Papillary Carcinoma in a Thyroglossal Duct Cyst with Concurrent Thyroid Micro-Carcinoma and Neck Nodal Metastases

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Introduction

Thyroglossal duct cyst (TDC) is most common benign midline mass seen in the neck, a small percentage (<1 %) may undergo malignant transformation [1]. Papillary carcinoma is the most common histologic subtype seen in about 80% of the thyroglossal duct cysts with malignant transformation [1]. Regional lymph node metastases of papillary carcinomas in thyroglossal duct cysts occur much less frequently than in primary cancer of the thyroid, with incidence of less than 7.7 % of cases [2]. Initially most of the diagnoses were made postoperatively from histopathology of the surgical specimen. But now with the use of fine needle aspiration cytology more and more cases of the thyroglossal duct carcinoma are being identified preoperatively.

We report a case of thyroglossal duct carcinoma with neck nodal metastases and a micro-carcinoma of the thyroid

gland a very rare presentation. We also discuss the management of this rare clinical presentation.

Case Report

A 40 year old female presented with 2 years history of a slowly enlarging upper midline neck mass (Fig. 1). Palpation revealed a 5×4 cm soft non-tender cystic mass that moved upward with protrusion of the tongue. There was 3×3 cm right level II cervical lymph node on deep palpation. Thyroid function tests were normal. CT scan revealed 6×4 cm lesion in maximum axial dimension in the upper neck region inferior to the body of the hyoid. Septae with eccentrically located mural nodule with calcifications were also noted in the lesion with homogeneously enhancing right level II and III nodes (Fig. 2). USG guided FNA from the lesion was consistent with papillary thyroid carcinoma and that from the nodes confirmed the metastases. Rest of the work up did not reveal any metastases anywhere in the body. Thyroid gland was normal both clinically and radiologically. Diagnosis of papillary carcinoma in the thyroglossal cyst with neck metastases was made. Total thyroidectomy with Sistrunk's operation which consists of removing the cyst, the mid portion of the hyoid bone and the entire thyroglossal tract with bilateral selective neck dissection was performed (Fig. 3). Left sided selective neck dissection was also done as there were enlarged nodes in left level III and IV. Gross examination disclosed a 6×4.5×2 cm greyish-tan cystic tumour with multilocular cystic areas with papillary excrescences. Right thyroid lobe also revealed a tiny nodule of 0.2×0.2×0.2 cm dimension. Microscopic examination confirmed the diagnosis of papillary carcinoma thyroid in the thyroglossal duct cyst with metastases in multiple nodes in level II, III

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Fig. 1 Preoperative photograph of thyroglossal duct carcinoma

and IV right side (Fig. 4). It also revealed a tiny focus of papillary carcinoma in the right lobe of thyroid.

After 5 weeks post operatively she underwent I131 scan and ablation. She is being regularly followed with Tg levels which are well below the normal limit. Three years post operative she is well controlled without evidence of local or distant metastases.

Discussion

Some form of TDC remnant is seen in 7 % of total population [3]. Although this is a common midline neck mass seen, only <1 % of the TDC may undergo a malignant transformation to carcinoma [1]. TDC carcinoma usually presents in 4th to 6th decade with female preponderance with male to female ratio of 1:3 [4]. Papillary carcinoma is the most common histologic subtype seen and again more often seen in females with male to female ratio of 2:3 [2].

The diagnosis of TDC carcinoma used to be generally made postoperatively by histopathological examination of the resected tissues [5]. Carcinoma must be suspected in

cases of TDC with recent changes in the clinical behaviour. If TDC's are hard, fixed, irregular or have undergone size change with palpable lymph nodes, carcinoma should be suspected. Imaging studies do not allow for full preoperative diagnosis, but will guide a clinician in suspecting a malignant transformation. Calcifications within the cyst wall is a characteristic feature of papillary carcinoma. Cyst wall invasion is seen in 21 % of patients. Imaging in the form of CT scan may be useful in diagnosing the carcinoma in TDC in the preoperative setting according to some authors [6–9]. Wide application of the FNA for the diagnosis of head and masses may be useful in diagnosing carcinoma in TDC preoperatively. Many authors have reported preoperative diagnosis with FNA but the number of cases is limited [10–15]. We also made a diagnosis of thyroglossal duct cancer by the preoperative fine needle aspiration cytology.

Due to the rare nature of the disease the management of TDC carcinoma is not well defined in literature. The most controversial part of the TDC malignancy is whether total thyroidectomy should be done or not. Bosch and LiVolsi stated that when the papillary carcinoma was entirely within the cyst, the Sistrunk procedure was adequate, but when the carcinoma had spread to the capsule of the cyst, wide resection and lymph node dissection were necessary [16, 17]. O'Connell et al. suggest the use of USG and CT scan or MRI with FNA and justify the use of total thyroidectomy and or lymphnode dissection in case significant doubt remains [18]. They also suggest for additional therapy in the form of ^{131}I ablation in case of thyroid gland involvement, soft tissue extension and lymph node involvement as is standard practice for the management of well differentiated thyroid carcinoma.

Patel et al. did not recommend other treatments in low-risk patients (those below 45, with tumor diameter <4 cm, and no sign of extensive metastasis) [19]. Patel and Chu also rejected the use of thyroidectomy when pathological findings were not present in the preoperative ultrasonographical and biochemical examinations, and when additional clinical and pathological findings were absent [19, 20].

Regional lymph node metastasis has been reported in 7.7–12.9 % of the malignant cases [1, 3, 4]. Since the case reports with neck metastases in case of TDC papillary is limited, we do not have standard treatment protocol. Literature is deficient in whether we should choose unilateral or bilateral neck dissection and also if one should perform modified neck dissection or selective nodal excision. Yamada et al. reported a case of TDC papillary carcinoma with bilateral neck metastases. They performed en-bloc resection of submental tumor and concurrent bilateral supraomohyoid neck dissection avoiding total thyroidectomy [15]. In our case we did total thyroidectomy with complete excision of the thyroglossal duct cyst and bilateral selective neck dissection clearing level II, III,

Fig. 2 CT scan axial view at the level of hyoid bone showing cystic swelling with septae and calcification of mural areas

Fig. 3 Surgical specimen of total thyroidectomy, TGD carcinoma and bilateral selective neck dissection

IV, V and VI. She was also treated with ^{131}I ablation and received adjuvant thyroxine as part of treatment.

These cases are followed up like any other differentiated thyroid cancer. In recent years, serum thyroglobulin (Tg) measurement has become a mainstay of surveillance for differentiated thyroid cancer recurrence. Tg is the major protein product of thyroid cell function and is not produced by other cells in the body. If all thyroid cells have been eliminated, serum Tg should be negligibly low. Elevated serum Tg level (0.2 ng/mL) is highly sensitive and specific for recurrence after surgery and radioablation.

Fig. 4 Microphotograph of the papillary carcinoma in the TDC

Conclusion

Although the papillary carcinoma in TDC is rare clinical entity, presence of simultaneous lymph nodes and a microcarcinoma is still unusual. Due to the rare nature of the disease the management is controversial. Although sistrunk's procedure is standard of management, total thyroidectomy with or without lymph node dissection is necessary in case with soft tissue extension, presence of metastatic neck nodes and pathology in thyroid gland. Adjuvant treatment in the form of ^{131}I ablation may also be indicated in such situation. In our case since there was preoperative diagnosis of metastatic TDC papillary carcinoma we performed total thyroidectomy and lymph node dissection along with the sistrunk's procedure.

Conflict of Interest None

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