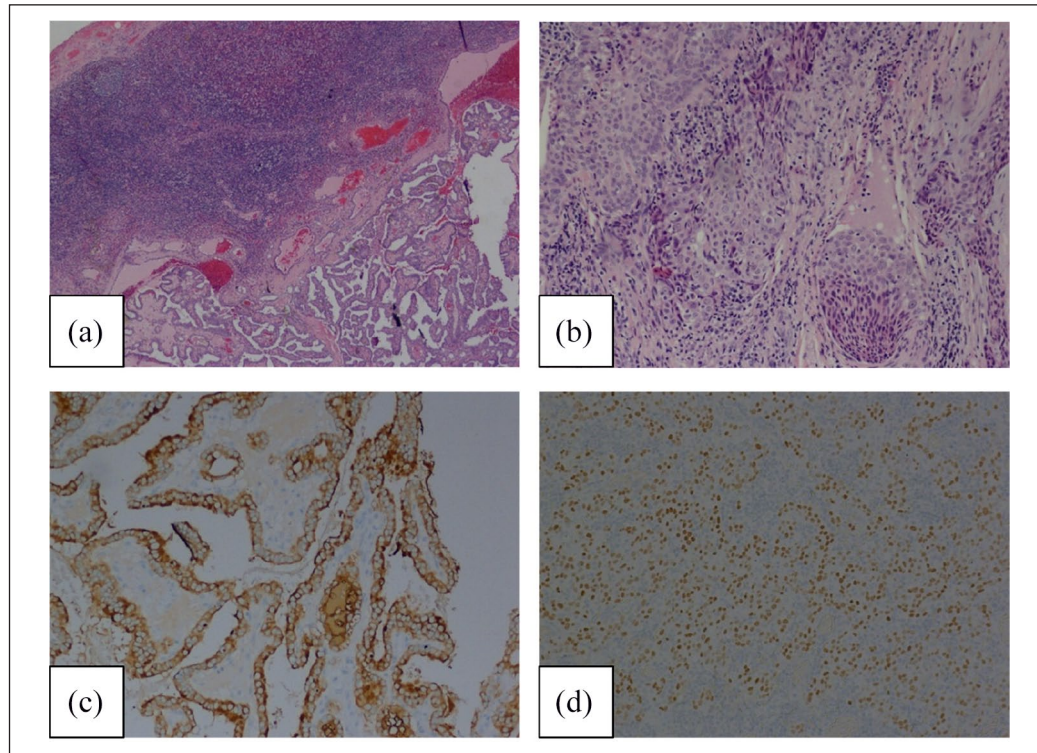


Figure 2. (a) Lymph node metastasis of papillary thyroid carcinoma (2× magnification, hematoxylin-eosin staining). (b) Lymph node metastasis of squamous carcinoma (10× magnification, hematoxylin-eosin staining). (c) Thyroglobulin expression of metastasis from papillary thyroid carcinoma (10× magnification, hematoxylin-eosin staining). (d) P40 expression of metastasis from squamous cell carcinoma (10× magnification, hematoxylin-eosin staining).

(last follow-up at 2 years from the operation). EBV tests (InformEBER Probe, Roche-Ventana) on the SCC of the TGDR and on a metastatic lymph node were positive, while human papillomavirus (HPV) tests (INFORM HPV III Family 16 Probe and INFORM HPV II Family 6 Probe) were negative. She did not yet undergo 131-iodine radiometabolic treatment, as her TG level is low (0.7 ng/mL).

Discussion

TGDR is the result of a missed involution of the tract of descent of the thyroid from the point of original migration from the endoderm (at the foramen cecum, base of the tongue) to its final destination in the anterior neck. The TGDR contains stratified epithelial squamous cells, pseudo-stratified columnar respiratory epithelium and follicular cells.^{2,4} It can form cysts (the most common situation, especially in children), fistulas, solid lumps, that can be observed along the whole tract, even if they are usually found in proximity of the hyoid bone. Tumors can rarely arise from TGDR (about 1% of cases), and most of them arise from the follicular component (95% of reported cases), while only 5% are SCC.⁵ In the children, TGDR derivatives represent the most common congenital midline neck mass, and usually consist of a benign thyroglossal duct cyst. In the adults, midline neck lumps are present in about 7% of population, and most

of tumors are reported in adults older than 30 years old.⁶ As a consequence, midline solid masses in the adults are at higher risk of malignancy and deserve a careful preoperative evaluation including a detailed ultrasound examination of the neck with echo-guided FNAC of all suspicious findings before proceeding with the management. The suspicion of malignancy should rise in case of hard, fixed lumps, irregular borders, concurrent enlarged lymph nodes in the neck and in subjects of age over 30 years (SCC develops at an even older age, average being over 50 years old).⁷

PTC from TGDR are rare and usually have a good prognosis after surgical treatment. SCC is even rarer (about 26 cases reported in the literature)³ and biologically more aggressive than PTC; the treatment is more complex, usually requiring adjuvant therapy, and the prognosis is worse, sometime ending with the death of the patient.^{4,5}

Concurrent SCC and PTC are extremely rare (four cases described including ours): Salve Ronan et al.⁸ described two cases of thyroglossal duct cyst, one of whom with a SCC component. She was a 19-year-old black woman with a 3-year history of a firm 6 cm mass and no other findings. The clinical evolution of the patient was not reported. Winkle Kwan et al.⁹ reported a 38-year-old man with a small component of moderately differentiated squamous cell carcinoma with early invasion, clearly separate from the PTC component, who was referred to him after the

removal of the mass along with the central portion of the hyoid bone. No other neck abnormalities were noticed, and they managed the patient by removing the thyroid and administering ablative radioactive iodine first, followed by the administration of 51 Gy in 20 fractions as adjuvant external beam radiotherapy to treat the SCC component (in an inverse order respect to us). Three years later, the patient was still alive without disease. Finally, Kiyoshi Gomi et al.⁶ reported an 11-year-old girl with a 9-month history of a 1.7 cm anterior-midline neck mass and no other findings. She underwent an open biopsy with a diagnosis of PTC with squamous metaplasia and was treated by a Sistrunk operation and regional lymphadenectomy. The diagnosis was confirmed with invasion of surrounding soft tissues, and metastases from PTC component were found in submandibular lymph nodes. This young patient received postoperative external beam radiotherapy, followed by thyroid hormone suppression therapy. The follow-up was 10 months with no signs of relapse. Other authors reported cases of adenosquamous carcinoma.^{7,10}

In our patient, after radical surgery the multidisciplinary team started adjuvant treatment for invasive SCC, according to the best current knowledge about head and neck tumors,¹¹ since the squamous component of the disease was predominant; the follicular component will be eventually treated later by radioiodine administration due to the presence of lymph node metastases from PTC.^{12,13} At 2-year follow-up, there are neither signs of recurrence nor any other occult source of head and neck SCC.

An interesting aspect to be discussed is the positive EBV tests on the primary SCC and on a metastatic lymph node, which was negative at the preoperative FNAC (performed on a lymph node at III level). If these data had been known before surgery, we could have hypothesized an occult nasopharyngeal primary carcinoma¹⁴ and we could have proposed multiple biopsies in this anatomic site. Against this hypothesis, there are the negative preoperative MRI and postoperative PET-TC imaging for lesions in the nasopharynx as well as the negative ENT evaluation and the fact that the subject is currently alive without disease 2 years after her surgery despite the nasopharynx was not included in the irradiation field. It is our opinion that the patient was not affected by an occult nasopharyngeal carcinoma, but TGDR was the primary site of her SCC; the presence of positive EBV testing in a metastatic lymph node was not correlated to this moderately differentiated SCC.

Conclusion

To the best of our knowledge, this is the fourth case ever reported of a simultaneous papillary thyroid and squamous cell carcinoma of the thyroglossal duct. The message is: in presence of a solid lump in the medial neck above the thyroid isthmus, especially if it is fixed, with irregular borders or associated to enlarged neck lymph nodes, it should be

investigated properly to exclude malignancy by a multidisciplinary working group in order to offer the patient the best chance of cure. The association of a follicular and a squamous cell carcinoma is possible though rare; an aggressive adjuvant treatment of SCC should precede that of differentiated thyroid carcinoma.

Declaration of conflicting interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Ethics approval

Our institution does not require ethical approval for reporting individual cases or case series.

Funding


The author(s) disclosed receipt of the following financial support for the research, authorship, and/or publication of this article: This report was partly supported for the publication by the University of Pisa.

Informed consent

Written informed consent was obtained from the patient(s) for their anonymized information to be published in this article.

ORCID iDs

Marco Puccini  <https://orcid.org/0000-0002-1678-8125>

Giacomo Fiacchini  <https://orcid.org/0000-0001-9832-4799>

References

1. Rayess HM, Monk I, Svider PF, et al. Thyroglossal duct cyst carcinoma: a systematic review of clinical features and outcomes. *Otolaryngol Head Neck Surg* 2017; 156(5): 794–802.
2. Wei S, LiVolsi VA and Baloch ZW. Pathology of thyroglossal duct: an institutional experience. *Endocr Pathol* 2015; 26(1): 75–79.
3. Huang Q, Shen Y, Wang AY, et al. Squamous cell carcinoma arising from a thyroglossal duct cyst: a case report and review of the literature. *SAGE Open Med Case Rep* 2018; 6: 2050313X18767050.
4. Carter Y, Yeutter N and Mazeh H. Thyroglossal duct remnant carcinoma: beyond the Sistrunk procedure. *Surg Oncol* 2014; 23(3): 161–166.
5. Motamed M and McGlashan JA. Thyroglossal duct carcinoma. *Curr Opin Otolaryngol Head Neck Surg* 2004; 12: 106–109.
6. Gomi K, Kitagawa N, Usui Y, et al. Papillary carcinoma with extensive squamous metaplasia arising from thyroglossal duct cyst in an 11-year-old girl: significance of differentiation from squamous cell carcinoma: a case report. *J Pediatr Surg* 2011; 46(4): e1–e4.
7. Chang YS, Su HH and Ho SP. Adenosquamous carcinoma arising from a thyroglossal duct cyst: a case report. *Oncol Lett* 2016; 11: 2668–2672.
8. Ronan SG, Deutsch E and Ghosh L. Thyroglossal duct carcinomas: light and electron microscopic studies. *Head Neck Surg* 1986; 8(3): 222–225.

9. Kwan WB, Liu FF, Banerjee D, et al. Concurrent papillary and squamous carcinoma in a thyroglossal duct cyst: a case report. *Can J Surg* 1996; 39(4): 328–332.
10. Kinoshita N, Abe K, Sainoo Y, et al. Adenosquamous carcinoma arising in a thyroglossal duct cyst: report of a case. *Surg Today* 2011; 41: 533–536.
11. Iocca O, Farcomeni A, Di Rocco A, et al. Locally advanced squamous cell carcinoma of the head and neck: a systematic review and Bayesian network meta-analysis of the currently available treatment options. *Oral Oncol* 2018; 80: 40–51.
12. Patel SG, Escrig M, Shaha AR, et al. Management of well-differentiated thyroid carcinoma presenting within a thyroglossal duct cyst. *J Surg Oncol* 2002; 79(3): 134–139; discussion 140–141.
13. Miccoli P, Pacini F, Basolo S, et al. Thyroid carcinoma in a thyroglossal duct cyst: tumor resection alone or a total thyroidectomy? *Ann Chir* 1998; 52: 452–454.
14. Katabi N and Lewis JS. Update from the 4th Edition of the World Health Organization classification of head and neck tumours: what is new in the 2017 WHO blue book for tumors and tumor-like lesions of the neck and lymph nodes. *Head Neck Pathol* 2017; 11(1): 48–54.