

# Case study

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

### **Abstract**

*BACKGROUND :- Adenoid cystic carcinoma is rare, slow-growing malignant tumour of the head and neck , which most commonly involves the salivary glands. It accounts for approximately 1% of all head and neck malignancies, and accounts for 10% of all salivary gland tumours. The tumour has a tendency of perineural spread along with haematogenous spread to distant organs.*

*CASE SERIES: - This is a case series of 3 cases, case 1 is a rare case of adenoid cystic carcinoma of buccal mucosa in a 65 year old women, diagnosed and managed surgically, with maxillectomy and temporalis muscle reconstruction; case 2 is a case of adenoid cystic carcinoma of palate treated surgically with maxillectomy and radiation; case 3 is a rare case of ACC of the posterior border of the tongue managed surgically with access mandibulotomy and partial glossectomy.*

*CONCLUSION: - All the cases were kept on long term follow up and no recurrence was seen. Various authors have concluded that surgery is the mainstay of managing ACC of head and neck, followed by radiation and chemo-therapy .*

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

### **1. INTRODUCTION**

Adenoid cystic carcinoma is rare, slow-growing malignant tumour of the head and neck , first described by Billroth in 1856 and termed cylindroma.<sup>(1)</sup>

ACC is a rare tumour entity and forms about 1% of all malignant tumours of the oral and maxillofacial region and about 22% of all malignant tumours of the major and minor salivary glands

<sup>(1)</sup>. Growth patterns are characterized as cribriform, tubular and solid . Lymphatic spread to the neck remains rare<sup>(1)</sup>. Haematogenous and perineural dissemination occurs often in the course of the

disease<sup>(1)</sup>. The treatment of choice consists of total resection. We present a case series of three cases of ACC of head and neck region with 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 lymph nodes 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. DISCUSSION:-**

ACC predominantly occurs in fourth and sixth decade of life with a slight female predilection , in our case also all the patients were in 4<sup>th</sup> and 6<sup>th</sup> decade of life.

ACC has been described as having an apparently indolent course; however, it has an aggressive long-term behaviour. The most common presenting symptom is a slowly growing mass, followed by pain attributed to its tendency for perineural invasion <sup>[2]</sup>. In major salivary glands the tumor produces a mass, and when located in the parotid, facial nerve palsy may occur; in the palate a mass is common, though ulceration or even oro-antral fistula 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.

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 .

#### **6. ETHICAL APPROVAL**

It is not applicable.

**Figures**



**FIGURE 1A**



**FIGURE 1B**



**FIGURE 1C**



**FIGURE 1D**



**FIGURE 1E**



**FIGURE 2A**



**FIGURE 2B**



**FIGURE 2C**



FIGURE 3A



FIGURE 3B



FIGURE 3C

UNDER PEER REVIEW

## 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                                     |

## References

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