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A Rare Case Report of Adenoid Cystic Carcinoma of the Anterior Maxilla in a Young Patient

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ABSTRACT

Background and Objective: Adenoid cystic carcinoma (ACC) of the maxilla is a rare malignancy arising from minor salivary glands. The clinical and radiological appearance may be similar to any odontogenic/nonodontogenic pathology. This study aims to report a rare case of primary central ACC of the anterior maxilla.

Case Report: A 31-year-old man was referred to the Department of Oral and Maxillofacial Surgery complaining of swelling and pain in the right anterior maxilla for 3 months. The patient has noticed swelling and pain in this side since about five months before and instead of diagnosing this tumoral lesion, false diagnosis and unsuccessful root canal therapy were considered. A biopsy was performed in the central region and microscopically, the lesion showed tumor cells arranged in sheets in fibrous stroma islands of epithelial cells showing a classical "Swiss cheese" pattern. Based on clinical, radiographic, and CBCT evaluation and positive immunohistochemistry of CD63, and C-Kit confirmed ACC diagnosis. After the diagnosis, the lesion was completely removed by enucleation and curettage performed by the surgeon. Postoperative radiotherapy was performed. Follow-up within 3 years since the initial diagnosis showed no sign of recurrence.

Conclusion: Based on the results of this study, early diagnosis and treatment of ACC can lead to successful treatment and patient survival.

Keywords: *Adenoid Cystic Carcinoma, Salivary Gland Malignancy, Anterior Maxilla, Immunohistochemistry.*

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Introduction

Adenoid cystic carcinoma (ACC) is a malignant neoplasm of epithelial origin and is very rare in the jaw and face, and less than 1% of head and neck malignancies can affect the major or minor salivary glands of the oral cavity (1, 2). The occurrence of ACC in the jaw bone is a very rare phenomenon, and among the salivary glands, mucoepidermoid carcinoma tends to occur in the jaw bone, and most of these tumors are reported in the posterior mandible of adults (3). Intraosseous adenoid cystic carcinoma (ACC) is an extremely rare neoplasm with only a few cases reported to date causing expansile bony destruction and affecting the mandible more commonly than the maxilla (1). Although the occurrence of ACC in the maxilla is rare in the paranasal and nasal sinuses, it is very rare in the anterior part of the maxilla and may arise and originate from the seromucous of minor salivary glands mucosa of the maxillary sinus, which is trapped during the embryonic stages of maxillary bone formation (4). The report of 26 cases of primary ACC of the mandible until April 2009 in the literature suggests its rarity (1). Also, a case of advanced ACC of maxillary sinus was reported by Lahjaouj et al., according to which adenoid cystic carcinoma of the right maxilla occurred in a 55-year-old man whose diagnosis was delayed due to failure to track symptoms (3).

The clinical findings of this malignant tumoral lesion usually include slow growth, local recurrence, pain due to peripheral nerve invasion (5, 6), and distant metastasis (7). This tumor frequently occurs in the 5th and 6th decade of life and usually affects women (8, 9). Among these subgroups, the solid type is the most aggressive (9). One of the unique features of ACC is the tendency to invade the peripheral nerves, which can be seen even in the early stages. Six cases with ACC were reported in 2015, which require long-term follow-up (10). ACC is one of the most biologically destructive and unpredictable head and neck tumors and is extremely difficult to treat.

This study aims to report a rare case of primary central ACC of the anterior maxilla with an unusual occurrence in terms of location, gender, and age with relatively fast growth within five months.

Case Report

This study was approved by the ethics committee of Mazandaran University of Medical Sciences with the code IR.MAZUMS.REC.1401.136. The patient was a 31-year-old man who was referred to the surgical department of Mazandaran Dental School in Sari in 2018. The patient had a history of pain and swelling in the anterior region of the maxilla in canine tooth, and after visiting the dentist and obtaining a periapical radiograph, a diagnosis of acute apical periodontitis was made for the mentioned tooth, and then root canal treatment was performed. However, after three months, the patient's toothache did not improve but the lesion was bigger and its swelling increased. The central and lateral teeth of the patient were loose, so the patient was referred to a maxillofacial surgeon. A wide lucent lesion was evident in the area of the canine to the central and lateral teeth. Root deviation or resorption was observed. The patient had no history of drug use, smoking, or systemic disease. Also, he had no history of head and neck surgery or chemotherapy. According to the patient's report, the lesion was created in the maxilla five months ago and started to grow rapidly with time. The extraoral examination indicated the absence of regional lymphadenopathy, and the skin surface was normal but slightly prominent due to the underlying swollen lesion. In the intraoral examination, the overlying mucosa appeared normal but a sore and painful swollen lesion with a broad base was observed on the right side of anterior maxilla. The mentioned lesion had a firm consistency with a smooth surface that progressed toward the alveolar ridge with approximately 3×4 cm extension (Figure 1).

Cone-beam CT (CBCT) and panoramic photo were performed for the patient and a large lucent lesion was observed in the right side of the maxilla from the central to canine teeth (Figures 2, 3). The possibility of a malignant tumor such as MEC, PLGA and also odontogenic cyst was considered based on clinical and radiographic findings. Routine hematologic assessments were normal.

Then, an incisional biopsy of the lesion was performed by the surgeon and it was sent to an oral, maxillofacial pathology center in 10% formalin (figure 4). The tissue was brown with an elastic consistency, measuring 1.5×1.5×5.3 cm.

Figure 1. swollen lesion on the right anterior side of the maxilla

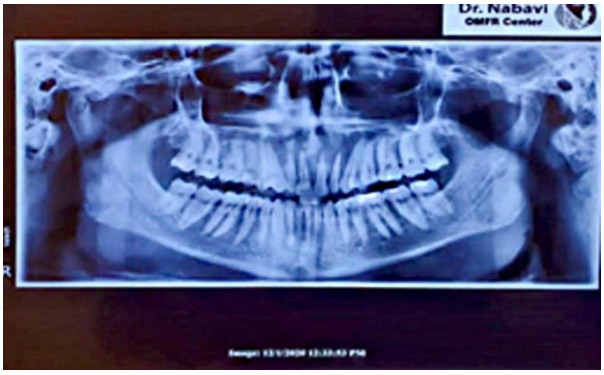

Figure 2. panoramic view of the lesion of the right side of the maxilla from the central to canine teeth

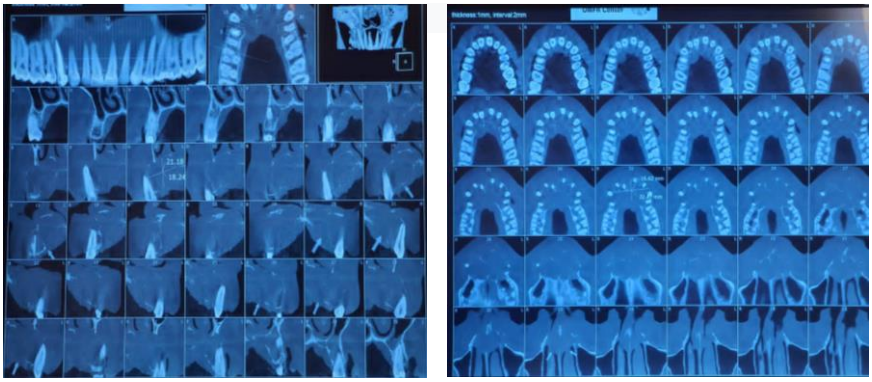

Figure 3. CBCT of the lesion

Figure 4. Biopsy from swollen lesion on the right anterior side of the maxilla

Microscopically, the lesion showed overlying mucosa with no abnormality. The tumor consisted of the proliferation of malignant neoplastic tissue of solid pattern with basaloid type tumor cells arranged as islands of basaloid epithelial cells containing numerous spherical spaces showing a classical "Swiss cheese". It included cyst-like spaces containing basophilic mucoid material that show stroma. Abundant plump fibroblasts, numerous blood vessels and few lymphocytes interspersed among fine moderately fibrillar collagen bundles were noticeable. The majority of the areas (>80%) showed a cribriform pattern; hence, the diagnosis of ACC was confirmed (Figures 5, 6).

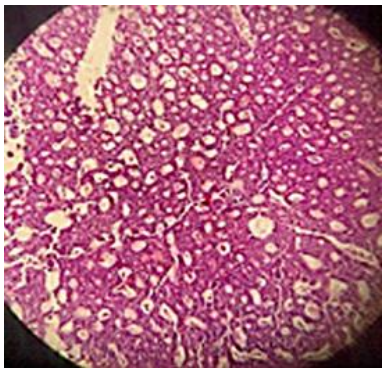

Figure 5. Microscopic view of adenoid cystic carcinoma using hematoxylin and eosin staining with 40x magnification with cuboidal cells with prominent hyperchromatic nuclei in duct-like tubules

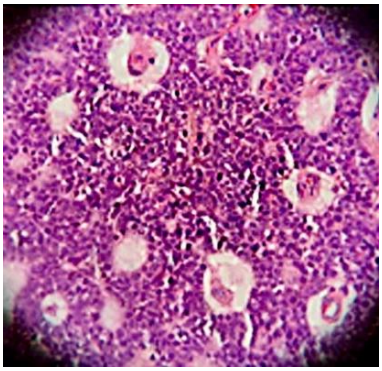

Figure 6. A microscopic view of adenoid cystic carcinoma using hematoxylin and eosin staining with 100X magnification

To definitively diagnose the immunohistochemical tissue, it was suggested that C-Kit (CD117) and CD63, markers were positive and confirmed the mentioned diagnosis (Figures 7, 8).

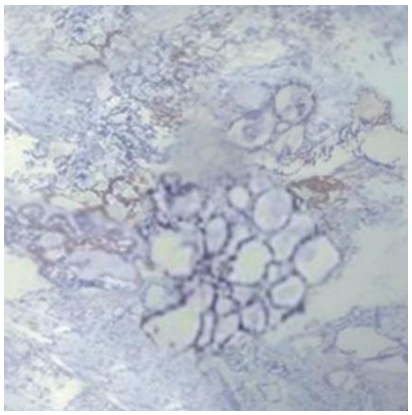

Figure 7. Moderate staining in some tumor cells CD117 immunopositivity

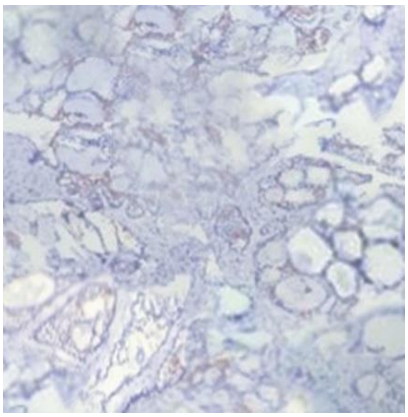

Figure 8. P63 positive in most tumor cells

Finally, after the diagnosis, the lesion was completely removed by enucleation and curettage and the pathology report showed the final diagnosis of adenoid cystic carcinoma and tumoral epithelial tissue without invading the surrounding connective tissue. At the time of tumor diagnosis, chest x-ray and bone scan were performed for the patient and no metastasis was observed. After two days, the patient was discharged in good general condition. Considering these results, the patient was then referred to the Department of Oncology for evaluation. The patient received 30 sessions of radiotherapy with a total dose of 60 Gy executed in the fractionation of 2 Gy/day for 3 days/week and every 45 days an MRI test was taken. In three-year follow-up of the patient, no recurrence or metastasis was observed until 2023.

Discussion

The most important finding that led to the report of this case was bone swelling in anterior maxilla and pain and a large lucent lesion in the radiographic view in a 31-year-old man. As seen in our study, swelling, and pain are the most common clinical findings of this tumor (8, 9). Contrary to other reports, our patient did not report any symptoms of paresthesia, hypoesthesia, or other unpleasant feelings, which is important in this respect (11, 12), which led to being mistaken for a dental infection in the first stage of treatment. In

some cases, tooth loss, loose teeth, and ear pain are observed with adenoid cystic carcinoma (1, 13). Loose teeth were also observed in our report.

The presence of salivary gland tumors inside the jawbone is rare. The pathogenesis of central salivary gland neoplasms is unknown. Some researchers have proposed the following factors which could play a role in the origin and formation of central malignant salivary gland tumors, which include: entrapped ectopic salivary gland tissues in jaws during embryological development of the jawbones as seen in our case. Neoplastic transformation of the mucus‑secreting cells commonly found in the epithelial linings of dentigerous cysts or any sinus pathology. Submandibular and sublingual glands closely apposed in bony defects or cavities in the lingual cortex of the mandible or fragments of these glands that have undergone embryologic evagination (4).

Differentiation between adenoid cystic carcinoma and other salivary gland tumors such as salivary duct carcinoma is histologically important. The immunohistochemical examination helps in the definitive diagnosis of these lesions (14). In our case, the tumor cells were positive for CD63, and C-Kit (CD117) markers, so the definitive diagnosis of adenoid cystic carcinoma was confirmed by immunohistochemistry.

The presence of intact cortical plates, radiographic evidence of bone destruction, covering the lesion in the mucous membrane, histological examination, and excluding the diagnosis of an odontogenic tumor or other primary salivary tumors are necessary to confirm the final diagnosis (12). In our patient, the tumor first occurred in the maxilla, after which the tumor mass spread widely and advanced toward the alveolar ridge.

Surgery is the predominant treatment modality for central salivary gland tumors of the jaws. It ranges from enucleation or curettage to en bloc or radical excision (3). Although surgery is the best treatment for adenoid cystic carcinoma, complete resection is often difficult to achieve due to vascular invasion and perineurial infiltration specialty in maxillary sinus such as an advanced case proposed by Lahjouj et al, which was extended to the maxillary sinus (3). Postoperative radiation therapy enhances local and regional control in ACC. In our case, enucleation and curettage with radiotherapy were performed and after 3 years of follow-up, there was no evidence of recurrence which was similar to the research of Indu et al., whose patient had ACC in anterior mandible (15). Therefore, in some cases, postoperative radiation therapy is necessary. Adenoid cystic carcinoma is sensitive to radiation therapy, however, it is not curable with this method (16). Postoperative radiation therapy improves local and regional control of adenoid cystic carcinoma. For inoperable tumors, a dose of 60 to 70 Gy is recommended for the primary lesion (17). In our patient, radiation therapy was performed 30 sessions after the operation, and local control was achieved.

Despite the local control of the tumor, distant metastases develop in about 40-60% of patients. The lung and bone are the most common sites of spread, but the very slow growth pattern of the tumor and the gradual appearance of metastases allow patients to live a normal life for years. A study has shown that in 17 cases of mandibular central adenoid cystic cancer, lung metastases developed in 4 cases, but bone metastases did not occur in any of them (12). However, adenoid cystic carcinoma with bone metastasis may progress faster than lung metastasis. Adenoid cystic carcinoma has 3 histological subgroups, including tubular, cribriform, and solid types. Histological subgroup is one of the most important factors in the prognosis of patients. Tubular type has the best prognosis. The solid type of adenoid cystic carcinoma causes distant metastases faster and indicates a more serious prognosis than tubular and cribriform types (18). In our case, according to the patient's faster referral and treatment, alveolar bone destruction and inferior nerve canal invasion were not reported. Early diagnosis with a 10-year survival rate is estimated at 75%, while the survival rate gradually decreases with the progression of the disease. It seems that in our reported case, the absence of

paraesthesia indicates the absence of perineural invasion, which justifies the presence of a tumor without metastasis. Finally, it should be stated that in malignant and dysplastic lesions, early diagnosis in early stages and performing biopsy, will lead to better treatment and prognosis of the patient (19, 20). Many patients with such disease suffer from its complications and ultimately die because of inappropriate treatments and delayed diagnosis. Therefore, detailed histopathological examination and definitive diagnosis can lead to successful treatment (21, 22).

Adenoid cystic carcinoma in our case is one of the most destructive and unpredictable head and neck tumors with very difficult treatment, so quick diagnosis was crucial for patient survival.

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