Case Report

Oral carcinoma cuniculatum

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Abstract

Oral Carcinoma cuniculatum(OCC) is rare variant of squamous cell carcinoma. The histopathological features of this type of carcinoma makes it special in the literature of Pathology. The appearance of deep invading epithelial islands with pool of keratin cores very similar to rabbit burrows gives the term cuniculatum to the carcinoma. Here we present a case of OCC on the lateral surface of the tongue. Early diagnosis of such case is very important to distinguish it distinctly from the other entities of squamous cell carcinoma like verrucous carcinoma. The knowledge of existence of such entity of squamous cell carcinoma occurring in the oral cavity is very important to facilitate correct treatment planning and management.

Keywords: keratin plugging, oral carcinoma cuniculatum, tongue squamous cell carcinoma

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INTRODUCTION

Oral carcinoma cuniculatum (OCC) is a rare, unacquainted variant of oral squamous cell carcinoma (OSCC).[1] It is similar to cutaneous CC (which was first described by Aird *et al.* in 1954) in its clinicopathological findings and biological behavior.[2,3]

OCC is confused clinically and histologically with verrucous carcinoma (VC) and is often misdiagnosed as either VC or OSCC. To the best of our knowledge, till date, only more than 50 cases of this tumor have been reported in the oral cavity (including the present case) and only a limited number of cases have been reported from the Indian subcontinent. OCC is characterized by the formation of keratin-filled crypts[4] extending down into the supporting tissues, including skeletal muscle and bone. Neutrophil microabscesses are frequently found within these channels. By contrast, VC features a broad "pushing" infiltrative front that lacks the complex interconnecting network of channels characteristic of OCC. In addition, mandibular invasion by OCC may result in a large radiolucent cavity, whereas VC abutting the jawbone tends to cause surface resorption without invasion. Furthermore, foci of conventional SCC may be found in up to 20% of VC.[5]

Thus, OCC is defined histologically by a characteristic infiltrative pattern of a deep, broad and complex proliferation of stratified squamous epithelium with keratin cores and keratin-filled crypts. Notably, there is a consistent absence of any significant cytological atypia[4] which, in the past, has led to failure in recognizing this neoplasm as malignant.[6]

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We present a case of a 32-year-old female patient who reported with a tongue ulcer.

CASE REPORT

A 32-year-old female came to the clinic with the complaint of nonhealing ulcer on the left lateral surface of the tongue which was mildly tender on palpation and left submandibular lymph nodes were tender.

She had a history of consumption of gutkha (chewable form of tobacco) but had quit the habit 5 years back. She complained of weight loss of 7 kg within the past 3 months along with loss of appetite.

Incisional biopsy of 0.7 cm × 0.5 cm was obtained from the lesion including adjacent tissue which was examined histopathologically [Figure 1].

Under low magnification, the superficial epithelium was thrown into papillary folds covered with thickened keratin.

The deeper sections revealed multiple branching crypts filled with keratin. There were numerous crypts with epithelial rete pegs pushing deep into the stroma with independent well-defined keratin pearls in the stroma [Figures 2 and 3].

Foci of nodules in the keratin pearls were seen. Crypts showed the characteristic burrowing type of invasion. The tumor cells in the crypts showed mild cytological atypia along with limited mitotic figures [Figure 4].

Looking at all the observations, a diagnosis of OCC was made.

DISCUSSION

OCC represents one of the rarest variants of OSCC. This

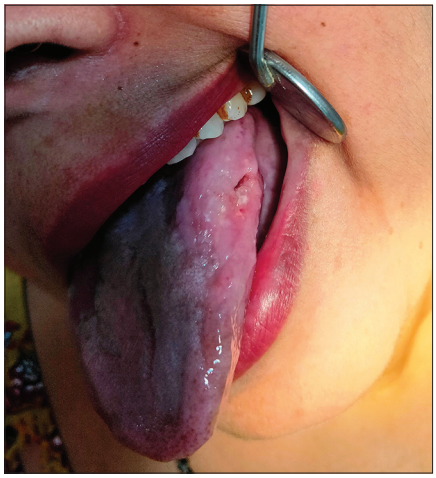

Figure 1: Deep ulcer on lateral border of tongue with no exophytic growth

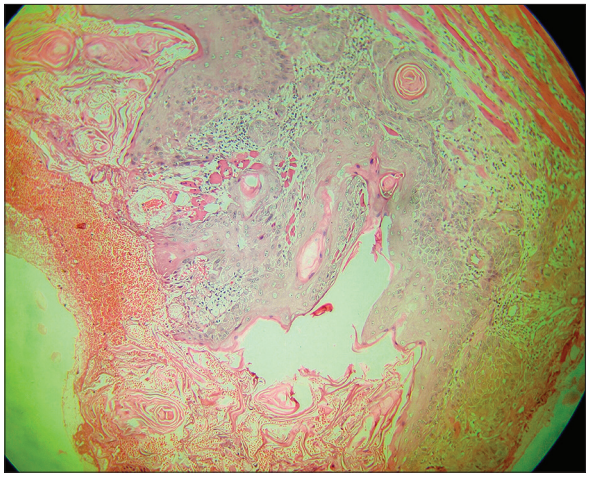

Figure 2: Excessive keratin flakes seen in superficial epithelium with keratin filled crypts (×10)

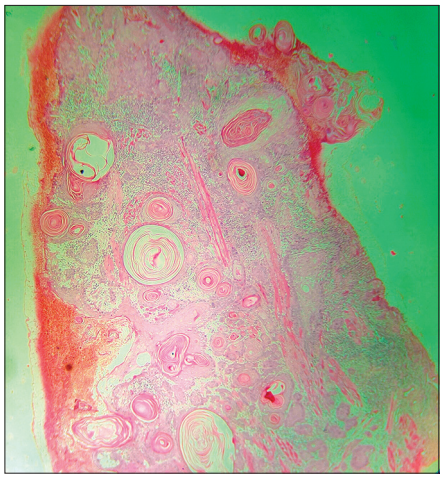

Figure 3: Low magnification shows networking or anastomosing cords of epithelial proliferation (×5)

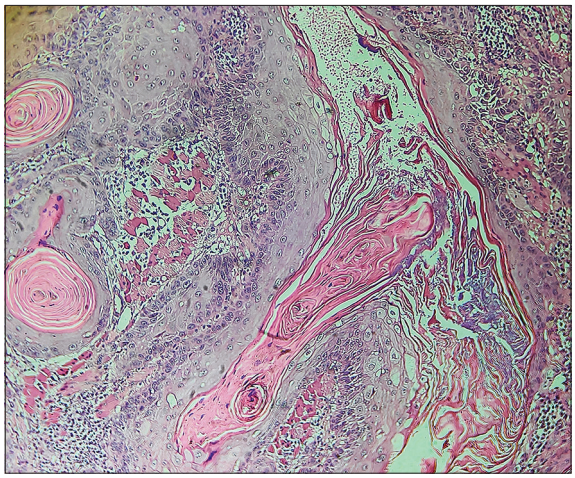

Figure 4: Keratin pearls in the connective tissue ,keratin plugging seen in the deeper portions of crypt. (×10)

term is least heard and discussed because of the rarity in reporting in medical literature. Fliegar and Owinski (1997) first reported a case of OCC after being described by Aird *et al.*[2] as a pathological entity of foot.

A systematic review has been reported by Farag *et al.* shows that the mandibular gingiva has been the most common site of occurrence, whereas the tongue was found the common site by Sun *et al.*[7] OCC is a rare entity and can be missed by pathologists.they are misdiagnosed as OSCC or VC.[5]

The clinical presentation was ulcer and induration similar to OSCC. The clinical course matches with that of oral VC which has a slow progression unlike OSCC which has faster progression.

The histopathological features are similar to that of VC, hence diagnosed as inverted VC in the older days. In addition, OCC is slightly aggressive than VC.[8]

Well-differentiated epithelial cells and epithelial islands lacking cellular atypia exhibiting blunt/pebbly surface with keratin filled crypts extending deep into stroma satisfied the diagnostic criteria.

The literature about this pathological entity is so rare and this had led to its misdiagnosis or under diagnosis. The etiological features are tobacco, alcohol, human papilloma virus (HPV), etc.

The presence of a mutated form of *P* 53 has been associated with malignant features but associated with OCC could not be much established or showed conflicting reports. Cutaneous form of CC shows correlation with HPV. OCC do not show such correlation, so pathogenesis with HPV as a factor remains unclear.[9,10] Clinically, it presents as an exophytic lesion.

The treatment of such cases is complete excision with 5 cm free margin. Chemotherapy and radiotherapy do not remain as main treatment modalities. It is locally aggressive and rarely shows metastasis.

The histopathological differential diagnosis includes OSCC (well differentiated) and VC. Epithelial clefting and parakeratin plugging are hallmark features of VC, whereas complex branching of keratin-filled crypts is diagnostic clue of OCC. Invasive epithelial components in the form of keratin channels are hallmarks for OCC. It burrows deep into muscles unlike VC. To date, there have been no reports of the DNA ploidy status of OCC prior to the current report. The diploid nature of this tumor suggests a more favorable clinical outcome.[11-17]

CONCLUSION

We report a case of OCC of the tongue and draw attention to its distinct clinicopathological features. OCC needs to be distinguished from other variants of OSCC since recognition of this entity and awareness of its clinical behavior lead to the appropriate management.

The management of such cases is always multidisciplinary with good communication between the oral pathologist and oral surgeon. The treatment of choice for OCC is complete surgical resection, which provides a highly favorable outcome.

It must be differentiated appropriately from other lesions.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient (s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initial s will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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