Kurzmitteilung

Polymorphous Low-Grade Adenocarcinoma of the Nasopharynx
Case Report and Review of the Literature

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Background: The polymorphous low-grade adenocarcinoma of the nasopharynx is a rare disease. Polymorphous low-grade adenocarcinoma is a minor salivary gland neoplasm which occurs frequently in the mucosa of the soft and hard palates, in the buccal mucosa and in the upper lip. To date this entity has been identified within the oral cavity and only one case within the nasopharynx and some cases in the parotid gland. It has a slow infiltrating growing pattern with frequent perineural invasion and low metastatic potential.

Case Report: We report on a patient with non-papillary polymorphous low-grade adenocarcinoma in the nasopharynx which extended intracranially. The patient underwent primary radiotherapy. The CT showed partial response to radiotherapy and the patient is alive 51 months after the diagnosis his state being unchanged.

Conclusion: The treatment for minor salivary gland tumor is primarily surgical. It is reported that the polymorphous low-grade adenocarcinoma has been known to have poor response to radiotherapy. However, we believe that in addition to its favorable biological behavior, the radiotherapy in this localization may result in longer survival.

Key Words: Nasopharyngeal neoplasm · Polymorphous low-grade adenocarcinoma · Radiotherapy

Polymorphes Adenokarzinom des Nasopharynx mit niedrigem Malignitätsgrad. Fallbeschreibung und Literaturübersicht


Schlussfolgerung: Die Therapie des Speicheldrüsenkarzinoms ist in erster Linie eine chirurgische. Es ist bekannt, dass das polymorphe Drüsenkarzinom mit niedrigem Malignitätsgrad auf Strahlenbehandlung nur wenig reagiert. Wir sind aber der Überzeugung, dass die Strahlenbehandlung zur Verlängerung des Überlebens beiträgt.

Schlüsselwörter: Nasenrachenraumkarzinom · Polymorphes niedrigmalignes Adenokarzinom · Strahlenbehandlung

Polymorphous low-grade adenocarcinoma of the nasopharynx is an extremely rare malignancy. It is a malignant epithelial neoplasm which occurs almost exclusively in minor salivary gland site and rarely in the major salivary gland [8, 9], nasopharynx and nose [16, 18]. In spite of the infiltrative grown pattern, this tumor has a low local recurrence rate and a remarkably low potential for distant metastasis [10, 14].

Primary treatment of the intra-oral polymorphous low-grade adenocarcinoma is surgical. Radiotherapy and adjuvant chemotherapy have been used, although there is no evidence for their benefit in case of tumors of intra-oral minor salivary gland origin [3, 4]. We present a patient with a non-papillary variant of polymorphous low-grade adenocarcinoma of the nasopharynx that responded to radiotherapy.
Case Report

Medical History: Forty-four-year-old man visited the hospital complaining of headache, left-sided hearing loss and nasal obstruction. The head and neck examination was unremarkable except for left secretory otitis media. Fiberoptic examination showed a reddish-grey mass filling the whole epipharynx. Biopsy was performed and histologic analysis revealed a polymorphous low-grade adenocarcinoma. Nasopharyngeal tumor and its invasion of the skull base and intracranial spread was diagnosed by CT (Figure 1). Cervical lymph node metastasis and distant metastasis were not evident in the patient at the time of diagnosis.

Pathological Examination: Macroscopically a 10 ¥ 8 ¥ 6 mm, firm, unencapsulated, light tan to gray coloured tumor mass was attached to the epipharyngeal mucosa.

Microscopically within solid nests small duct-like structures, tubules and trabeculae were seen. Pseudocystic spaces contained eosinophilic homogenous material. The cells forming the tubular structures in a single layer and the solid areas were uniform, cuboidal or low columnar having scant eosinophil cytoplasm with indistinct borders, isoform round or oval nuclei with inconspicuous or absent nucleoli. The nuclear membrane was thin, slightly irregular, the chromatin finely dispersed or granular. There was one mitosis per 10 high-power fields. The relatively limited stroma showed hyalinisation.

Treatment: Total surgical removal was not executed because the tumor infiltrated the base of the skull. The first part of radiation treatment included 2 lateral opposed fields for the nasopharynx and upper cervical lymph nodes (dose prescribed to the midline) with the dose of 60 Gy. The target volume (sizes of portals 8 ¥ 10 cm) was irradiated by 60Co, with a daily fraction of 2 Gy over 5 days per week. The primary lesion persisted after radiation, the tumor extension into the intracranium required boost therapy with cobalt radiation with reduced bilateral fields centralizing to the sella (sizes of portals 6 ¥ 6 cm). Then another 20 Gy were delivered to the nasopharynx up to a total dose of 80 Gy. Chemotherapy was contraindicated by the cardiac status of the patient.

Follow-up CT scans were made at 3- to 6-month intervals. Twelve months after the complete treatment CT showed partial response to radiotherapy in both the nasopharynx and the intracranium (Figure 2). The subsequent CT scans showed unchanged status. The patient lives with this disease for 51 months. To date regional or distant metastasis has not been detected. MRI-examination was impossible because metal cardiac valve had been implanted in mitral position.

Discussion

Polymorphous low-grade adenocarcinoma has also been called low-grade papillary adenocarcinoma [1, 12], terminal duct carcinoma [5] and lobular carcinoma [4]. The papillary low-grade adenocarcinoma of minor salivary gland origin is considered to be a variant of the polymorphous low-grade adenocarcinoma. Recent distinction between polymorphous low-grade adenocarcinoma and low-grade papillary adenocarcinoma on clinical and histological grounds emphasizes the more aggressive behavior of the papillary type [13, 15]. Distinguishing polymorphous low-grade adenocarcinoma from other tumors such as intra-oral salivary gland carcinoma arising within a pre-existing pleomorphic adenoma and adenoid cystic carcinoma is important because these tumors have much worse prognosis and they are more likely to recur and metastasize to regional and distant sites and they lead to the death of the patients [10, 13].

Polymorphous low-grade adenocarcinoma frequently occurs in the 5th and 6th decades of life and there is a 2 : 1 female predominance. There may be a predilection for this tumor in blacks [10]. The palate is the most common localization, and histologically polymorphous low-grade adenocarcinoma is the second most common type in this localization following adenoid cystic carcinoma. Waldron et al. [17] published 426 minor salivary gland tumors and polymorphous low-grade adenocarcinomas represented 11% of all of them and 26% of all of the tumors were malignant [17]. Over 60% of the polymorphous low-grade adenocarcinomas arise from the mucosa of either the soft or the hard palate, about 12% occurred in the buccal mucosa and 12% in the upper lip [3]. The remaining tumors were found in the retromolar region, floor of the mouth, tongue and some cases were seen in the major salivary glands [8, 9, 11] and the nasopharynx [7, 16, 18]. In contrast polymor-