Concurrent papillary and squamous carcinoma in a thyroglossal duct cyst: a case report

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Carcinomas of thyroglossal duct cysts are rare. Most are papillary carcinomas; only about 5% are squamous cell carcinomas. Only one case of mixed papillary and squamous cell carcinoma of a thyroglossal duct cyst has been reported so far. The authors present a second case, that of a 38-year-old man who was first seen with a midline neck lump. It was diagnosed clinically as a thyroglossal duct cyst and was locally excised. Pathological examination showed both a concurrent papillary carcinoma and a squamous cell carcinoma. Treatment consisted of a near-total thyroidectomy, ablative radioactive iodine and adjuvant external radiation therapy. The authors review the literature and explain the rationale behind their choice of treatment.

Les cancers de kystes du canal thyréo-glosse sont rares. Il s'agit de carcinomes papillaires dans la plupart des cas. Environ 5 % seulement sont des épithéliomas spinocellulaires. On n'a signalé jusqu'à maintenant qu'un seul cas d'épithélioma spinocellulaire et de carcinome papillaire mixte d'un kyste du canal thyréo-glosse. Les auteurs décrivent un deuxième cas, celui d'un homme de 38 ans dont le premier examen a révélé la présence d'une masse à la ligne médiane du cou. On a posé un diagnostic clinique de kyste du canal thyréo-glosse qui a été excisé au niveau local. L'examen pathologique a révélé un carcinome papillaire conjugué à un épithélioma spinocellulaire. Le traitement a consisté en une thyroïdectomie quasi totale, une ablation à l'iode radioactif et une radiothérapie externe. Les auteurs présentent une recension des écrits et justifient leur choix de traitement.

Only about 150 cases of carcinoma arising in a thyroglossal duct cyst have been reported,[1] of which 85% were papillary carcinomas of the thyroid type. Squamous cell carcinomas constitute approximately 5% of such cases.[1] There has been one previous report of a concurrent papillary and squamous cell carcinoma in a thyroglossal duct cyst,[2] but it focussed primarily on the pathological features of the case and gave little clinical information, particularly on treatment and follow-up. We describe the case of a patient who had a concurrent papillary and squamous carcinoma of the thyroglossal duct cyst, his treatment plan and outcome.

Case report

A 38-year-old man was referred to the Princess Margaret Hospital (PMH) in January 1992 after local excision of a thyroglossal cyst. He presented initially with a midline neck lump at the level of the hyoid bone. It was associated with a slight sensation of pressure but was otherwise asymptomatic. There was no history of chronic inflammation in his head and neck region or of previous radiation exposure. He smoked two packets of cigarettes per week. At operation, the cyst was found to be attached to the hyoid bone, and the cyst along with the middle portion of the hyoid bone was excised.

Microscopic examination revealed a cystic structure with chronic granulomatous inflammation within the cyst wall. There was a single focus of pro-

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liferation of atypical squamous cells with dysplastic cells and microinvasion into the underlying stroma (Fig. 1). Separate from the squamous cell carcinoma, there were small glandular structures with papillary ingrowths associated with psammoma bodies (Fig. 2). Immunohistochemical staining confirmed thyroglobulin expression in the papillary component but not in the squamous component. The squamous component expressed high-molecular-weight keratin. The overall picture was one of moderately differentiated squamous cell carcinoma with early invasion, coexisting with multifocal papillary carcinoma of the thyroid type. The squamous and papillary carcinoma components were quite separate and distinct, and the pattern did not seem to arise from squamous metaplasia of the papillary carcinomatous component. Because of the invasive nature of the squamous component, it was not considered merely to represent dysplasia of metaplastic thyroglossal duct epithelium. Some of the papillary carcinoma foci were very close to the outer margins of excision, and a positive margin could not be excluded.

At the time of consultation at the PMH, there was no evidence of disease on examination. Specifically there was no cervical lymphadenopathy or abnormalities of the thyroid gland. The findings on radioactive iodine uptake (RAIU) scanning, ultrasonography of the neck, chest radiography and computed tomography of the neck were all normal. Thyroid stimulating hormone, free thyroxine index and total T_4 levels were also normal.

Our plan of management was to approach this problem by addressing each component separately. Given that this lesion was, in part, a papillary carcinoma of the thyroid type in a thyroglossal duct cyst, there was a 10% to 14% risk that there was another focus of carcinoma in the thyroid gland.[1] Therefore, we proceeded first to a near-total thyroidectomy, which revealed a normal thyroid gland with several small lymph nodes at the thyroid isthmus. One of the lymph nodes had a focus that suggested involvement by the follicular variant of papillary carcinoma. A post-thyroidectomy RAIU scan indicated a focus of uptake in the thyroid bed, compatible with a small amount of remnant thyroid tissue. Therefore, the patient was given ablative radioactive iodine (^{131}I) treatment with an activity of 1.07 GBq (29 mCi).

Because there was also a separate component of squamous cell carcinoma in the tumour and there was concern about the completeness of excision, we administered a course of adjuvant external beam radiotherapy to the initial operative site. The target volume encompassed the scar in its entirety, the hyoid bone and a surrounding margin of 3 cm. The technique involved a lateral parallel pair of fields, administering a dose of 51 Gy in 20 fractions over 4 weeks. By this time, the patient was on thyroxine replacement, and he tolerated the external beam therapy well, with only temporary pharyngitis.

At his last follow-up, around 3 years after the initial diagnosis, the patient was well with no evidence of disease.

Discussion

The occurrence of concurrent, separate foci of papillary and squamous cell carcinoma arising in a thyroglossal duct cyst is extremely rare. Only one case of mixed papillary and squamous cell carcinoma has been described in the literature.[2] Ronan, Deutsch and Ghosh[2] described a 19-year-old woman who presented with a 3-year history of a submental mass. A total thyroidectomy and excision of the submental mass were performed. The mass was found to consist of glandu-

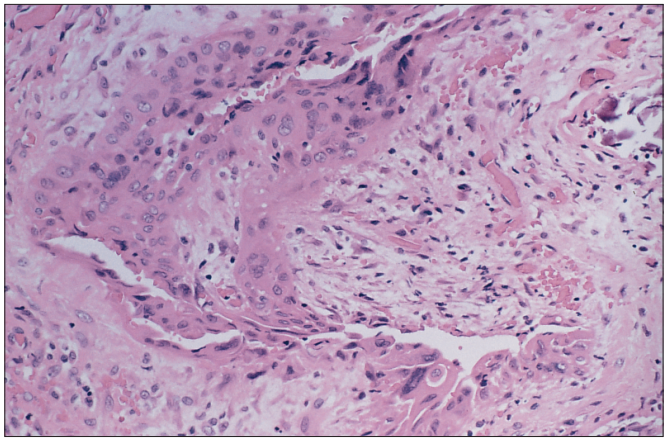

FIG. 1. Squamous cell carcinoma (hematoxylin–eosin, original magnification × 250).

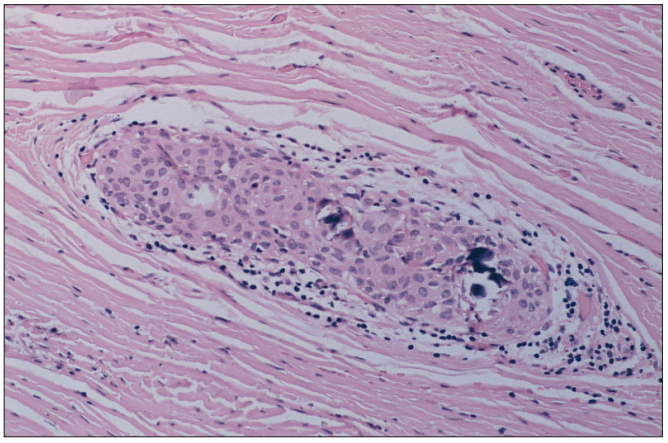

FIG. 2. Papillary carcinoma with psammoma bodies (hematoxylin–eosin, original magnification × 100).

lar proliferations with papillary infoldings and with areas of transition from neoplastic glandular to squamous epithelium. Squamous carcinoma cells were seen between the muscle fibres adjacent to the hyoid bone. This case probably represents squamous differentiation arising in a papillary carcinoma, a phenomenon that occurs in 15% to 45% of thyroid papillary carcinomas.[3] This patient evidently did not receive any adjuvant treatment, and no further clinical information was provided.

Our treatment rationale was based on the available literature for papillary or squamous carcinomas occurring in a thyroglossal duct cyst. Two recent reviews on papillary carcinomas of thyroglossal duct cysts were compiled by Weiss and Orlich[4] and Renard and colleagues[1] Of the 146 patients reviewed by Renard and colleagues, the thyroid gland was examined histologically in 43. In six cases (14%), papillary adenocarcinoma was found in the thyroid gland. The prognosis of papillary carcinoma arising from a thyroglossal duct cyst appears to be generally favourable. It is thought to be similar to that of papillary carcinoma of the thyroid gland, having a cure rate in excess of 95%.[5] McNicoll and colleagues[6] reviewed 48 patients who were treated with the Sistrunk procedure alone. No patient experienced a recurrence after 4 years. In our patient, because of his relatively young age and the risk (14%) of a concomitant papillary carcinoma in his thyroid gland, we elected to perform a near-total thyroidectomy. To facilitate follow-up with radioiodine scanning and serum thyroglobulin, RAIU therapy was given to ablate the thyroid remnant. It has been shown that the recurrence rate for papillary thyroid cancer is lowest after total ablation of thyroid tissue with thyroidectomy and radioactive iodine.[7]

Patients with squamous cell carcinoma of a thyroglossal duct cyst appear to have a poorer prognosis. The most complete review to date was conducted by Yanagisawa, Eisen and Sasaki.[8] A more detailed scrutiny of the literature revealed that 13 cases in total have been described

Table 1

Summary of Literature on Patients With Squamous Cell Carcinoma of Thyroglossal Duct Cyst

Series	Sex/age, yr	Clinical course	Outcome	XRT
Clute and Smith, 1929[9]	M/56	Received postoperative XRT. Progressed rapidly after XRT	Died 15 mo after diagnosis	Yes
Nachlas, 1950[10]	?	Reviewed 128 cases of TDC. One case of epidermoid carcinoma found. Grade 2 cancer	?	?
Dalgaard and Wetteland, 1956[11]	F/44	Suprahyoid mass for 10 yr. Excised, recurred 13 yr later. Pathology of recurrence = SCC	NED 15 yr after second operation	—
Ruppmann and Georgsson, 1966[12]	F/51	22 years of painless swelling. Recurrent drainage, 5 resections needed. Last resection pathology = SCC	NED 1 yr after last surgery	—
Shepard and Rosenfeld, 1968[13]	F/28	3 years of cervical mass. Excised. Pathology = SCC. Postoperative XRT	Recurred locally with advanced disease. Died 4 yr after diagnosis	Yes
Mobini, Krouse and Klinghoffer, 1974[14]	F/50	1 yr of neck mass. Mass infiltrated isthmus and right lobe of thyroid. Mass, right lobe and isthmus excised with difficulty. Postop XRT	NED 2 yr after treatment	Yes. 67 Gy
Saharia, 1975[15]	F/81	4 mo of neck mass. Cyst, left side of hyoid, strap muscles excised. Neck exploration negative. Pathology = well-differentiated SCC invading surrounding muscle	NED 3 years after treatment	—
Benveniste, Hunter and Cook, 1980[16]	M/75	2 mo of neck swelling. Mass grew rapidly, became fixed to skin in 2 wk. Mass and overlying skin excised. Pathology = SCC infiltrated into strap muscles and subcutaneous tissue. Postop XRT	Necrotic ulcer and tracheal fistula. Biopsy negative for recurrence. Alive 7 mo after presentation	Yes. 55 Gy

(Table I[2,8–19]). Among 11 patients with follow-up information, 4 died of the cancer, and 1 had a necrotic ulcer and tracheal fistula 7 months after presentation. If the patient described by Clute and Smith,[9] treated before 1929, was excluded, 6 of 12 patients received modern-era postoperative radiotherapy. Three of these six patients remained alive with no evidence of disease from 18 months to 3 years after completion of treatment. It is of interest that we also observed a poor prognosis for squamous cell carcinoma of the thyroid gland in a previous review at the Princess Margaret Hospital.[20] Our rationale of administration of postoperative radiotherapy was based on the concern about the completeness of surgery, since the risk of local microscopic residual disease was uncertain. The dose of 51 Gy in 20 fractions over 4 weeks is our standard regimen for squamous cell carcinomas in other head and neck sites.

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Table 1 (continued)

Summary of Literature on Patients With Squamous Cell Carcinoma of Thyroglossal Duct Cyst

Series	Sex/age, yr	Clinical course	Outcome	XRT
White and Talbert, 1982[17]	M/61	Submental mass. Excised. Pathology = 2 cavities: TDC and pseudocystic mass surrounded by SCC	NED	—
Ronan, Deutsch and Ghosh, 1986[2]	F/19	3 yr of submental mass. Thyroidectomy and excision of mass. Mass infiltrated skeletal muscle. Pathology = mixed papillary carcinoma and SCC	?	—
Bosch, Kummer and Hohmann, 1986[18]	M/54	20 yr of midline neck swelling. Surgery: large cystic swelling with destruction of hyoid bone. Only partial excision possible. Postop XRT	Weight loss, local recurrence after 6 mo. Died 7 mo after surgery	Yes
Lustmann, Benoliel and Zeltser, 1989[19]	F/80	3 wk base of tongue pain. Cystic mass on left side of tongue. Radical neck dissection and removal of thyroglossal sinus tract and excision of mid-hyoid bone. Pathology = SCC, 2 LN positive. Cancer infiltrated submandibular gland. Postop XRT	Recurred 5 mo later. Died 2 wk after readministration of XRT	Yes. 50 Gy
Yanagisawa, Eisen and Sasaki, 1992[8]	M/65	10 yr midline neck mass. Local excision of TDC. Chronic draining sinus, recurrence of midline mass. Sistrunk procedure done. Pathology = SCC arising from TDC. Postop XRT	NED after 18 mo	Yes. 60 Gy

XRT = radiotherapy, TDC = thyroglossal duct cyst, ? = not described, SCC = squamous cell carcinoma, NED = no evidence of disease, LN = lymph node

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