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Case Report

Adenoid cystic carcinoma of the palatal minor salivary gland – case report

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ABSTRACT:
A patient with a history of asymptomatic gum pain reported swelling in his upper back region for one year. The pain was sudden, dull, and associated with nosebleeding. The pain spread to the palate and ear, and the patient struggled with chewing food. Upon inspection, the swelling was found in the posterior left maxillary region, extending from the mesial aspect of the canine to 2 cm anterior to the tuberosity on the left side, involving the palatal area but not crossing the midline. Radiographic and histopathological findings were conclusive for the diagnosis. The treatment plan suggested was surgical removal and chemotherapy, radiotherapy, and synergistic immunotherapy. The patient chose to get the treatment done at a higher centre in the country's capital.
Keywords: carcinoma, salivay gland, tumor, malignant, metastases

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INTRODUCTION

Some names for adenoid cystic carcinoma (ACC) include cyclinoma, adenomatous basal cell carcinoma, basaloid mixed tumour, and adenocystic basal cell carcinoma.[1] This aggressive neoplasm grows slowly but can recur remarkably. Adenoid cystic carcinoma ranks as the fifth most prevalent malignant epithelial tumour of the salivary glands, as reported by the American Cancer Institute (AFIP).[2] ACC of the maxilla is an uncommon but important salivary gland malignancy with variable clinical behaviour. This disorder is known for local invasion and distant metastases, making treatment difficult.[3-5] Distant metastasis to the lungs and bones occurs frequently, while metastasis to regional lymph nodes is rare.[6] ACC begins in the small salivary glands and often manifests as a slow-growing, painless lump in the maxilla that can be lethal.[7] Little is known about the molecular mechanisms that cause ACC. The fusion of the MYB (myeloblastosis) oncogene to the transcription factor gene NFIB (nuclear factor I/B) is caused by the ACC-specific t(6;9)(q22-23;p23-24) translocation, which was initially reported by Persson et al.[8,9] Chimeric transcripts including MYB and NFIB were generated by the chromosomal translocation t (6;9)(q22-23;p23-24), which leads to dysregulation of the MYB gene.[8] This further causes the constitutive activation of MYB-targeted genes, which in turn dysregulates important physiological processes involved in cell adhesion, cell cycle regulation, and death.[10] In addition to expressing elevated amounts of the transcription factor Sox4, ACCs also express genes linked to myoepithelial differentiation. In addition to being a possible human oncogene, the second one generally controls embryonic development.[11] Frizzled-7 and casein kinase 1-epsilon are two more overexpressed genes that have been linked to cancer and the Wnt/b-catenin signalling pathway.[12] They also tend to overexpress additional growth factor receptors, such as FGFR1, EGF receptor, and/or human epidermal receptor-2, and to generate excessive amounts of the receptor tyrosine kinase c-KIT.[8,11] The clinical features include its origin in 50 to 70% of occurrences in minor salivary glands with a higher predilection for females.[5,13] One percent of all head and neck cancers

are caused by this.[1,7] Still, it ranks second among big salivary gland tumours and first among small ones. In all, 10% of tumours in the salivary glands are caused by this. Although it's more common in the 50s and 60s, it can happen at any age; in fact, it's not uncommon in the 30s.[7] Early local discomfort, paraesthesia, fixation to deeper structures, local invasion, and paralysis of the facial nerve in cases of parotid tumours are some of its associated clinical symptoms.[3,5,7] Immunohistochemical studies showed that pseudocysts include basement membrane components like type IV collagen, heparin sulphate, and laminin isoforms and are positive for PAS and Alcian blue.[6,12] Epithelial cells express CEA and EMA. S-100 protein, calponin, p63, smooth muscle actin, and myosin are found in myoepithelial cells, and C-kit (CD117) is in duct lining cells.[7]Perineural invasion is associated with S-100, GFAP, and neural cell adhesion molecule expression.[5-7] P53 mutations may promote tumour growth and recurrence.Surgery, radiation, chemotherapy, and combination therapy can treat ACC.[5,8] Tumour cells spread beyond clinical and radiographic boundaries, requiring largest surgical margins.[7,9] Neutron therapy, which uses larger, more energetic particles, may manage localisation well.[8] Recent research suggest encouraging results for adoptive immunotherapy and chemoradiotherapy.[14] E-cadherin expression is helpful in prognosis.[15] High AgNOR levels may indicate metastases.[16] Anatomic location, primary lesion size, diagnosis-time metastases, perineural invasion, and histological variation are key survival variables.[5,7,8,14]

This article in the form of a case report presents a case of ACC that had originated in the maxilla nearly one year before its diagnosis. The article presents a brief overview of the aetiology, diagnosis, and management of such cases with a brief discussion of the probable similar conditions.

CASE REPORT

A 45-year-old male from Rampur, Uttar Pradesh, had experienced a growth in his upper back gum region for one year. The patient experienced pain in his upper back gum region a year ago, followed by swelling of the palate. The pain was dull, sudden, and associated with nosebleeding. The swelling initially was small but gradually increased to the present size. The patient also had difficulty chewing food. His dental history includes extractions [26 and 27] one year ago, with the patient reporting daily brushing with toothpaste and a toothbrush and being a chronic smoker for 2 years. The patient has no abnormalities in gait, nails, blood pressure, skin, pulse rate, or cyanosis. Patient has a height of 5ft 8 inches, weight 58 kg, built was endomorphic, afebrile temperature, no pigmentation and had a respiratory rate of 17 breaths per minute. The extraoral features included bilateral facial symmetry, normal temporomandibular joint, mouth opening, no tenderness, clicking, deviation, or deflection, and mandibular range of motion within physiological limits, with no evidence of tender masticatory muscle, and palpable lymph nodes **(Figure 1A)**. Soft tissue examination revealed no abnormalities in buccal, labial, gingival, alveolar, floor of mouth, and tongue. Periodontal status showed staining, calculus, bleeding on probing, mobility, pocketing, and recession. The patient's teeth had decay, with few missing. Occlusion was normal, with no signs of trauma. Saliva was normal, and the tooth wear pattern was absent of attrition, abrasion, and erosion.

Analysis of area of interest: The posterior part of the left maxillary bone was swollen, and it reached all the way to the palate **(Figure 1B, C)**. It was observed to go from the back of the canine all the way to the left side, two centimetres before the tuberosity, touching the palatal region but without going across the midline. Over the bulge, the mucosa was smooth and joined the surrounding mucosa seamlessly. On palpation, the swelling felt soft. Palpation verified the results of the inspection. Even though the affected teeth were immobile, probing revealed that they were painful. The swelling was solid in texture, not painful, and it didn't move. In addition, a greyish patch could be made out. After gauging a differential diagnosis that included mucoepidermoid carcinoma, ameloblastoma, central giant cell granuloma, and adenoid cystic carcinoma, investigations in the form of an orthopantomograph (OPG), cone beam computed tomography, and excisional biopsy were considered. The OPG showed a multilocular lesion that was present on the left side of the maxilla **(Figure 1C)**. It was clear from the OPG and CBCT that the buccal cortical plate had thinned **(Figure 2 A - C)**, and the lesion has a corticated and non-sclerotic appearance. Invasion of the maxillary sinus, palatine process of the maxilla, and osteomeatal complex of the ipsilateral side were all characteristics of the lesion, which also exhibits deviation of the nasal septum to the right side. The lesion on the CBCT measured 32.25 by 46.63 by 32.55 millimetres. Alveolar mucosa was subjected to an incisional biopsy for histopathological purposes. The results of the biopsy showed that the tumour cells are basaloid and grouped in the form of nests, sheets, and strands of varied sizes and shapes, with a cribriform and tubular pattern (Figure 2 D). Mucin pooling was seen in the cystic spaces, and there were a few regions that exhibited substantial amounts of cystic degeneration. The response of the stroma was characterised by a modest persistent inflammation. In addition, the stroma exhibited areas of hyalinization and haemorrhage in certain regions. Histopathological features indicated the presence of adenoid cystic carcinoma with a poor prognosis. The treatment plan suggested to the patient included surgical and non-surgical interventions, followed by a restoration and maintenance phase, which included counselling on tobacco cessation, surgical resection, and chemotherapy and radiotherapy.

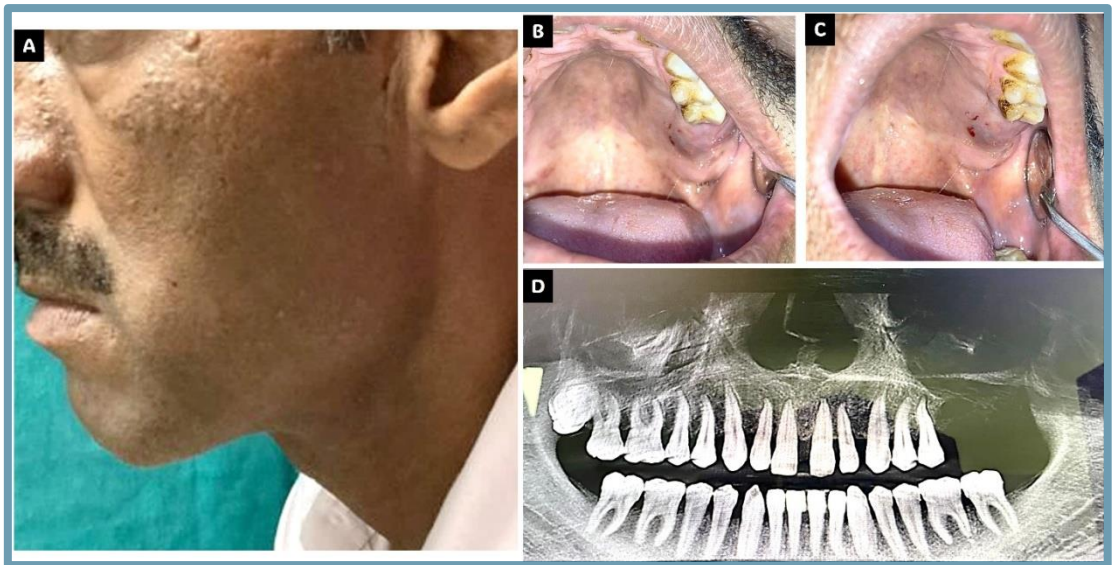

Figure 1: (A) Lateral view of the face (B) Intral oral view showing maxillary tuberosity region lesion (C) Intra oral view showing palatal aspect of the lesion (D) Orthopantomograph showing a lesion on the left maxilla

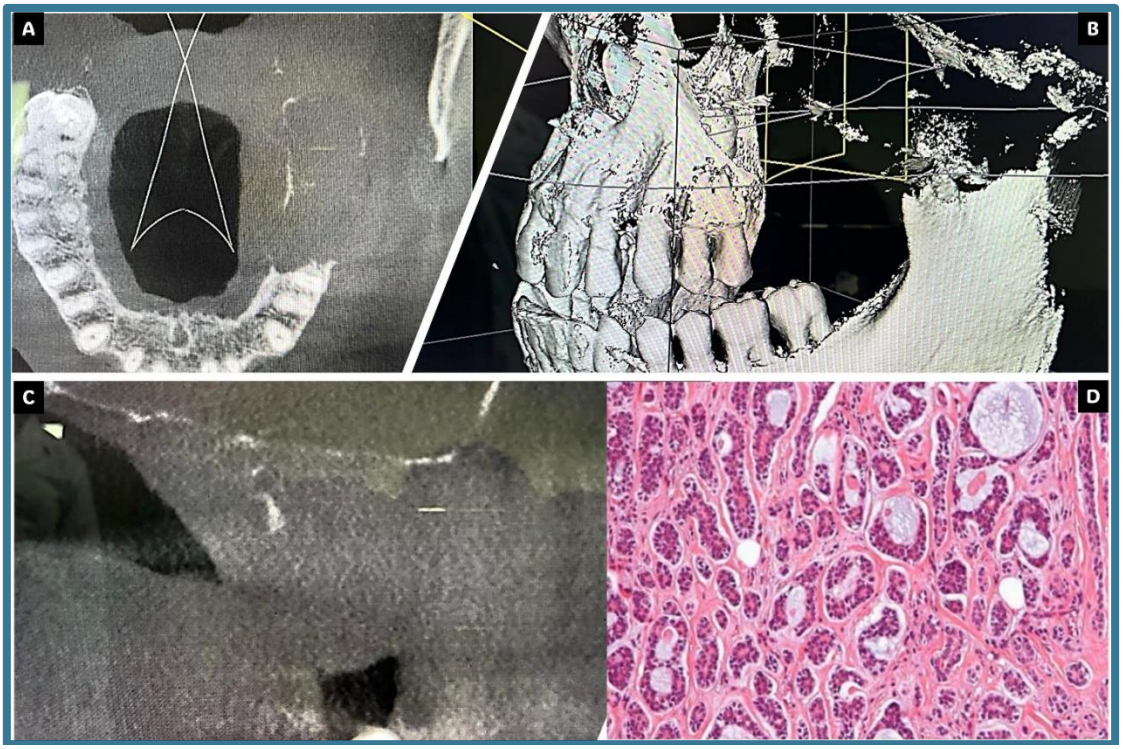

Figure 2: (A) Cone beam computed tomography occlusal view showing the extent of lesion (B) Frontal view with CBCT (C) Detailed CBCT image showing relation of lesion with bone (D) Histopathological image showing the cells involved.

DISCUSSION

A clinical case report presenting a case of ACC on the left palatal region of the maxilla has been presented. The case is unique as it is one of the rarest tumours involving minor salivary glands of the maxilla. The type associated in this case presented a typical gradually growing mass with no evidence of numbness or nerve involvement. The final diagnosis is through histopathological examination confirming the classical presentation of salivary gland cells. Myoepithelial cells and ductal cells, arranged in a variety of ways, make up adenoid cystic carcinoma.[18] Tubular, cribriform (classic), and solid (basaloid) are the three morphological growth patterns.[5,7,9] Basaloid epithelial cell nests create numerous cylindrical cyst-like formations in the cribriform pattern, which is similar to a honeycomb or Swiss cheese pattern.[7,9,17] Structures bordered by stratified cuboidal epithelium are revealed by the tubular pattern. Visualising solid groups is the solid pattern of cuboidal cells with little

tendency towards duct or cyst formation.[9] The cribriform pattern is the most common pattern, whereas the solid pattern is the least common one.[7] Invasion of perineural space and peripheral nerve is a characteristic feature of the tumour, although it was not found in this case. Invasions of neighboring structures and poorly delineated boundaries with radiographic evidence of bone deterioration are key radiographic findings, as observed in this case. Advanced artificial intelligence-based radiography has resulted in better radiographic pictures and understanding of most of the carcinomas.[19] ACC must be differentiated clinically from other similar clinically appearing conditions like mucoepidermoid carcinoma, pleomorphic adenoma, and squamous cell carcinoma.[7,14,17] Mucoepidermoid carcinoma is the most prevalent minor salivary gland cancer in adults. Mucoepidermoid carcinomas typically develop in the parotid gland and hard palate, causing harmless swelling with or without facial nerve involvement.[18] Clear mucin-containing cells usually stain reddish pink with mucicarmine stain in histology. Common radiographic characteristics of low-grade mucoepidermoid carcinomas include well-defined masses with cystic components.[9] Calcification and solid component enhancement can occur. One mobile, slow-growing, painless lump may be present for years in pleomorphic adenoma. When the parotid gland tumour grows or becomes malignant, facial nerve paralysis occurs. Rapid tumour nodule growth suggests malignancy.[7,17] Depending on the location, minor salivary gland tumours might cause dysphagia, hoarseness, dyspnoea, chewing difficulties, and epistaxis.[20] Radiographs show a radiolucent defect with well-defined cortical margins, bevelled margins indicating an intraoral origin, confinement within the osseous defect or its projected circumferential diameter if complete erosion has occurred, and no periosteal reaction or hard palate involvement. The most frequent oral cancer is SCC. It appears as a red or pink ulcer that bleeds easily over the alveolus, jaw, or hard palate bone. Crepitus, lip anaesthesia, and jaw discomfort from pathologic fracture are other complaints.[14] It is typical for cancer to cause radiolucent lesions in the jawbones.7 Peripheral or mucosal SCC damage bone. Peripheral radiographs indicate lytic defects. Both types of radiolucent lesions may have ragged bone sequestra.[18] One must always rule out similar clinically looking lesions, including the ones that are anatomical, like bony exostosis.[21, 22]

Kim GE, et al. analysed the clinical features and outcomes of patients with ACC of the maxillary antrum. Results showed that local recurrence was the most common site of failure (72.7%), with a lower local recurrence rate (40%) in patients treated with surgery alone or radiation alone. Neck node failure was rare, and distant metastases were documented in seven patients (32%). Overall survival and disease-free survival rates were 37.6% and 13.6%, respectively.[5] Horrée WA found 28 patients with adenoid cystic carcinoma of the maxilla had poor prognosis, indicating doubts about the effectiveness of complete tumour removal. The study suggests a "three-modality therapy" including fluorouracil infusion, maxillary resection, and radiotherapy for a possible curative effect.[6] Cleveland D, et al., in their seven cases of ACC of the maxilla, with clinical and radiographic characteristics suggesting its origin in the maxillary alveolar bone. Four of those five patients died, confirming the lethality of this solid variant.[7] The age of this patient conforms with earlier reports of patients being in their forties.[23,24] The clinical feature of painless swelling also is in accordance with studies that report it as a common clinical sign. [25] Surgical correction is usually extensive and therefore leads to permanent deformation of the face, which can be cosmetically corrected using a maxillofacial prosthesis.[26]

CONCLUSION

The study of adenoid cystic carcinoma (ACC) highlights its unique pathophysiological characteristics, necessitating effective surveillance protocols and personalised treatment strategies. It also underscores the need for further research into biomarkers for early detection and prognosis. A multidisciplinary approach, including surgical resection and radiation therapy, is crucial for improving patient management and survival rates.

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