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Basal cell adenocarcinoma of submandibular gland mimicking a vascular mass – An unusual presentation of a rare tumor

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Abstract: Basal cell adenocarcinoma (BCAC), the malignant counterpart of basal cell adenoma, is a rare salivary gland neoplasm, the most common site being parotid gland. It has been classified as a low-grade tumor with a favourable prognosis. Only a few cases have been reported to occur in submandibular gland. This report describes a case of right submandibular swelling which was diagnosed as a vascular tumor on initial imaging studies, thus, precluding aspiration cytology studies. Surgical excision of this tumor surprisingly revealed it to be a basal cell adenocarcinoma. Histopathological features of this entity have been described together with a literature review of the treatment modality of this rare tumor.
Keywords: Basal cell adenocarcinoma; submandibular gland; vascular mass; basaloid neoplasm

INTRODUCTION

Basal cell adenocarcinoma (BCAC) is a rare salivary gland neoplasm which occurs predominantly in the major salivary glands, most commonly in the parotid. Only a few cases have been reported to occur in the submandibular gland. Although, it is a low grade tumor with favourable prognosis, it has a propensity for local recurrences. FNA is highly sensitive at detecting basaloid neoplasms such as basal cell adenoma and adenocarcinoma, but distinction between several of the basaloid entities in the differential diagnosis is often not possible.

We report a case of right submandibular swelling which was diagnosed as a vascular tumor on initial imaging studies. Surgical excision of this tumor, however, revealed it to be a basal cell adenocarcinoma. Histopathological features of this entity have been described together with a review of the treatment modality.

CASE REPORT

A 28 year old man presented to our outpatient department with complaints of a painless and progressively increasing swelling in his right submandibular region for 4 months. There was no variation in size of swelling associated with food intake. Physical examination revealed a 5X4 cm firm, non-tender, mobile and non-pulsatile swelling in the right submandibular region (Figure 1). There was no palpable lymphadenopathy. Ultrasonography showed two highly vascular lesions measuring 3.5X3.7X3.1 cm and 2.1X2.2 cm close to the right submandibular gland. They showed heterogenous parenchyma and few hypo echoic areas. In view of these findings, a contrast-enhanced CT scan was done to better characterise as well as evaluate the extent of mass. It revealed a 5x3.5 cm well-defined mass in the right submandibular region with intense post-contrast enhancement. Owing to the loss of fat planes, it appeared to arise from the outer surface of the submandibular gland which was also displaced medially (Figure 2). These findings were suggestive of a vascular tumor originating from the submandibular gland. Digital subtraction angiography was done to evaluate the vascular supply of the tumor. It showed a well-defined, moderately hyper vascular blush supplied by hypertrophied submandibular branches of the facial artery and draining into the internal jugular vein via retromandibular vein. No intratumoral arteriovenous shunting was seen.

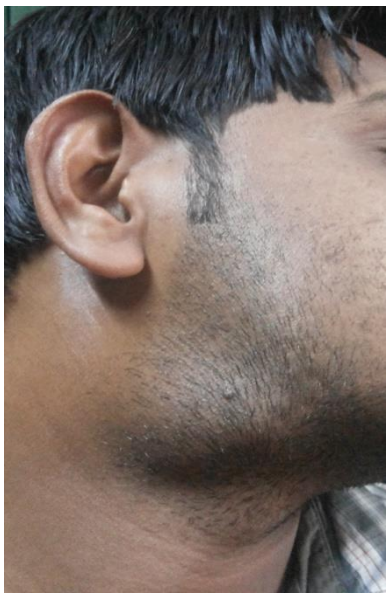

Fig-1:Right submandibular swelling

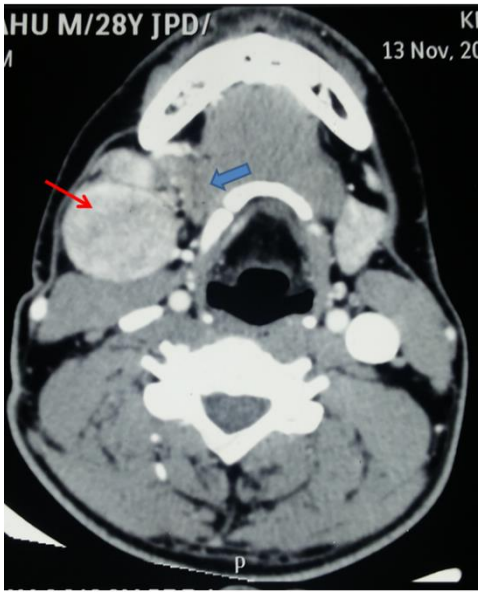

Fig 2. CECT showing intensely enhancing tumor (red arrow) arising from submandibular gland (blue arrow), which is displaced medially

The patient was taken up for transcervical excision of the vascular tumor under general anaesthesia. As anticipated, it was arising from the submandibular gland, and hence en-bloc excision of the tumor and submandibular gland was done (Figure 3-5). Marginal mandibular function was intact post-operatively (Figure 6).

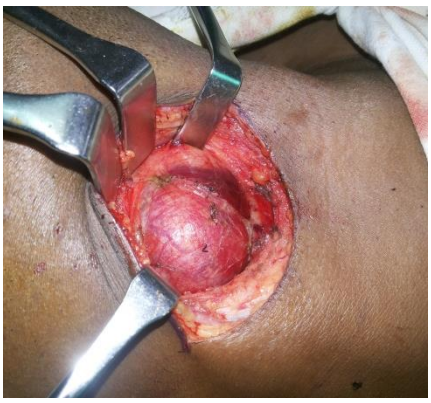

Fig 3.Intraoperative view of tumor

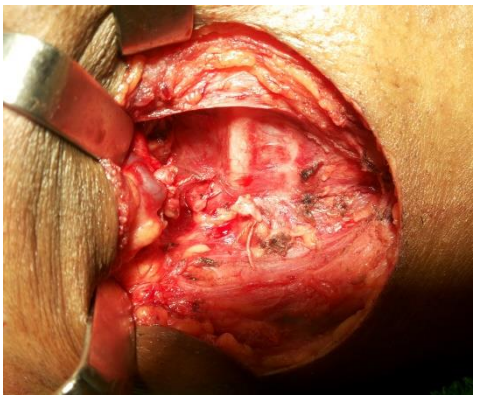

Fig 4.Tumor bed after excision of the mass during transcervical excision

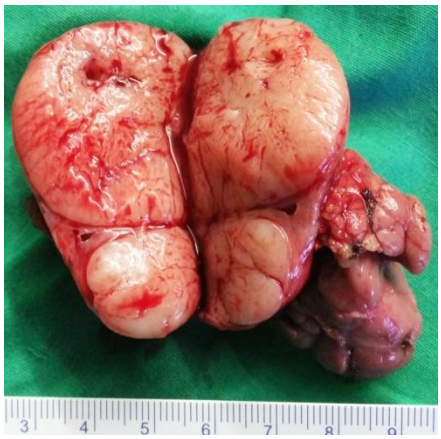

Fig 5.Resected specimen

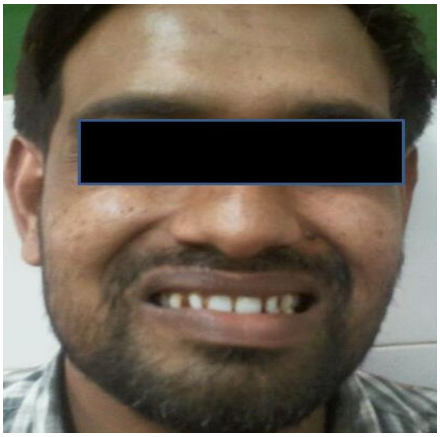

Fig 6. Postoperative intact marginal Mandibular function

Histopathological examination of the resected specimen revealed a partially encapsulated multinodular tumor with cells arranged in trabecular, tubular, solid sheets and nests separated by thin basement membrane material. Tumor cells showed peripheral palisading, cuboidal to columnar with round to oval nuclei with scant cytoplasm. The tumor was focally devoid of capsule and infiltrated the surrounding salivary gland and adipose tissue. The findings were suggestive of basal cell adenocarcinoma. The patient was advised long term follow-up. He has been recurrence free for more than two years now.

DISCUSSION

Although first recognised in 1978 [1], Basal cell adenocarcinoma (BCAC) was included in the 1991 World Health Organization classification of salivary gland neoplasms. Before the term BCAC was universally accepted, it was reported under a variety of names such as malignant basaloid tumor, malignant basal cell tumor, hybrid basal cell adenoma/ adenoid cystic carcinoma, basaloid salivary gland carcinoma and atypical monomorphic adenoma [2]. The histologic features of this tumor were described by Ellis and Gnepp in 1988. Subsequently, Ellis and Wiscivitch defined the clinicopathological features of basal cell adenocarcinoma [3]. Basal cell adenocarcinoma (BCAC), the malignant counterpart of basal cell adenoma, is a rare salivary gland neoplasm. It comprises 1.6% of all salivary gland neoplasms and 2.9% of malignant salivary gland neoplasms [2]. Ninety percent of cases arise from major salivary glands; usually the parotid.[4] Occasional cases have been reported in the submandibular gland and the minor salivary glands. We could find eight cases of BCAC of submandibular gland reported in literature till date [2, 5-7].

Salivary gland enlargement is the main presenting symptom, and uncommonly, mild pain or tenderness may also be present. The peak incidence is reported in the sixth decade of life without any gender predilection. FNA is highly sensitive at detecting basaloid neoplasms such as basal cell adenoma and adenocarcinoma, but distinction between several of the basaloid entities in the differential diagnosis is often not possible. Most basal cell adenocarcinomas are microscopically identical to basal cell adenomas except for the presence of an invasive histologic growth pattern. Because FNA does not detect parenchymal invasion, basal cell adenomas and adenocarcinomas are, for the most part, indistinguishable by FNA [8]. Recently, diagnostic fine-needle aspiration cytomorphology of these tumors have been described in isolated case reports. These include basaloid-cell clumps with hyper chromatic nuclei and scant cytoplasm, accompanied by characteristic peripheral features, including palisading poor cohesiveness, and/or intermingling with fat cells [9]. According to Tse *et al.;* the cytological features of BCAC are not distinctive, but the presence of two-cell populations with moderate pleomorphism and a rosette-like pattern with central, eosinophilic globules may assist with its differentiation from other salivary gland neoplasms [10].

Our patient had a similar presentation with a painless swelling of the salivary gland. However, the distinctive feature in our case was the highly vascular nature of the tumor as revealed on imaging studies. The intense post-contrast enhancement on CT scan and the hyper vascular blush on digital subtraction angiography confirmed it to be a vascular tumor. The presumptive diagnosis of a vascular tumor precluded any cytological studies by FNA. Hence, the patient was taken up for surgical excision of the tumor without any pre-operative tissue diagnosis.

BCAC is believed to arise from pluripotent ductal reserve cells. Histologically, BCAC is a tumor similar to BCA except that it grows in an invasive destructive fashion, often with perineural and/or vascular invasion. True invasion must be distinguished from both multi-modularity with a pushing type of growth pattern and multi-focal origin in adjacent salivary lobules, both of which are the features of BCA [11]. BCAC can be divided into four subtypes based on histological patterns: solid, trabecular, tubular and membranous [3]. Histopathologic appearance of the cells and presence of infiltration of the surrounding salivary gland and adipose tissue prompted the distinction between BCAC and basaloid adenoma in the present case.

The 2005 World Health Organization classification categorizes basal cell adenocarcinoma (BCAC) as a low-grade tumor with a favourable prognosis [12]. Although it does seem to have a propensity for local recurrences, metastatic disease is uncommon. Ellis and Wiscovitch reported recurrences

in 7 out of 29 cases of BCAC of major salivary glands. 3 of these metastasized to lymph nodes and lung [3].

Surgical excision with a wide margin to ensure complete removal has been suggested as the primary treatment for BCAC [13]. Enucleation or curettage is to be avoided. With complete and wide excision of a low stage tumor, adjunctive radiotherapy is not warranted. Zhan *et al.;* reviewed a large series of 509 cases of BCAC of major salivary glands and found that for high T-stage disease, surgery with radiation had significantly better survival than surgery alone [14]. Adjunctive radiotherapy has also been proposed for lesions in the minor salivary glands because of the higher likelihood of vascular and neural invasion [15]. Singh *et al.;* reported a case of BCAC of submandibular gland who underwent excision of the mass with 3 mm margins. Histopathological examination revealed positive margins in the resected specimen, thus, necessitating adjuvant radiotherapy [7]. Their case reiterates the fact that the surgeon should aim for complete excision of the tumor with wide margins during the first surgical procedure to provide the best chance for survival.

CONCLUSION

BCAC is a rare tumor of the submandibular salivary gland. Although FNA is highly sensitive at detecting basaloid neoplasms, basal cell adenoma and carcinoma cannot be reliably distinguished on the basis of cytological features. It may present as a vascular tumor on imaging, thus, precluding any pre-operative cytological studies. It is essential to perform complete surgical excision of the tumor with wide margins to reduce local recurrence and to provide best chance for survival.

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