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Chemodectoma in A Patient of Papillary Thyroid Carcinoma: A Case Report

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Abstract

Differentiating the neck mass between carotid body paraganglioma and recurrent papillary thyroid carcinoma is occasionally difficult. Methods: We report a 66-year-old woman with previous history of papillary thyroid carcinoma, and a 3-cm-sized, non-tender, rapid-growing mass was found over right submandibular region two years later. Recurrent thyroid cancer was suspected initially. Results: Repeated cytology investigations showed metastatic carcinoma. Neck magnetic resonance imaging (MRI) and biopsy finally revealed paraganglioma over carotid bifurcation. Conclusion: Carotid body tumors is highly related with neck paraganglioma, but difficultly indistinguishable from thyroid lesions or metastatic lymphadenopathy. Biopsy is better diagnostic choice than cytology. It might have related with germline mutations. Surgical removal is first-line treatment, and radiotherapy are suggested if unresectable. (J Intern Med Taiwan 2018; 29: 317-322)

Key Words: Chemodectoma, Carotid body tumor, Neck mass, Papillary thyroid carcinoma, Paraganglioma

Introduction

Carotid body tumors (CBT), chemodectoma, arise from paraganglionic tissue in the bifurcation of the common carotid artery (CA). Generally, they are clinically silent and found incidentally. The carotid body is derived from both mesoderm of the third branchial arch and neural crest ectoderm, while accessory thyroid tissue, arising from endoderm, is usually not to existing in carotid body[1,2]. Magnetic resonance imaging (MRI) plays a pivotal role in the diagnosis and preoperative workup of these tumors[1]. Here we report a 66-year-old woman with previous history of papillary thyroid carcinoma, newly-existing with chemodectoma, which was suspected as recurrent thyroid cancer via cytology. Series publications are reviewed for introducing the forgotten term of carotid body paraganglioma, chemodectoma.

Case report

A 66-year-old woman was noted of palpable thyroid nodules on health check-up. The nodules were elastic without tenderness and estimated smaller than 1 cm. Thyroid ultrasonography revealed hypoechoic nodules with peri-nodular vascularity (Fig. 1A). Aspiration cytology in the left nodular goiter showed atypical cells and right total thyroidectomy and left enucleation thyroidectomy were carried out. Papillary thyroid microcarcinoma was found in the both side thyroid lesions (pT1aN0M0,

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stage I) (Fig. 1B). Radioactive iodine is not indicated for ablation.

Post-operatively, she took Levothyroxin 50mcg once daily for replacement. Serial thyroglobulin levels were all below 0.20 ng/mL in the following months. Serial anti-thyroglobulin antibody levels downgraded from 379.8 IU/mL to 149.7 IU/mL. Two years later, one 3-cm-sized, non-tender, rapid growing, and hard mass was noted over the right lower neck, and neck ultrasonography revealed one hypoechoic mass with prominently intra-nodular vascularity (Fig 1C). Fine needle aspiration cytology was diagnosed as carcinoma (Fig. 1D). Neck MRI was performed because of its multiplanar capability, high sensitivity for contrast enhancement, lack of ionizing radiation, and higher sensitivity than CT in soft tissue tumors. It reported one 4.3 x 2.7 cm well-defined tumor displacing carotid artery, (Fig. 2A & B), and further fluorodeoxyglucose positron emission tomography (FDG-PET) was suggested for the character of rapid growing tumor, and higher standardized uptake value (SUVmax, 8.4) was measured (Fig. 2C). Biopsy pathology showed nested tumor cells with regular-sized nuclei and some vessels in the stroma. (Fig. 3A & B). Paraganglioma in right carotid body was impressed. Considering tumor location and surgical risk (Shamblin classification),[2,3,4] cisplatin-based radiotherapy (6600 cen-

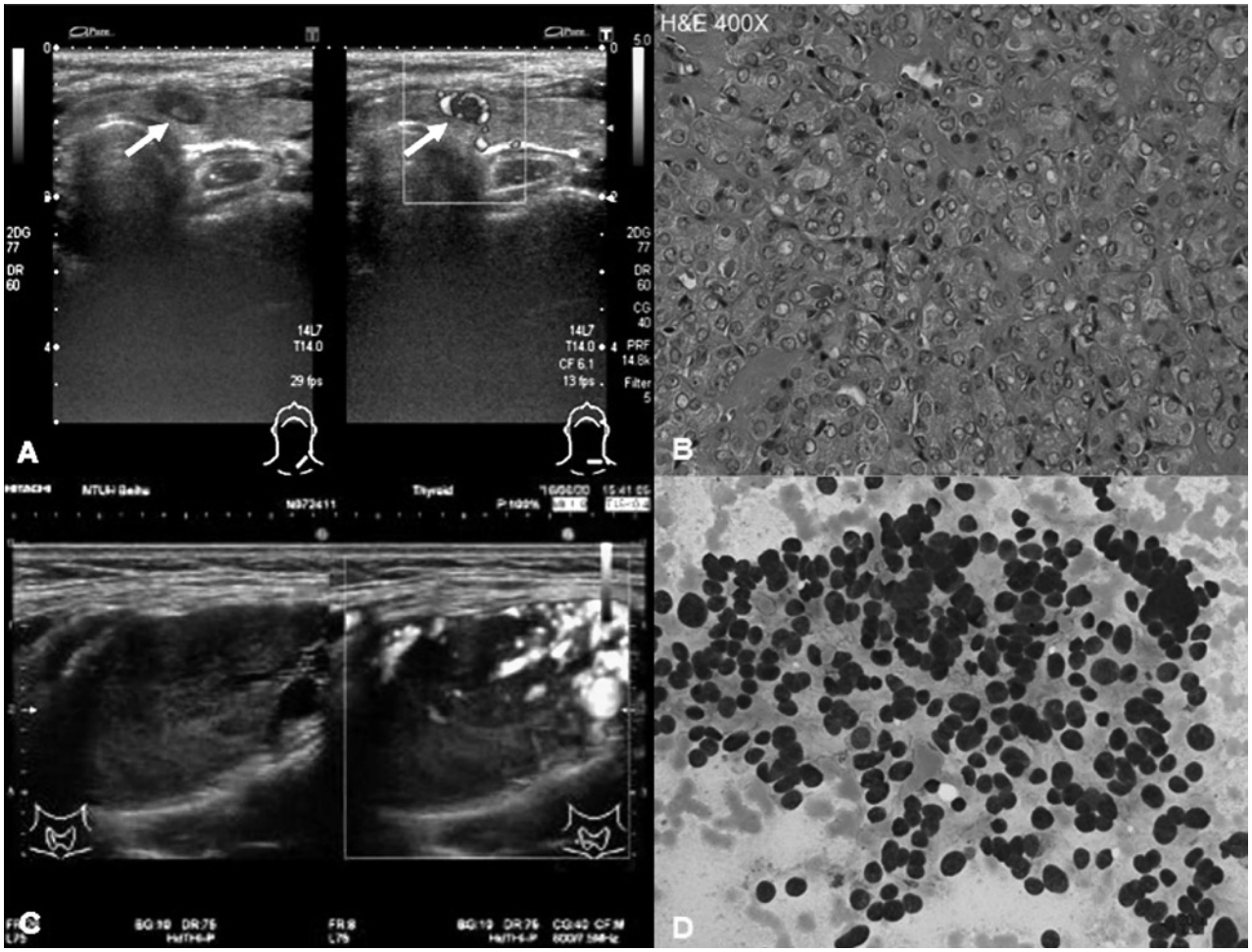

Figure 1. (A) Thyroid sonography revealed a left hypoechoic nodular goiter with peri-nodular hypervascularity, which was proved to be papillary thyroid microcacinoma. (B) Pathology of original papillary thyroid microcarcinoma without metastasis of lymph nodes. (C) Neck sonography revealed one hypoechoic nodular lesion (3.53 x 3.18 x 1.99 cm), over level II area, right side. (D) Cytology of right hypoechoic nodular lesion.

tigray) was given for tumor rapid growing character rather than surgical removal. The patient is under stable disease condition with shrinkage carotid body tumor till now after treatment started for five months.

Discussion

Carotid body tumors (CBT), chemodectoma, account for more than 50% of head and neck paraganglioma and 60 % located at the carotid bifurcation[2]. Additional sites of origin include the jugular bulb, the vagal nerve at the nodose ganglion, and the middle ear. Carotid body paraganglioma often present as slow growing, non-tender neck masses located just anterior to the sternocleidomastoid muscle at the level of the hyoid. Occasionally the tumor mass may transmit the carotid pulse or demonstrate a bruit or thrill.

To establish the diagnosis of a CBT, imaging modalities such as ultrasonography, computed tomography, magnetic resonance imaging (MRI), and digital subtraction angiography are important.[1,2] The MRI findings show low signal intensity in T1-weighted sequences and a hyperintense

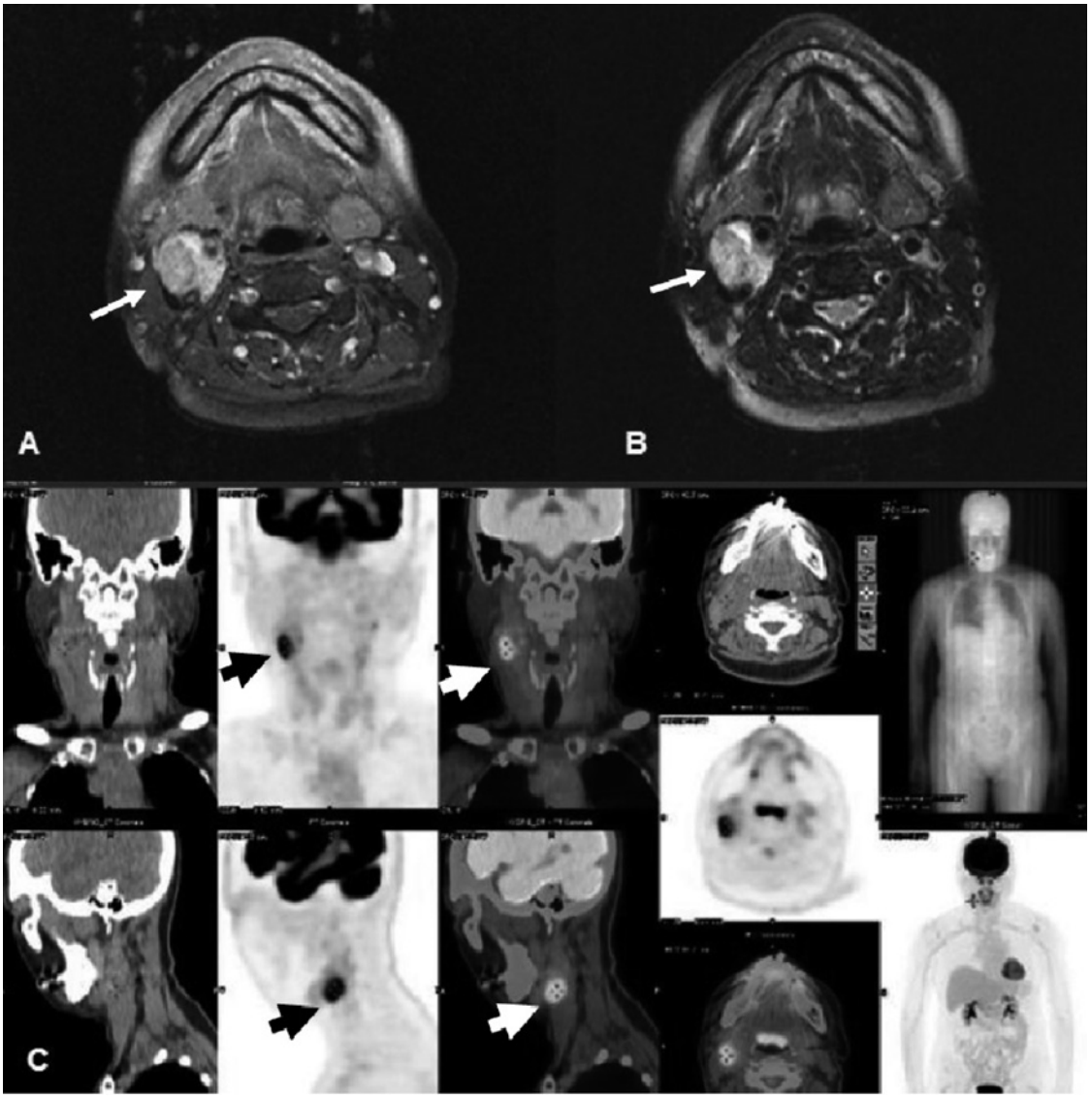

Figure 2. Neck MRI showed a well-defined tumor (A & B, arrow), and FDG-PET showed higher standardized uptake value (SUVmax, 8.4).

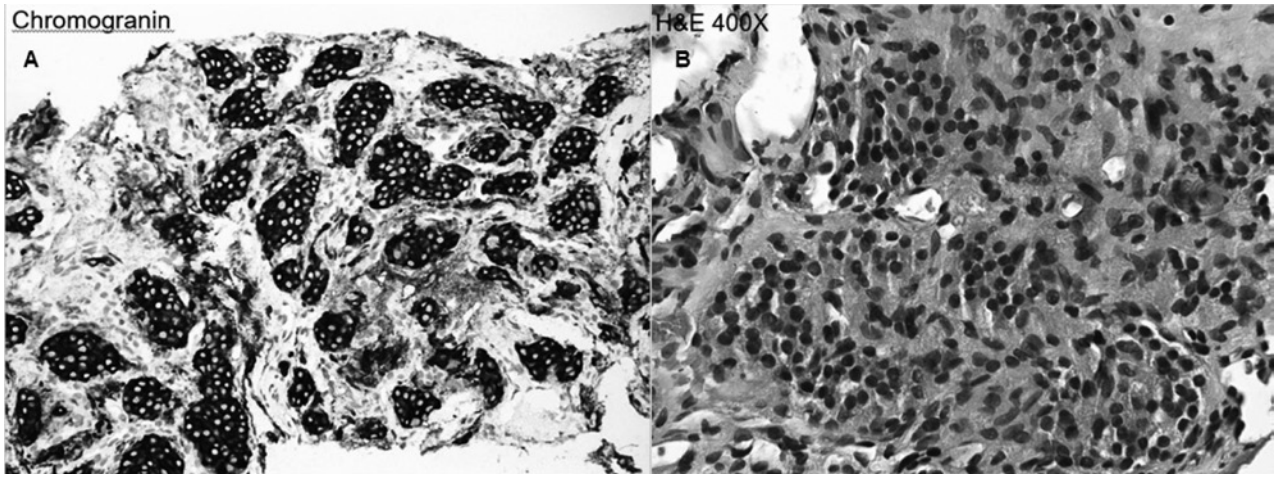

Figure 3. Nested tumor cells with regular-sized nuclei (immunocytochemical stains: synaptophysin (+), chromogranin (+), GATA-3 (+), sustenticular cells S-100 (+), and very scanty Ki-67 (+).

signal in T2-weighted sequences. Shamblin et al,[3] in the 1970s suggested a surgical classification of CBTs into 3 groups, which was according to the gross tumor vessel relationship and was based on the intraoperative findings and postoperative specimen examinations.[2] Group 1 tumors were minimally attached to the vessels and easily resectable. Group 2 tumors seemed partially surrounding the vessel and were more adherent to vessel adventitia, which were difficult to dissect but may be able to careful resection. Group 3 tumors had an intimate adherent relationship to the entire circumference of the carotid bifurcation, and surgical dissection was impossible even in the hands of experienced vascular surgeons. Therefore, required sacrifice of the ICA with vessel replacement.[2,3,4]

Shamblin classification has been widely used as a predictor of vascular morbidity and for surgical policy.[2,3,4] It helps make therapeutic options including surgical excision, conventional radiotherapy, and stereotactic radiosurgery. If the internal carotid is encased in tumor or damaged during resection, immediate repair/replacement should be performed. Radiotherapy is a treatment option for giant and recurrent carotid body paragangliomas, advanced and unresectable, as well as in multiple tumors,[7] and with malignant carotid body paragangliomas metastatic to the regional lymph nodes. [5,6,7] The incidence of cranial nerve injury remains strikingly high, ranging from 20% to 40%.[5,6,7] In 20% of patients the neurological deficits are permanent. [7] Our patient's tumor located at right carotid space, displacing external carotid artery (ECA) and internal carotid artery (ICA), so she received CCRT instead of surgery. The tumor shrinks gradually in series image follow-up.

Papillary thyroid carcinoma is the most common thyroid malignancy. In recent decades, numerous clinical reports have revealed an increased incidence of both papillary thyroid carcinoma (PTC) and papillary thyroid microcarcinoma due to high-resolution ultrasonography[8]. Most PTC patients show a good prognosis with long-term mortality rate around 5-10%. Prior retrospective review in Taiwan, the non-incidental papillary thyroid microcarcinoma (PTMC) presenting with distant metastases have a much higher incidence of multifocal lesions and a high mortality rate. Subtotal thyroidectomy is sufficiently therapeutic strategy for incidental PTMC. [8] Another retrospective review in patients with multifocal papillary thyroid carcinoma (PTC) showed that patients in the multifocal PTC group were older and had a smaller mean tumor size, a more advanced tumor staging. They also

revealed higher percentage of non-remission status in comparison with those having solitary PTC[9]. As our patient initially diagnosed as multifocal PTC (right and left side), a more aggressive tumor pattern is concerned. Therefore, repeat cytology and biopsy were arranged to differentiate diagnosis from other neck pathology. It is difficult to diagnosis via neck ultrasonography and cytology. Therefore, biopsy is important if equivocal cytology result.

Pheochromocytomas (Pheo) and paragangliomas (PGL) are rare tumors with heterogeneous genetic background. Malignancy is rare but it frequently associates with SDHB mutations. Currently, we know that thoracic paraganglioma might related with germline of SDHx, SDHAF2, MAX and TMEM127 mutations.[10,11] In 2013, a case was diagnosed as papillary thyroid carcinoma associated with paraganglioma and Dandy-Walker Malformation was reported whose SDHD mutation positive. The authors have noted that SDHx mutations can cause familial paraganglioma syndromes. These combinations currently supposed to be an atypical MEN syndrome or a subtype of MEN syndrome.[10,11] Unfortunately, our patient refused genetic survey.

In conclusion, carotid body tumors are rare tumors but highly related with neck paraganglioma, which are mostly benign tumor. A neck mass should be evaluated thoroughly and carefully, either by neck sonography, images, fine needle aspiration cytology or even tissue biopsy, especially for recurrent tumors or previous history of papillary thyroid carcinoma.

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化學受器瘤與其共存乳頭狀甲狀腺癌之病例報告

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摘 要

區分頸動脈體副神經節瘤和疑似復發性乳頭狀甲狀腺癌的頸部腫瘤有時很困難。方法：我們報告了一名66歲的女性，既往有甲狀腺乳頭狀癌病史，兩年後在右下頜下區發現了一個3厘米大小的一個硬質且快速生長的腫塊。最初經細胞學檢查懷疑復發性甲狀腺癌。結果：反覆細胞學檢查疑似轉移性癌病變。但後來，組織切片與頸部磁共振成像(MRI)均顯示了頸動脈分叉處的副神經節瘤。結論：頸動脈體瘤與頸部副神經節瘤高度相關，但常因淋巴結或甲狀腺病變而誤導抑或忽略此診斷的可能性。生成原因可能與胚系突變有關。鑑別診斷的重要性不僅對治療而且對後續監測都有影響。手術切除是目前第一線的治療方法，但若此腫瘤解剖位置或大小因素考量亦可考慮放射線治療。頸動脈體瘤有一被遺忘的專有名詞稱為化學受器瘤(chemodectoma)，以此文介紹給大家。