Papillary cystadenocarcinoma: Report of a case of high-grade histopathologic malignancy

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Abstract

Papillary cystadenocarcinoma is an extremely rare malignant tumor of the salivary gland which is cytologically considered to be of low-grade malignancy, showing an indolent biological behavior. Histologically, the tumor is characterized by cysts and papillary endocystic projections. This tumor most commonly arises in the major salivary glands, mainly the parotid gland, but involvement of the minor salivary glands has also been reported [2,4,5]. Most patients present a mass of slow and painless growth [1].

We report here a rare case of PC arising in the palate of a 79-year-old man, which was histologically considered to be of high-grade malignancy. We found no case of high-grade histologic PC located in the palate in the literature. We describe the clinical and histologic features of the tumor, as well as the immunohistochemical findings obtained using prostate-specific antigen (PSA), thyroglobulin, p53 and Ki 67 as markers.

2. Case report

Patient M.R.N., a 79-year-old black man, was seen at a surgery and oromaxillofacial traumatology service complaining of an asymptomatic volume increase in the palate for 6 months. The patient reported swallowing difficulties.
Review of the medical record revealed no noteworthy systemic alterations. Extraoral physical examination showed discrete facial asymmetry in the left hemiface. No palpable regional lymph nodes were noted.

Intraorally, an extensive exophytic tumor lesion measuring approximately 5.0 cm was identified in the hard palate, extending from the alveolar margin, crossing the midline and provoking expansion of cortical bones. In addition, the tumor presented a lobular aspect and an area of central ulceration (Fig. 1). Axial computed tomography scans with contrast injection showed a tumor mass measuring about 5.0 cm in diameter in the anterior region of the left maxilla and destruction of the buccal cortical bone, with the mass invading the maxillary sinus and extending in the direction of the hard palate (Fig. 2). In coronal sections with contrast injection the same tumor mass was found to be destroying the alveolar margin, body of maxilla and hard palate on the left side. Invasion of the inferior portion of the maxillary sinus and nasal fossa was also noted.

With the diagnostic hypothesis of squamous cell carcinoma, an incisional biopsy was performed whose anatomopathologic analysis revealed malignant salivary gland neoplasm. The tumor exhibited cystic cavities that contained papillary projections consisting of the proliferation of columnar or cubic cells with voluminous nuclei arranged in a single or double layer, as well as stratified areas (Fig. 3). The cells were characterized by atypias such as nuclear and cellular pleomorphism, numerous, sometimes aberrant, mitotic figures, and nuclear hyperchromatism, in addition to prominent nucleoli (Fig. 4). The stroma was scarce and exhibited a moderate, predominantly mononuclear inflammatory infiltrate. The histopathologic diagnosis was PC. The patient was referred to the Service of Head and Neck Surgery, but refused to undergo excision of
the tumor and any treatment. An immunohistochemical study was subsequently carried out by the streptavidin-biotin method using PSA and thyroglobulin as markers to rule out the hypotheses of metastases from prostate and thyroid tumors, respectively. The specimens were negative for these antibodies. Labeling for protein p53 showed strong positivity in the nucleus of neoplastic cells, while only few cells were positive for Ki 67.

3. Discussion

Cystadenocarcinomas of the salivary gland represent a distinct group of epithelial malignancies characterized by diverse cytomorphologic features and a predominantly cystic and invasive growth pattern. In many cases, the neoplastic cysts are associated with a papillary component [5]. Malignancy is confirmed by nuclear pleomorphism, an infiltrative growth pattern and mitoses [1].

In a review of 57 cases, PC showed no preference for gender or patient age, the latter ranging from 20 to 86 years, with a mean of 58.8 years [5]. The present patient was 79 years old, in agreement with the former study. In contrast, another review of 22 cases reported a strong predominance in males (17/22) and a mean age of 37 years (range: 17–61 years) [6].

Particularly interesting in the present case is the uncommon location of the tumor in the palate, considering that in a review of 57 cases PC was found in the major salivary glands in 65% of cases and in the minor salivary glands in 35%, with the lips being the most affected site, followed by the buccal mucosa, palate and tongue, with only four cases occurring in the palate [5].

Another intriguing finding was the size of the tumor, which measured 5 cm, and its time of progression which was only 6 months according to the patient. These facts disagree with literature findings indicating a mean size of 2.2 cm in the minor salivary glands (range, 0.4–6 cm) and a slow growth. In addition, we observed an area of ulceration, a characteristic not reported for the cases reviewed by other investigators [5]. Ulceration was reported in only one case but was associated with irritation due to the use of a partial prosthesis [2].

Histologically, the present case showed the features reported in the literature [4,7,8], differing in terms of the presence of intense nuclear and cellular pleomorphism, numerous mitoses, some of them atypical, nuclear hyperchromatism, and numerous prominent nucleoli. Some of these features have also been observed in a case of PC located in the tongue [9]. Furthermore, other investigators also described cases of high-grade histologic malignancy which were divided into well- and poorly differentiated tumors, with an association being observed between recurrence and nodal metastases and the poorly differentiated subtype [6]. Thus, the rapid progression and large extent of the present tumor might be related to the degree of histopathologic differentiation observed.

Variable proliferation of different cell types ranging from small cuboidal and large cuboidal cells to columnar cells was observed, with the present case thus belonging to the fourth type reported in the literature, which classifies PC into four types based on cellular morphology: the small cuboidal cell type which is the most common; the large cuboidal cell type, the columnar cell type, and a combination of these cell types [5]. In that study, the authors also observed a relationship between cases with a predominance of the columnar cell type and a higher metastatic potential. Among 57 cases analyzed, three of four metastasized tumors were of the columnar cell type [5].

The present tumor was negative for PSA and thyroglobulin, ruling out the hypotheses of metastases from prostate and thyroid tumors, respectively, and was therefore considered to be a primary carcinoma of the salivary gland. Strong staining for protein p53 was observed, in contrast to the findings of Pollett et al. [9] who reported a case of high-grade PC located in the tongue that was negative for this protein. Regarding Ki 67, only few cells were positive, indicating a low proliferative activity of the tumor, in contrast to the authors cited above who observed a proliferation rate of 60%. It would have been expected that a rapidly expanding tumour of 6 months duration would have had a high Ki-67 labelling index, which was not the case with this patient.

In conclusion, PC seems to present a broader histomorphologic spectrum than that generally recognized by the WHO, and at least some cases of this tumor should be considered to be high-grade malignant variants.

References


