Auris Nasus Larynx 42 (2015) 59–62

Contents lists available at ScienceDirect

Auris Nasus Larynx

journal homepage: www.elsevier.com/locate/anl

A case of carotid body tumor concomitant with carcinoid tumor☆

Mi Jin Mun [a], Jin Choon Lee [b], Byung Joo Lee [b,*]

[a] Department of Otorhinolaryngology – Head and Neck Surgery, Busan St. Marry's Medical Center, Busan, Republic of Korea
[b] Department of Otorhinolaryngology – Head and Neck Surgery, Pusan National University School of Medicine and Biomedical Research Institute, Pusan National University, Busan, Republic of Korea

ARTICLE INFO

Article history:
Received 5 June 2014
Accepted 18 August 2014
Available online 6 September 2014

Keywords:
Paraganglioma
Carcinoid tumor
Carotid body

ABSTRACT

Neuroendocrine tumors typically fall into two broad categories: those of epithelial origin and those of neural derivation. The former group includes carcinoid tumors and the latter includes paraganglioma. Although paraganglioma and carcinoid tumor have different biologic behaviors, their overlapping histological appearance can pose diagnostic challenges. Carcinoid tumors are rare, slow-growing neuroendocrine tumors arising from the enterochromaffin cells disseminated throughout the gastrointestinal and bronchopulmonary systems. Carotid body tumor is the most common type of extra-adrenal paraganglioma. Paraganglioma tends to grow slowly but can compress adjacent vessel and nerve. A 63-year-old woman showed huge mass extending from carotid body to skull base, encircling internal and external carotid arteries on magnetic resonance image. Surgical removal of carotid body tumor was done after embolization procedure. Postoperative histopathologic examination and immunohistochemical analysis were consistent with paraganglioma concomitant with carcinoid tumor in carotid body. Primary cervical carcinoid tumor is extremely rare, and to the best of our knowledge, this is the first case of concomitant existence of paraganglioma and carcinoid tumor in carotid body.

1. Introduction

Neuroendocrine tumors are divided into two categories according to their origin: those of epithelial origin and those of neural derivation. The former group includes carcinoid tumor and the latter includes paraganglioma. Even though paraganglioma and carcinoid tumor have different biological behavior, differential diagnosis can be hard due to their overlapping histological appearance [1].

The most common type of extra-adrenal paraganglioma is carotid body tumor [2]. Carotid body tumor is derived from paraganglionic cell of carotid body that is located at carotid bifurcation. The growth rate of tumor is very slow but tumor itself can invade adjacent tissues or cause compressive effect on vessels and nerves [3]. Incidence of carotid body tumor is high in those who live in high altitude and who are affected by chronic obstructive pulmonary disease. It is therefore concluded that chronic hypoxia might be the cause of carotid body tumor [2]. The diagnosis and treatment of carotid body tumor remain difficult due to low incidence [4]. Most of carotid body tumor is benign and malignant form is rare. Malignant transformation rate is known as 6–30%, average 10% [2].

The other form of neuroendocrine tumor is carcinoid tumor. Carcinoid tumor produces serotonin which cause carcinoid syndrome like diarrhea, hot flush, and bronchospasm. The majority of carcinoids are found within the gastrointestinal tract (55%) and bronchopulmonary system (30%). Far less common anatomic sites have been reported in the literature, including carcinoids of the breast, larynx, thymus, and gallbladder [5]. Carcinoid tumor in head and neck field is rare.

Although carcinoid tumor and paraganglioma have overlapping histological features, there has been no report about concomitant existence of both carcinoid tumor and paraganglioma in one tumor so far. This is the first case of concomitant existence of paraganglioma and carcinoid tumor in carotid body.

2. Case report

A 63-year-old female presented with right side neck mass for 4 years. On the laryngoscopic examination, her right side vocal cord was paralyzed and there was a huge mass bulging from the right side of oropharynx. Magnetic resonance image (MRI) results

☆ This work was supported for two years by Pusan National University Research Grant.

* Corresponding author at: Department of Otorhinolaryngology – Head and Neck Surgery, Pusan National University School of Medicine and Medical Research Institute, 1-10 Ami-Dong, Seo-Gu, Busan 602-739, Republic of Korea.
Tel.: +82 10 8550 1300; fax: +82 51 246 8668.
E-mail address: voiceleebj@gmail.com (B.J. Lee).

http://dx.doi.org/10.1016/j.anl.2014.08.015
0385-8146/

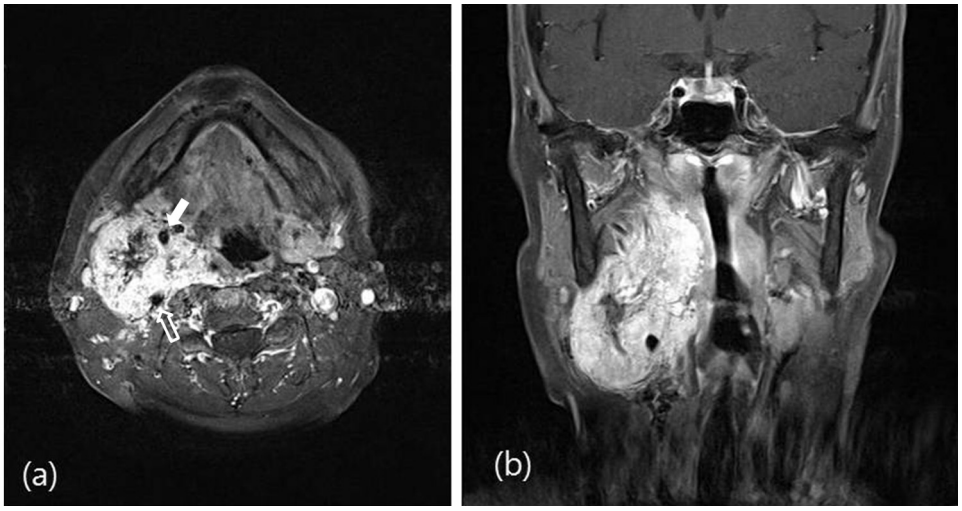

Fig. 1. Preoperative MR images. (a) Coronal image of carotid body tumor. Mass is placed between external (white arrow) and internal carotid artery (empty arrow). (b) Sagittal section of main mass. The mass extends from carotid body to skull base causing compressive effect to adjacent structures.

showed 5.1 cm × 3.1 cm sized round shape mass extending from carotid body to skull base, encircling internal and external carotid arteries. The mass showed increased signal intensity on T2 weighted image extending from parapharyngeal space to skull base (Fig. 1). Patient got embolization procedure using polyvinyl alcohol (PVA) particles to reduce peri-operative bleeding one day before surgery.

Under general anesthesia, right carotid body tumor removal was done. The tumor was located on carotid bifurcation in carotid sheath. There were severe adhesion between carotid artery and the tumor, so we extracted external half of the tumor and external carotid artery ligation was inevitable. During operation, frozen biopsy resulted with paraganglioma. We thought that complete surgical removal of the tumor was possible before surgery. But the tumor extended to the skull base and there were severe adhesion between adjacent structures and the tumor, so complete resection was impossible. After operation, she had no specific complication including Horner syndrome. Permanent tissue analysis of the specimen revealed 4.5 cm × 3.0 cm × 2.6 cm in size, with clear margin and including solid mass. In low power view, the tumor was composed of two different tumors (Fig. 2a). The tumor cells of paraganglioma have a "zellballen" growth pattern. This refers to a well developed nested or organoid growth pattern of the tumor cells with an intervening stromal component of delicated fibrovascular tissue and supporting cells at the periphery of the zellballen or cell nests. The nests are containing round or vacuolated cells with moderate amounts of cytoplasm and vesicular nuclei with clumped chromatin (Fig. 2b). The carcinoid tumor, which is growing in insular, ribbon, cords or pseudoglandular pattern, is containing uniform polygonal cells showing scant cytoplasm, dark nuclei with fine granular chromatin, and inconspicuous nucleoli (Fig. 2c). Both sides of the tumor showed strong positive reaction to immunohistochemical stain for synaptophysin, chromogranin, and CD56 which were neuroendocrine tumor markers. Immunohistochemical stain for S-100 protein demonstrated delicate networks of sustentacular cells in paraganglioma. S-100 protein was negative in carcinoid tumor (Fig. 3). Final diagnosis was reported as concomitant existence of paraganglioma and carcinoid tumor.

She got additional radiation therapy due to residual tumor after operation and no signs of recurrence have been detected in 2-year follow-up period.

3. Discussion

Carcinoid tumor and paraganglioma have different biologic behaviors, but they share similar histological appearance. Carcinoid tumor originates from epithelial components of neuroendocrine cells, whereas paraganglioma is derived from neural components [1].

Carcinoids are rare neuroendocrine tumors arising from the enterochromaffin (Kulchitsky) cells. The diagnosis of carcinoid tumor is based on histology with confirmation by positive

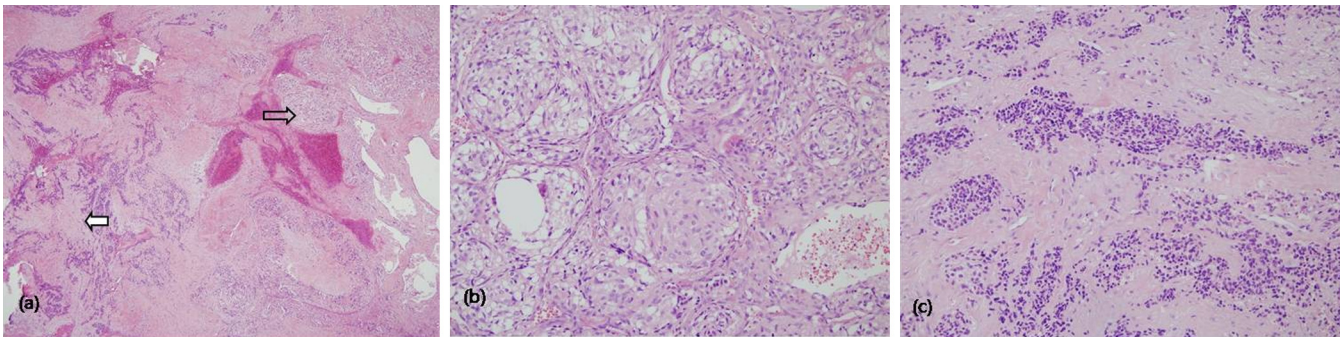

Fig. 2. (a) In low power view, the tumor is composed of two different tumors which are showing zellballen pattern (white arrow) and organoid pattern (empty arrow) (H&E, 40×). (b) Tumor cells of paraganglioma are arranged in small nests around vessels. The nests are containing round or vacuolated cells with moderate amounts of cytoplasm and vesicular nuclei with clumped chromatin. (c) The carcinoid tumor which is growing insular, ribbon, cords or pseudoglandular pattern, is containing uniform polygonal cells showing scant cytoplasm, dark nuclei with fine granular chromatin, and inconspicuous nucleoli (H&E, 200×).