CASE REPORT

Adenoid cystic carcinoma of buccal mucosa

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SUMMARY

Adenoid cystic carcinoma is a malignant neoplasm most commonly originating in the salivary glands of head and neck region. The clinical and pathological findings typical of this tumour include slow growth, perineural invasion and potential local recurrence. Up to 50% of these tumours occur in the intraoral minor salivary glands usually in the hard palate. We present a case report of a 26-year-old woman who was diagnosed with adenoid cystic carcinoma of the right buccal mucosa. The peculiarity of the lesion and the approach we made is the key factor in the presentation.

BACKGROUND

Adenoid cystic carcinoma (ACC) was first described by Theodor Bilroth in 1856, as cylindromas. The term 'ACC' was introduced by Ewing (Foote and Frazell) in 1954.¹⁻² ACC accounts for about 5–10% of all salivary gland neoplasms, representing 2–4% of malignant occurrences of the head and neck area.³ It presents a widespread age distribution, peak incidence occurs predominantly among women, between the fifth and sixth decades of life.⁴ The majority of the tumours arise in the major salivary glands, minor salivary glands of the oral cavity and mucous glands of the upper respiratory tract.⁵ According to the Armed Forces Institute of Pathology's (AFIP's) records, of their own diagnoses of ACC, 26.8% of cases occur in the parotid, 24% in the submandibular gland, 20.5% in the palate, 5% in the tongue, around 4% each in the lips and buccal mucosa and 1.2% in the sublingual gland.⁶ In the present case, ACC was located on the rare region of the oral cavity (buccal mucosa) of a 26-year-old patient with the histopathological features showing cribriform pattern with no perineural invasion.

CASE PRESENTATION

A 26-year-old woman presented to the PSM dental college for the evaluation of a mass on the right side of buccal mucosa. The patient reported that the lesion initially presented as a small swelling in the buccal mucosa region over a period of 1 year. Extraorally, there was facial asymmetry with swelling seen on the right side of the face on the cheek (figure 1). Head and neck examination revealed no evidence of paraesthesia or motor nerve deficiency. On palpation, it was hard and tender. An intraoral examination revealed an oval, red growth measuring about 3 cm in the maximum dimension which was tender on palpation. The surface over the skin was smooth. There was no clinical evidence of regional nodal involvement or metastatic disease.

Figure 1 Swelling on the right side of the face.

On the basis of clinical details, adenoma was given as provisional diagnosis.

INVESTIGATIONS

CT of the maxilla was suggestive of soft tissue density lesion seen in the right buccal mucosa anterior to the masseter muscle showing good enhancement with contrast, and ultrasound showed a 1.6×1.5 cm well-circumscribed hyperechoic lesion (figure 2).

Complete surgical removal of the lesion was performed (figure 3).

On gross examination, the specimen was oval in shape, brownish white in colour, measuring about 1.5×1 cm and 1.2×1.2 cm in diameter (figure 4). Histologically, the lesion was composed of isomorphic hyperchromatic basaloid tumour cells arranged in cribriform and tubular patterns (figures 5 and 6). The tumour cells were polygonal in shape.

Figure 2 CT scan of maxilla showed soft tissue lesion in right buccal mucosa.

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Figure 3 Complete surgical removal of the lesion.

The cribriform spaces contain eosinophilic and basophilic materials and there is an evidence of hyalinised stroma surrounding the cribriform islands (figure 7). Expression for cytokeratin 10 (CK10) was seen within the ductal cells of the tumour (figure 8). The histological features were suggestive of ACC.

DIFFERENTIAL DIAGNOSIS

Based on the patient's history and the clinical and radiographic examination, the differential diagnosis of mucoepidermoid carcinoma, adenoid cystic carcinoma or carcinoma of maxillary sinus was suggested.

TREATMENT

Complete surgical removal of the lesion was performed (figure 3).

OUTCOME AND FOLLOW-UP

After surgical excision of the tumour, the margins of the tumour were clear microscopically and a regular follow-up of the patient with repeated scans and radiographs has not revealed any signs of recurrence or metastasis until today.

Figure 4 Gross specimen.

Figure 5 Histopathological view (H&E $\times 4$).

DISCUSSION

The term cylindroma, a pathological entity later called 'ACC', was coined by Billroth.¹ Until 1940, the tumour was thought to be a benign variant of the mixed salivary gland tumour. In 1943, Dockerty and Mayo emphasised the malignant nature of this tumour.⁷ In 1953, Foote and Frazell renamed the lesion as ACC.²⁻⁸ Conley and Dingman described ACC as "One of the most biologically destructive and unpredictable tumours of the head and neck." WHO in 2005 defined ACC as "A basaloid tumour consisting of epithelial and myoepithelial cells in various morphological configurations, including tubular, cribriform and solid patterns." It has a relentless clinical course and usually a fatal outcome.⁹⁻¹⁰ ACC of the salivary gland is a slow-growing but aggressive cancer which is reflected by the good short-term but very poor long-term outcome of patients with this disease.¹¹ In advanced cases, fixation to skin or deeper tissues can occur due to its growth pattern, it should also be noted that at the time of presentation, these tumours have often invaded beyond the clinically apparent borders.¹¹⁻¹² Histologically, ACC can present three different variables: glandular (cribriform), tubular and solid. The mitotic figures are generally scarce in cribriform and tubular areas.¹³ The prognostic factors of ACC depend on tumour site, tumour stage, the

Figure 6 Histopathological view (H&E $\times 10$).

Figure 7 Histopathological view (H&E ×40).

presence of perineural invasion and tumour grade. Tubular and cribriform subtypes have better prognosis than solid subtypes.^{14–16} We performed a thorough microscopic evaluation of the specimen for marginal clearance.

CKs are a group of antigenically distinct intermediate filaments, and they consist of approximately 20 different keratin polypeptides.¹⁷ There is some restriction of the type of CKs expressed in different epithelia and their neoplasms.^{18–19} The immunohistochemical stains proved positive for pan-CK AE1/AE3 and epithelial membrane antigen among ACC.²⁰ The differential expression of CKs 7 and 20 by carcinomas can help discriminate between carcinomas of different primary sites and can be useful in the determination of the primary site of origin of metastatic carcinomas.²¹ CK10 is the salivary gland duct-specific marker.²² In our case CK10 was positive within the ductal cells of the tumour (figure 8), which is in accordance with the study performed by Missall *et al.*²²

The treatment aspect of the tumour is chiefly surgical, although in some cases surgery has been successfully coupled with radiotherapy. Lymphatic spread is far less common in this tumour. In our patient, we preferred surgical excision as the sole treatment because histopathologically the lesion was cribriform type which has better prognosis than the other variants.

Figure 8 Expression for cytokeratin 10.

Learning points

- ▶ The primary treatment objective in the patients with adenoid cystic carcinoma is distant metastasis prevention.
- ▶ Surgical therapy involving associated radiotherapy remains as the modality of choice for most cases.
- ▶ Regular follow-up of the patient with repeated scans and radiographs are advised to prevent recurrence or metastasis.

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Competing interests None.

Patient consent Obtained.

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