Case Report

Nasopharyngeal adenoid cystic carcinoma: a case report and review of the literature

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Received May 18, 2014; Accepted June 12, 2014; Epub June 15, 2014; Published July 1, 2014

Abstract: ACC derived from nasopharyngeal epithelial cells is rare, usually benign. In this article, we reported a nasopharyngeal adenoid cystic carcinoma (NACC) in a 31-year-old woman with a symptom of hoarseness, headache, epistaxis slightly, diplopia, facial numbness and dysphagia near 3 months. A tumor on the right side of the nasopharynx was confirmed by laryngoscope check and MRI of the skull base. Histopathological findings showed that tumor cells were arranged in cord-like or acinar-like by atypical hyperplastic epithelial cells forming a cribriform and tubular pattern, and immunohistochemical findings showed that tumor tissues were immunopositive for p63 (+), CK7 (+), CK19 (+), CK8 (+), CK18 (+), SMA (+), CK (+), p53 (++), S-100 (+) and Ki-67 (5%+), and negative for CD34 (-), CK5/6 (-), CEA (-) and CD117 (-). Patient was treated by surgical operation and radiotherapy, and was followed-up near 10 months, no local recurrence and distant metastasis.

Keywords: Adenoid cystic carcinoma, nasopharynx, immunohistochemistry

Introduction

Adenoid cystic carcinoma (ACC) was first described by the Billroth in 1895. Typical features of ACC shows a slow growth rate, high propensity for perineural spread, local recurrence and distant metastasis (like lung, liver, bone, et al) [1, 2]. Now, ACC is one of the most common malignant salivary gland neoplasms, but occurs rarely in the nasopharynx, accounting for 0.13~0.48% of all malignant nasopharyngeal tumors [3, 4]. ACC of the nasopharynx (NACC) has a characteristic with low lymphatic metastases rate, slow growth rate and high local invasiveness. And, partly NACC patients show perineural spread, intracranial involvement and cervical lymphadenopathy [5, 6]. So, the treatment of NACC should be combined by surgical operation and radiotherapy. Here, we reported a NACC in a 31-year-old woman with a symptom of hoarseness, headache, epistaxis slightly, diplopia, facial numbness and dysphagia near 3 months.

Case report

A 31-year-old woman had a symptom of hoarseness, headache, epistaxis slightly, diplopia, facial numbness and dysphagia both for liquid or solid food randomly last for 3 months. A nasopharynx tumor was observed by laryngoscope check. MRI of the skull base, signal uneven, high signal of T2W1, also confirmed an irregular mass in the right side of the nasopharynx with infiltration of right pterygopalatine fossa and cavernous. She was hospitalized in Department of Otolaryngology at Taiping People’s Hospital of Dongguan for endoscopic surgery. And, two irregularly gray mass measured 1 cm × 0.5 cm × 0.4 cm in volume from nasopharynx were taken out in succession. Wound healing was good after surgery one week, and radiation was used begin in another one week. Histopathological findings showed that tumor cells were arranged in cord-like or acinar-like by atypical hyperplastic epithelial cells forming a cribriform and tubular pattern,
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and some glass-like eosinophilic materials or mucilages were observed in tumor interstitials (Figure 1). Immunohistochemical findings showed that tumor tissues were immunopositive for p63 (+), CK7 (+), CK19 (+), CK8 (+), CK18 (+), SMA (+), CK (+), p53 (++), S-100 (+) and Ki-67 (5%+), and negative for CD34 (-), CK5/6 (-), CEA (-) and CD117 (-) (Figure 2). The patient was followed-up near 10 months, no local recurrence and distant metastasis.

Discussion
ACC can arise from parotid, submandibular, palate, intraoral site, buccal, tongue and nasopharynx glands [1-9]. Although most ACC arising from the head and neck glands, ACCs are

Figure 1. The NACC tissues were stained with hematoxylin and eosin. A: Tumor cells were arranged in cord-like or acinar-like by atypical hyperplastic epithelial cells forming a cribriform and tubular pattern (40 ×). B: Some glass-like eosinophilic materials or mucilages were observed in tumor interstitials (100 ×).

Figure 2. NACC tissues were stained with immunohistochemistry. The tissues were immunopositive for CK (A), CK7 (B), CK8 (C), CK18 (D), CK19 (E), Ki-67 (F), P53 (G), P63 (H), S-100 (I), SMA (J) and immunonegative for CD34 (K), CD117 (L), CEA (M), CK5/6 (N) in NACC tissues (100 ×).
common in uterine cervix, lungs, kidneys, prostate, skin, esophagus and breast [2, 4, 10, 11]. When ACC patients present high stage disease, skull base involvement, tumor recurrence, lymphovascular invasion, bone invasion and perineural invasion, suggest an increased incidence of recurrence or dying in patients [2-4].

The best treatment for ACC is unanimously considered endoscopic surgery followed by radiotherapy [2, 4, 12]. MRI of the skull base is necessary for NACC patients, although there is no consensus on the imaging characteristics of NACC [2, 6, 13]. This approach ensures an optimal therapeutic outcome via accurate tumor mapping with special attention to the perineural spread. It is important to distinguish NACC from nasopharyngeal squamous cell carcinoma (NSCC). The skull base MRI of NSCC patients have a uniform signal on lesions. However, most NACC patients have a non-uniform signal on lesions.

Besides MRI, histological findings and immunohistochemical findings are very important in the diagnosis and distinguish of NACC. The cribriform and tubular pattern on tumor tissue and immunopositive for p63, CK7, CK19, CK8, CK18, SMA, CK, p53, S-100 and Ki-67 (5%), and negative for CD34, CK5/6, CEA and CD117, lead us to diagnose this patient as having a NACC. Cranial nerves invasion, stage and treatment approach might be important factors affecting the prognosis of NACC patients. Here, patient with no visible nerves invasion was treated by surgical operation following by radiotherapy, and followed-up showed no local recurrence and distant metastasis near 10 months after leaving hospital.

Acknowledgements

This work was supported by the National Natural Science Foundation of China (81273237), the Key Project of Science and Technology Innovation of Education Department of Guangdong Province (2012KJCX0059), the Science and Technology Project of Dongguan (20131051010006) and the Science and Technology Innovation Fund of Guangdong Medical College (STIF201110).

Disclosure of conflict of interest

None.

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