

# Case study

## **ADENOID CYSTIC CARCINOMA, AN ENIGMA UNFOLDED – A CASE SERIES**

### **Abstract**

*Adenoid cystic carcinoma is a rare, slow-growing cancer that typically affects the salivary glands in the head and neck area and accounts for only 1% of all head and neck malignancies. The cancer tends to spread around nerves and can also spread to other parts of the body through the bloodstream. In this case series, we present three rare cases of adenoid cystic carcinoma. The first case involved a 65-year-old woman with follicular adenocarcinoma of the oral mucosa, which was successfully managed with surgery involving resection and removal of the upper jaw, followed by reconstruction of the temporal muscle. The second case involved a patient with nasopharyngeal adenocarcinoma who received maxillary surgery and radiation therapy. The third case was a rare case of adenoid cystic carcinoma at the posterior edge of the tongue, which was treated with mandibular surgery and partial tongue resection. All three cases were followed up for an extended period, and there was no recurrence observed. Based on our experience and that of other authors, we conclude that surgery is the primary treatment for head and neck adenoid cystic carcinoma, and radiation therapy and chemotherapy can also be used.*

**KEYWORDS:-** Adenoid, carcinoma , malignant, tumour , cystic.

### **1. INTRODUCTION**

“Adenoid cystic carcinoma (ACC) is a rare, slow-growing malignant tumor of the head and neck that was first described by Billroth in 1856 as cylindroma”.<sup>(1)</sup> “It accounts for approximately 1% of all malignant tumors in the oral and maxillofacial region, as well as about 22% of all malignant tumors in the major and minor salivary glands”.<sup>(1)</sup> The tumor growth patterns are characterized as cribriform, tubular, and solid, and lymphatic spread to the neck is uncommon<sup>(1)</sup>, while hematogenous and perineural dissemination occurs frequently in the course of the disease.<sup>(1)</sup> “ACC can involve lacrimal and ceruminous glands, as well as other sites in the head and neck, including the nasal and paranasal sinuses, trachea, and larynx”.<sup>(2)</sup> “It is the most commonly reported malignant tumor of the minor salivary glands and is also one of the most common cancers of the major salivary

glands. The treatment of choice for ACC is total resection, followed by postoperative radiotherapy<sup>(2)</sup>. Although chemotherapy has been studied in patients with advanced ACC, the indolent nature of the disease makes it difficult to observe clinical responses. In this article, we present a case series of three patients with ACC of the head and neck region and discuss their management.

## 2. CASE REPORT :-

### CASE 1 :

A 55 year old female patient reported with swelling in the right side of the face .History revealed slow growing swelling over past 6 months with no complaint of pain or discharge (Fig 1a ) . Extra oral examination revealed a well-defined swelling on the right side of the face measuring approx.. 5x5x5 cm ,extending from infra orbital margin to the corner of the mouth inferiorly (Fig 1b ) . The temperature of the overlying skin was not raised, and the consistency was firm and the tumour mass was well capsulated. The overlying skin was normal and not involved. No lymphnodes were palpable in the neck region. CT revealed sub mucosal bulging, mass in right maxillary sinus involving the buccal mucosa that measured 3\*3\*4 cm .

Biopsy revealed adenoid cystic carcinoma of minor salivary gland . Treatment in this case was surgical excision followed by reconstruction with temporalis flap(Fig 1c, Fig1d ,Fig 1e)

### CASE 2 :-

A 35 year old female patient reported with a complaint of swelling in the posterior region of right palatal shelf for the past 4 months associated with mild pain and discomfort. Intra oral examination revealed a well-defined swelling on the right palatal shelf measuring approximately 3\*3 cm and extending from the area distal to 3<sup>rd</sup> molar upto the mesial of 1<sup>st</sup> molar area (Fig 2a) . The overlying skin was ulcerated and erythematous. A MRI revealed a well-defined swelling measuring 2\*3 cm.

Biopsy revealed ACC of palate. Treatment of choice in this case was surgical excision of tumour mass and maxillectomy followed by radiation therapy(Fig 2b ,Fig 2c ).

### **CASE 3 :-**

A 65 year old female patient reported with a complaint of swelling on the right lateral border of the tongue for 1.5 years. History revealed an ulcerated swelling on the right lateral border of the tongue, from the posterior 1/3<sup>rd</sup> to the middle 1/3<sup>rd</sup> of the tongue, swelling was non tender , ulcerated and erythematous(Fig 3a) . The swelling was firm and well demarcated, measuring 2\*3 cm. No palpable lymph node was in the neck.

Fnac confirmed ACC of tongue. In this case tongue was managed surgically with access mandibulotomy and partial glossectomy(Fig 3b, Fig 3c). All cases were kept on long term follow up and no recurrence was seen.

### **3. DISSCUSSION:-**

ACC occurs mainly in the fourth and sixth decades of life with a slight predisposition to women, in our case all patients were in the 4th and 6th decades of life.

ACC is described as having a seemingly slow course; however, it has an aggressive behavior in the long run. The most common symptom is a slow-

growing tumor, followed by pain due to a tendency to invade around the nerve.<sup>[2]</sup>

In the major salivary glands, the tumor forms a mass, and when located in the parotid gland, paralysis of the facial nerve may occur; in the palate, a common mass, although a nodule or even a fistula in the antrum may be seen.<sup>[3]</sup>

Three distinct architectural patterns have been described: tubular, cribriform and solid. In our 3 case reports the biopsy samples showed cribriform and solid pattern.

AdCC treatment is influenced by tumor location, stage at diagnosis, and biological behavior, as evidenced by histological grade. AdCC's "gold standard" treatment, considered resectable after thorough work-up, is radical resection, ensuring a free margin, and postoperative radiotherapy.

Mendenhall et al. compared radiation therapy alone with radiation therapy plus surgery and concluded that combined treatment was better. AdCC has a strong tendency to infiltrate adjacent tissues, especially due to perineural invasion, so that even in 'resectable' AdCC, the 'free margin' goal is often not achieved.<sup>(2)</sup>

Due to the historically low rate of occult lymph node metastasis, cervical lymph node dissection is performed only for clinically positive lymph nodes. Clinically obvious lymph node metastases are uncommon in AdCC, especially for the parotid gland. However, for minor salivary gland subsites, the incidence of involved lymph nodes appears to be higher. minima et al.<sup>(2)</sup>

The differential diagnosis of ACC is (a) polymorphous low grade adenocarcinoma, (b) Basal cell adenoma and basal cell adenocarcinoma, (c) Pleomorphic adenoma.

Treatment of choice in all our cases was removal of complete tumour mass, no lymph nodes were found in neck so neck dissection was not performed. In all the cases no recurrence was seen on follow up upto 5 years.

#### **4. CONCLUSION .**

In conclusion Adenoid cystic carcinoma are seemingly innocuous lesion , which shows slow growth but due to their propensity for the perineural spread and distant metastasis ,require prolonged follow up.

#### **5. CONSENT :-**

Written consent has been taken from all the patients .

#### **Ethical Approval:**

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

**Figures**



FIGURE 1a

FIGURE 1b

FIGURE 1c



FIGURE 1d

FIGURE 1e



FIGURE 2a

FIGURE 2b

FIGURE 2c



FIGURE 3a

FIGURE 3b

FIGURE 3c

### LEGENDS

Figure 1a	well-defined swelling on the right side of the face measuring 5*5*5 cm
Figure 1b	Well defined swelling from worms view
Figure 1c	Weber fergusons approach for resection of tumour mass
Figure 1d	Resected tumour mass along with right palatal shelf
Figure 1e	Temporalis reconstruction
Figure 2a	well-defined swelling on the right palatal shelf measuring approximately 3*3 cm
Figure 2b	Maxillectomy performed
Figure 2c	Resected ACC of palate
Figure 3a	ulcerated swelling on the right lateral border of the tongue
Figure 3b	Mandibulotomy along with partial glossectomy
Figure 3c	Plating of mandible after removal of tumour

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