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Case Report

Myoepithelial carcinoma of pharynx expressing KIT and PDGFRA

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Abstract: KIT and PDGFRA expression has rarely been examined in myoepithelial carcinoma (MC) of the salivary glands. An 89-year-old Japanese woman presented with a pharyngeal mass. Gross and imaging examinations revealed an elevated mass in the middle pharynx next to the oral cavity. A biopsy revealed atypical cells, and tumorectomy was performed. The tumor was composed of atypical epithelioid cells arranged in solid nests, cords, and vague acinar patterns. Mitotic figures were recognized in 3 per 50 high power fields. Immunohistochemically, the tumor cells were positive for myoepithelial markers including cytokeratin (CK) 14, α-smooth muscle antigen, S100 protein, and p63. They were also positive for KIT, PDGFRA, pancytokeratin AE1/3, CK34βE12, CK5/6, vimentin, p53, and Ki-67 (labeling=28%). They were negative for neuron-specific enolase, CD45, CD34, CD56, chromogranin, synaptophysin, melanosome, desmin, epithelial membrane antigen, CK18, CK20, pancytokeratin (CAM5.2). A pathologic diagnosis of myoepithelial carcinoma arising from minor salivary glands was made. No metastatic lesions were found by various imaging techniques. The patient is now receiving palliative radiation therapy 2 months after the operation. The present case showed that MC can express KIT and PDGFRA.

Keywords: Myoepithelial carcinoma, pharynx, KIT, PDGFRA

Introduction

KIT and *PDGFRA* genes, both mapped to 4q12, encode receptor tyrosine kinase oncoproteins called KIT (CD117) and platelet-derived growth factor receptor-α (PDGFRA), respectively [1-3]. Both molecules are transmembranous oncoproteins involved in tumorigenesis of some neoplasms including gastrointestinal stromal tumor, acute myeloid leukemia, mast cell neoplasms, germ cell tumors, melanoma, neuroendocrine carcinomas, large cell neuroendocrine lung carcinoma, small cell lung carcinoma, and adenoid cystic carcinoma [1-3].

Salivary gland tumor shows diverse morphologies. Among them, myoepithelial carcinoma (MC) is rare. MC is defined as malignant salivary gland tumor in which the tumor cells almost exclusively manifest myoepithelial differentiation [4]. It is well known that adenoid cystic carcinoma expresses KIT in the salivary glands carcinomas. However, there are only several reports of KIT and PDGFRA expressions in salivary gland carcinomas other than adenoid cystic carcinoma, to the author's knowledge [5-11]. Here reported is a case of pharyngeal MC expressing KIT and PDGFRA.

Case report

An 89-year-old Japanese woman complained of a right pharyngeal mass, and consulted to Otolaryngology section of our hospital. Gross and imaging examinations revealed an elevated mass in the right middle pharynx adjacent to the oral cavity. A biopsy revealed atypical cells suggestive of atypical carcinoid. A tumorectomy with wide margins was performed. Post-biopsy imaging modalities identified no metastatic lesions. Grossly, the tumor measured 1.2 x 1.3 x 1.3 cm, and whitish hard. Microscopically, the tumor was composed of atypical epithelioid cells arranged in solid nests, cords, ribbon-like, and vague acinar patterns (**Figures 1A** and **1B**). Epithelial-myoepithelial pattern was not seen. Adenoid cystic pattern was not noted. No lymphocytic infiltration was recognized. Mitotic fig-

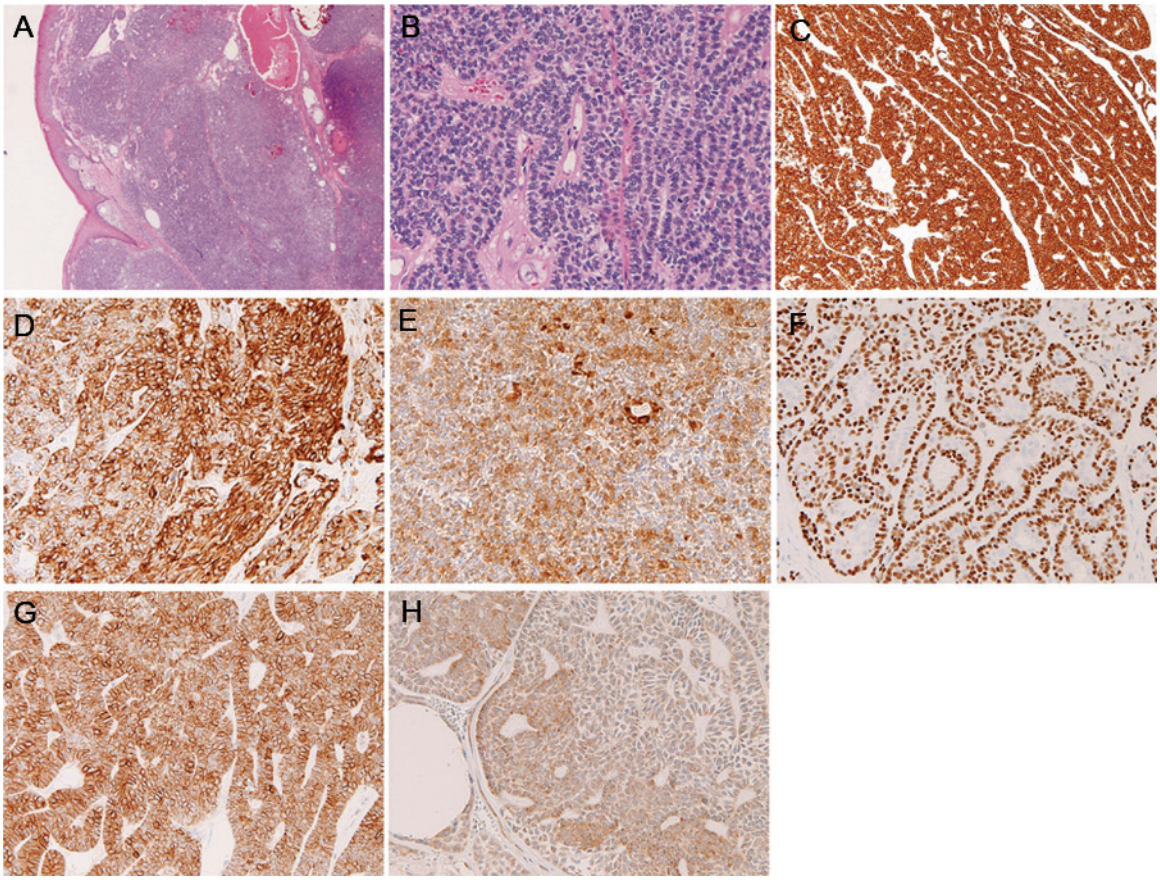

Figure 1. A. Low power view of the tumor. The tumor is solid and medullary, and focally invasive into the squamous cells. Acinar formations are recognized in some areas. HE, x20. B. High power view of the tumor. The tumor cells are arranged in cord pattern. They have hyperchromatic nuclei, and relatively clear cytoplasm. HE, x200. C. Cytokeratin 14 is diffusely positive. Immunostaining, x200. D. α-smooth muscle actin is strongly positive. Immunostaining, x200. E. S-100 protein is positive. Immunostaining, x200. F. p63 is positive in the nuclei. Immunostaining, x200. G. KIT is strongly positive in the membrane. Immunostaining, x200. H. PDGFRA is positive in the membrane. Immunostaining, x200.

ures were recognized in 3 per 50 high power fields. The tumor cell was infiltrative into mucosa and surrounding tissue (**Figure 1A**).

An immunohistochemical study was performed with the use of Dako's envision method, as previously described [12-15]. Immunohistochemically, the tumor cells were positive for myoepithelial markers including cytokeratin (CK) 14 (**Figure 1C**), α-smooth muscle antigen (**Figure 1D**), S100 protein (**Figure 1E**), and p63 (**Figure 1F**). They were also positive for KIT (**Figure 1G**), PDGFRA (**Figure 1H**), pancytokeratin AE1/3, CK34βE12, CK5/6, vimentin, p53, and Ki-67 (labeling=28%). They were negative for neuron-specific enolase, CD45, CD34, CD56, chromogranin, synaptophysin, melanosome, desmin, epithelial membrane antigen, CK18, CK20, pancytokeratin (CAM5.2).

A pathologic diagnosis of myoepithelial carcinoma arising from minor salivary glands was made. No metastatic lesions were found in various imaging techniques. The patient is now receiving palliative radiation therapy 2 months after the operation.

Discussion

The present tumor arose in the pharynx. The present tumor appears to be derived from minor salivary glands of the pharynx. Salivary glands tumors show diverse morphologies. Normal salivary glands contain myoepithelial cells, which can give rise to MC. MC is a rare tumor among salivary malignancies [4].

The present tumor showed epithelioid morphologies whose cell arrangement was reminiscent

of atypical carcinoid. However, it was found that the tumor cells showed immunoreactive myoepithelial markers, i.e. S100 protein, p63, CK14, and α-smooth muscle actin. These four antigens were well known markers of myoepithelial cells [4]. Therefore, the present case showed predominant myoepithelial differentiation. The cellular atypia, infiltrative growth, high Ki-67 labeling, and positive p53 expression are indicative of malignant potential of this tumor. Therefore, the present case was though to be MC. Epithelial-myoepithelial carcinoma, adenoid cystic carcinoma, and basal cell carcinoma are unlikely.

Several studies of KIT expression of salivary gland tumors have been published [5-9]; KIT expression have been observed in adenoid cystic carcinoma, polymorphous low-grade adenocarcinoma, acinic cell tumors, epithelial-myoepithelial carcinoma, basal cell carcinoma, lymphoepithelial carcinoma, pleomorphic adenoma, and monomorphic adenoma [5-9]. However, KIT expression in MC has been reported only once by Jeng et al [5] who demonstrated KIT expression in two cases of MC. They also reported no mutations of *KIT* gene in various types of salivary gland tumor [5]. The findings of the KIT of the present case are consistent with those of Jeng et al [5]. More studies of KIT protein and *KIT* gene are required in MC.

To the author's knowledge, there are only two studies of PDGFRA in salivary glands tumors [10, 11]. PDGFRA expression was observed in adenoid cystic carcinoma and carcinoma ex pleomorphic adenoma [10, 11]. There have been no reports of PDGFRA in MC. The present study showed that MC can express PDGFRA. Much more studies of PDGFRA in salivary gland tumors are required.

In summary, the author reported a case of MC of the pharynx expressing KIT and PDGFRA.

Declaration of conflict of interest

The author has no conflict of interest.

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