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Adenoid Cystic Carcinoma of Head and Neck Region: Experiences at a Tertiary Care Center in South India

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ABSTRACT

Background: Adenoid cystic carcinoma is a rare malignant tumor of the salivary glands. It accounts for less than 1 percent of all head and neck malignancies. It is a slow-growing aggressive tumor with increased chances of recurrence.
Objectives: To report the varied presentations and to discuss the clinical profile and management of six diagnosed cases of adenoid cystic carcinoma of the Head and Neck region.
Method: A retrospective study consisting of 6 diagnosed cases of adenoid cystic carcinoma of the Head and Neck region which were treated at our tertiary care center between the period of January 2009 till December 2022.
Results: In our study, the mean age of occurrence of the disease was 52.5 years and 4 out of 6 patients (66.7%) were females. Out of the 6 cases, 3 patients were diagnosed with adenoid cystic carcinoma of the parotid gland, 1 patient with the maxilla, 1 sphenoid sinus, and 1 patient with the base of the tongue. All patients underwent surgical excision of the primary tumor and two of the patients had recurrence. Positive microscopic margins were reported in only one patient. Microscopic perineural invasion was seen in 2 patients.
Conclusion: Primary treatment is still surgical resection with radiotherapy. Adenoid cystic carcinoma of minor salivary glands has a worse prognosis than those of major salivary glands. Post-operative radiation is a must to delay rather than prevent local recurrence.

Keywords: Adenoid cystic carcinoma; Parotid; Maxilla; Sphenoid sinus; Tongue

INTRODUCTION

Less than 1% of head and neck cancers and 10- 15% of salivary gland neoplasms are caused by uncommon, slow-growing adenoid cystic carcinoma (ACC). [1] It was first described by Billroth in 1859 and he named it “cylindroma”. [2] It is also known as pseudo adenomatous basal cell carcinoma, basaloid mixed tumor, or basalioma (Krompecher, 1908) due to its analogous nature to the basal cell growths of the skin. In 1954, Ewing coined the term "adenoid cystic carcinoma. [3,4] It occurs in both major and minor salivary glands. [5] The most common site of ACC in the Head and Neck region is the posterolateral aspect of the hard palate. [1] Extra orally, the parotid gland (25%) is the single most common site of origin. The involvement of paranasal sinuses by the tumor is very rare, constituting only 0.3% to 1.0% of all sinonasal tumors. [6] The peak incidence is in the sixth and seventh decades of life with a wide range; there is a slight female preponderance. [7] The clinical signs and symptoms of ACC depend on the site involved. Pain is an important symptom of this tumor due to its tendency for perineural

spread. Perineural invasion of ACC is prevalent and is noted in almost half of the reported cases. Surgery remains the mainstay of treatment due to local recurrence. It is also known for distant metastasis. [8] Radiotherapy is instituted in the presence of positive resection margins, infiltrative growth patterns, or perineural invasion. [9] Histopathology frequently displays a mixture of solid, cribriform, and tubular forms. [3] And usually, the tumor is named according to the predominant pattern. [1,3] The objectives of our study are to report the varied presentations of six diagnosed cases with adenoid cystic carcinoma of the Head and Neck region and to discuss the clinical profile and management of these cases.

MATERIALS & METHODS

A retrospective study was held which consisted of six histopathologically proven and treated cases of adenoid cystic carcinoma of the Head and Neck region at our tertiary care center between the period of January 2009 and December 2022. All the required patient data were taken from the medical records section. The radiological and pathological images of the selected cases were collected from the Department of Radiology and Pathology respectively. Before the commencement of the study, written consent was taken from the patients/patient's attendant. The institutional ethical committee gave its approval to the project.

RESULT

CASE REPORT 1

A 46-year-old male patient, presented to the Department of Otorhinolaryngology with complaints of right-side progressive nasal obstruction and swelling over the right cheek for 6 months and watering of the right eye for 4 months. On clinical examination, a solitary swelling of around 3x3 cm was noted on the right side of the ala of the nose, non-tender, firm in consistency and the overlying skin was normal and pinchable (Figure 1 A and B). Anterior rhinoscopic examination showed deviated nasal septum to the left with a fleshy mass occupying the whole of the right nasal cavity which was sensitive on probing. Oral cavity examination showed a bulging over the right side of the hard palate. Vision and extraocular movements were normal. The diagnostic nasal endoscopic examination revealed a reddish friable mass arising from the middle meatus (Figure 2). A computed tomographic scan of the nose and paranasal sinuses showed a complete soft tissue opacification of the right maxillary sinus with bulging of walls of the maxillary sinus with erosion and destruction of the walls, anteriorly involving the soft tissue over the maxilla, bulging into the right infratemporal fossa pushing the septum (Figure 3) – suggestive of neoplastic etiology. A provisional diagnosis of malignancy of the right maxilla was made. Biopsy was taken from the mass in the right middle meatus which was reported as adenoid cystic carcinoma. Subsequently, he underwent prophylactic tracheostomy with right total maxillectomy and orbital exenteration with split skin grafting (Figures 4 A and B). The decision of orbital exenteration was made, based on the intraoperative findings. Intraoperatively, the anterior wall of the maxilla was eroded and induration of overlying subcutaneous tissue was noted with the superior extent of the tumor into the orbital floor and medial wall involving the periorbital fat until the orbital apex. The final histopathology was reported as adenoid cystic carcinoma: except for the posterior margin, all the margins were negative with the perineural invasion of the infraorbital nerve and pterygopalatine ganglion (Figure 5 A, B, and C). Post-operatively, the patient received radiotherapy, and rehabilitation was provided through the prosthesis and is on periodic follow-up with no recurrence to date.

CASE REPORT 2

A 68-year-old female patient, presented with complaints of nasal discharge from the

right nasal cavity and watering from the right eye for the past 3 months. She had been diagnosed previously with type 2 diabetes and hypertension and was on medications for the same. On anterior rhinoscopy, mucopurulent discharge was noted in the right nasal cavity. Clinical examination of the eye was normal. Diagnostic nasal endoscopy showed a polypoidal mass in between the septum and the middle turbinate on the right side of the nasal cavity (Figure 6). Contrast-enhanced MRI of the paranasal sinuses and the orbits, revealed mucosal thickening at the right maxillary, ethmoidal and sphenoid sinus. Heterogenous contrast enhancement of the right maxillary sinus and the middle turbinate was noted which was suggestive of fungal etiology (Figure 7). Considering her co-morbidities and the presentation during the surge of mucormycosis cases during the second wave of covid 19 pandemic, a provisional diagnosis of right rhino-orbital mucormycosis was made. The swab from the right nasal cavity for KOH mount was negative for fungal elements and the fungal culture was reported as Rhizopus species. Injection Amphotericin was empirically started. After initial stabilization, she was taken up for endoscopic sinus surgery with right medial maxillectomy (lateral rhinotomy approach) and posterior septectomy with clearance of disease under general anesthesia (Figure 8). Intra-operatively, the mass was seen arising from the right sphenoid sinus and the spheno-ethmoidal recess. The tissue from the right sphenoid sinus which was sent for frozen section during the procedure revealed the presence of glandular malignant cells. Subsequently, the injection of amphotericin was stopped because of adverse effects. Post-operatively, the patient recovered well. The final histopathology was reported as adenoid cystic carcinoma. The patient was on regular follow-up and free of disease for up to 1 year. Later she was shown to have a recurrence of the disease both radiologically and clinically. Following this she underwent revision surgical debridement of the lesion under general anesthesia.

CASE REPORT 3

A 45- year-old female patient, with symptoms of swelling in front of the left ear from the past year and severe pain over the swelling for 15 days presented to the Department of Otorhinolaryngology. On examination, a solitary swelling of 5x4 cm was noted in the left preauricular and infra-auricular region, with tenderness over the swelling and firm consistency. Clinically the facial nerve functions were intact. The fine needle aspiration cytology from the swelling was reported as pleomorphic adenoma. Subsequently, the patient underwent total parotidectomy under general anesthesia. Intra-operatively, the tumor was found adhering to the upper branches of the facial nerve, hence was sacrificed. Post-surgery the patient developed grade III facial palsy. The histopathology was reported as adenoid cystic carcinoma. Given the adherence of the tumor to the branches of the facial nerve, postoperatively she was advised for radiotherapy. However, the patient did not receive any further treatment and lost for follow-up. After 4 years, she came back with complaints of swelling at the upper part of the neck and in front of the left ear for the past 6 months. On examination, the swelling was noted in the infra and preauricular region and zygoma on the left side, and fixity to the underlying structures was noted. Contrast-enhanced MRI scan showed a heterogeneously enhancing lesion consisting of the parotid tissue on the left side measuring 43x32x71mm (Figure 9). Later she underwent revision surgery with left radical neck dissection (Figure 10). Postoperatively, the patient received radiotherapy and is on regular follow-up with no recurrence of the disease for 1 and a half years.

CASE REPORT 4

A 45-year-old female patient presented with swelling in front of the left ear for the past 3 months with no other accompanying

symptoms. On clinical examination, a swelling of ~ 5x4 cm was noted in the left pre-auricular region which was firm in consistency, non-tender, and mobile. The facial nerve functions were normal. Fine needle aspiration from the swelling was reported as a pleomorphic adenoma of the parotid gland. Following this, she underwent left superficial parotidectomy under general anesthesia. Intraoperatively, the tumor was found completely engulfing the facial nerve. However, an attempt was made to preserve the facial nerve. The final histopathology was reported as adenoid cystic carcinoma of the parotid gland. After a few days, the patient was taken up for revision surgery and a total parotidectomy was performed. The patient received postoperative radiotherapy with no evidence of recurrence during 2 years of follow-up, after which she was lost for follow-up.

CASE REPORT 5

A 36-year-old female patient, presented with swelling below the right pinna for 1 month and pain over the swelling for the past 20 days. Clinical examination showed a solitary swelling of 1.5x1cm in the right infra-auricular region with tenderness and firm consistency. Contrast-enhanced CT scan showed a well-defined lesion in the superficial part of the right parotid gland measuring 12.5x10.6mm. Fine needle aspiration from the swelling was reported as pleomorphic adenoma. Subsequently, she underwent a right superficial parotidectomy. The final histopathology was reported as adenoid cystic carcinoma with perineural invasion. Postoperatively, the patient received radiotherapy and is on regular follow-up.

CASE REPORT 6

A 75-year-old male patient presented with complaints of progressive difficulty in swallowing and foreign body sensation on the right side of the upper throat for 1 month. On examination, the oral cavity was within normal limits. A smooth globular swelling of around 2 x 2cm was noted towards the right side of the base of the tongue, non-tender, and firm in consistency was noted. Contrast-enhanced CT scan showed a well-defined soft tissue density lesion of 2.5 x 2.4 cm involving the base of the tongue on the right side, likely neoplastic etiology. The patient was taken up for an excisional biopsy under local anesthesia. Complete excision of the mass was achieved with cauterizing of the base of the tumor and sutures were applied. Postoperatively, he was stable and was allowed to take orally the same day. Later the histopathology was reported as adenoid cystic carcinoma. The patient was advised for postoperative radiotherapy and is on regular follow-up.

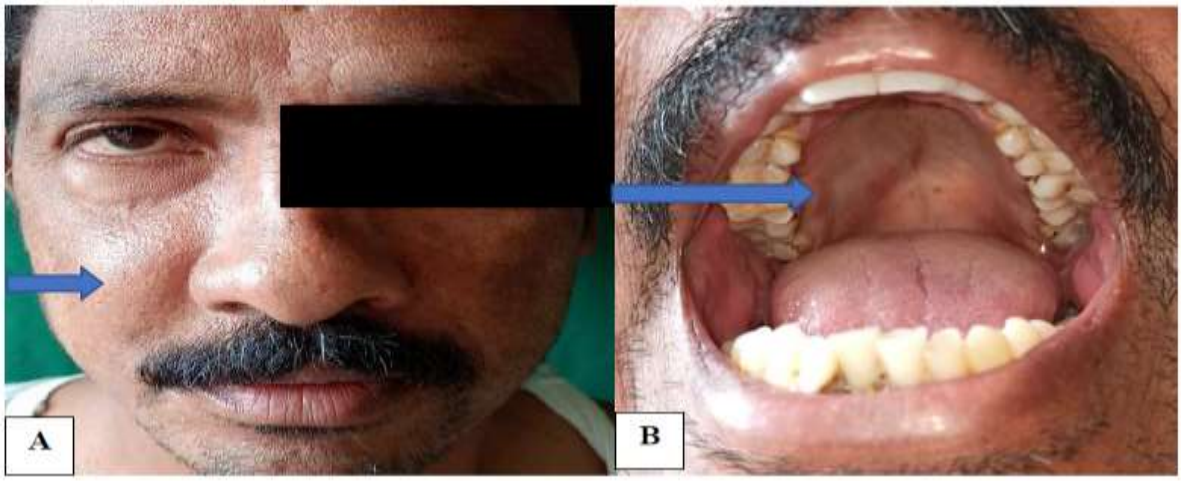

Figure 1 A) Diffuse swelling of ~ 3x3 cm + right side of the ala of the nose, firm, non-tender, skin over the swelling pinchable, esotropia of the right eye. B) Swelling over hard palate on the right side.

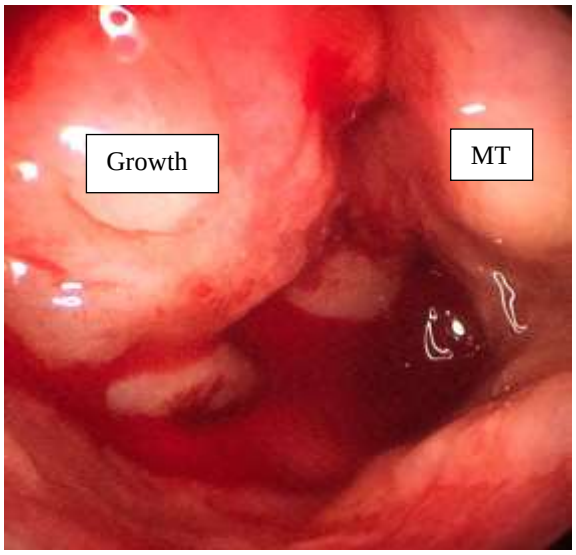

Figure 2 shows the diagnostic nasal endoscopic image showing reddish friable mass arising from the middle meatus.

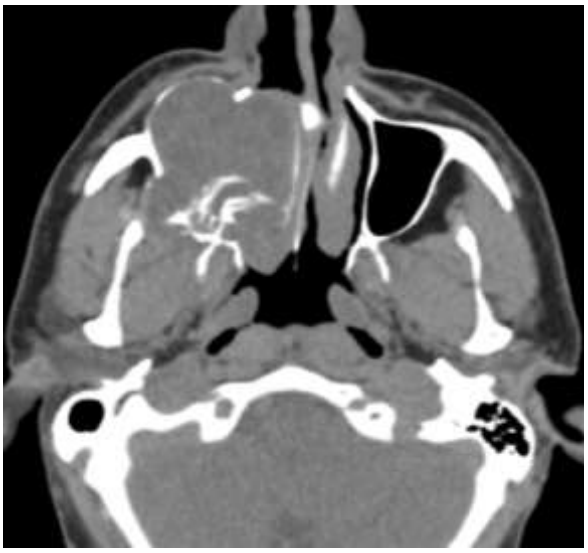

Figure 3 CT scan of nose and paranasal sinuses (Bone window) showing complete soft tissue opacification of right maxillary sinus. Bulging of walls of maxillary sinus with erosion and destruction of the walls, anteriorly involving the soft tissue over the maxilla, bulging into right infratemporal fossa pushing the septum.

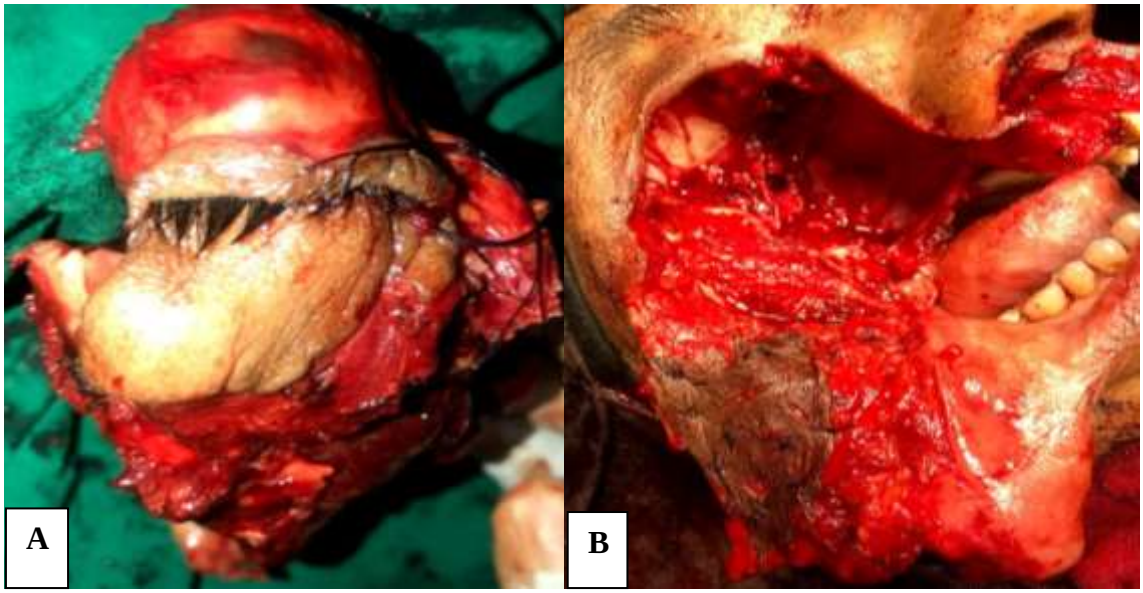

Figure 4 A) shows the operative specimen and B) shows the recipient site after right total maxillectomy and orbital exenteration along with split skin grafting.

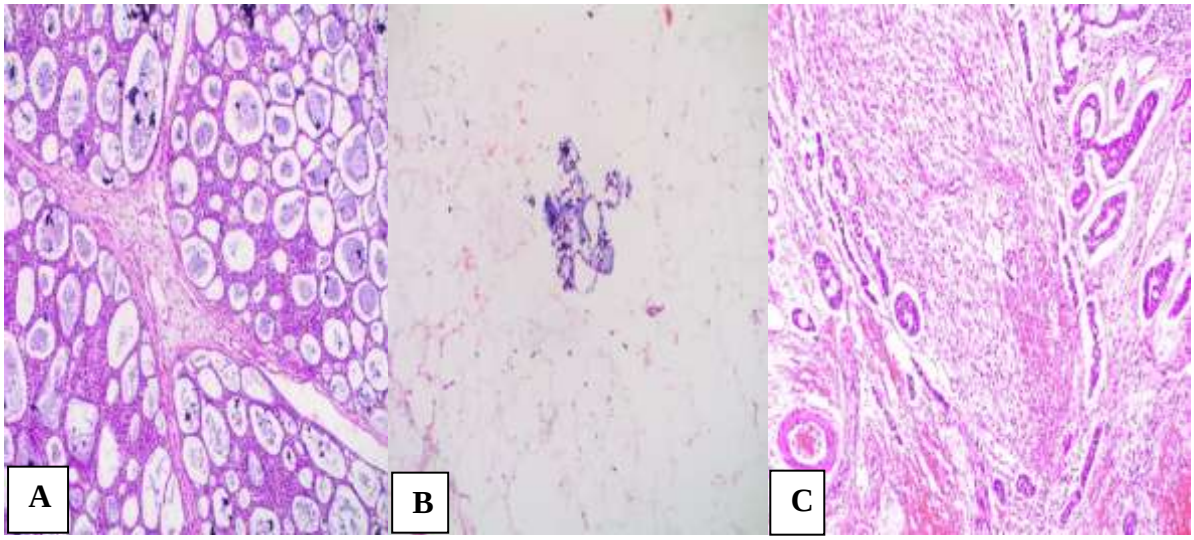

Figure 5 Histopathology images: A) 40X slides of adenocystic carcinoma- cribriform pattern B) Periorbital fat showing tumor infiltration C) 10X image showing perineural invasion of infraorbital nerve by tumor cells.

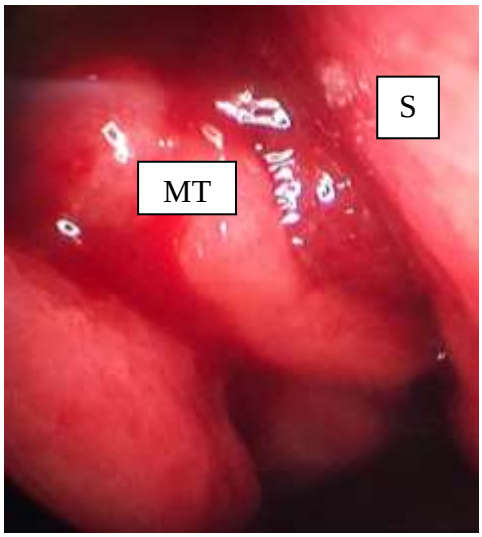

Figure 6 Diagnostic nasal endoscopic image showing polypoidal mass between the septum and the middle turbinate on the right side of the nasal cavity.

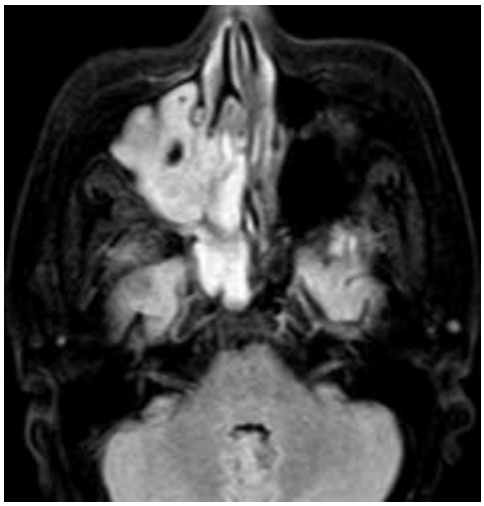

Figure 7 CEMRI PNS/ ORBITS- mucosal thickening noted at right maxillary sinus, ethmoid sinus and sphenoid sinus. Post contrast- right maxillary sinus and middle turbinate heterogenous contrast enhancement – suggestive of fungal etiology.

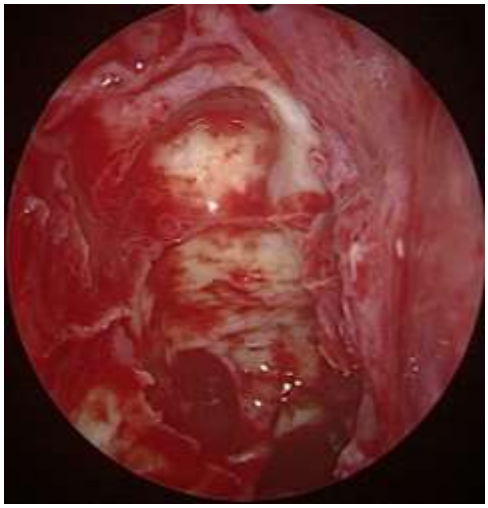

Figure 8 shows the intra operative endoscopic image after clearance of the disease from the right sphenoid sinus.

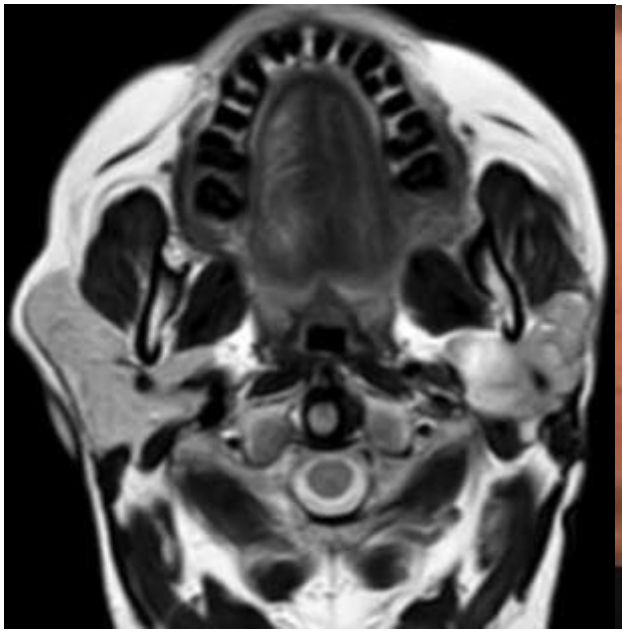

Figure 9 shows heterogeneously enhancing lesion involving superficial and deep lobe of left parotid gland measuring 43x32x71mm.

Figure 10 shows the post operative scar of the patients following revision surgery for recurrent disease.

DISCUSSION

A malignant condition affecting both the main and minor salivary glands, adenoid cystic carcinoma. It has an infrequent occurrence with less than 1 percent incidence among head and neck tumors. [1] It has aggressive behavior and a high incidence of both local recurrence and distant metastasis. The most common age of predilection is in the 4th to 6th decade with female preponderance. In our study, 4 patients were aged between the 4th to 6th decade and 2 patients were above that range. The mean age of occurrence was 52.5 years and 4 out of 6 patients (66.7%) were females which is consistent with other studies. [7]

Though ACC is the most common malignant tumor of the minor salivary glands with the palate being the most common location, [1,9] it can occur in other rare sites in the Head and Neck region like the nose, nasopharynx, paranasal sinuses,

middle ear, external auditory canal, maxilla, mandible, etc. [7] Three of our patients were diagnosed with ACC of the parotid gland, one patient with the maxilla, one patient with sphenoid sinus disease and one patient with the tumor over the base of the tongue. The symptoms usually depend on the site of origin. Pain and swelling are usually the presenting symptoms. Pain may be due to the involvement of peripheral nerves. The perineural invasion can be clinically evident with facial paralysis as in cases of parotid gland lesions or incidental findings during histopathological examination. In our study, no patients had clinical perineural involvement. The duration of the symptom depends on the site and slow-growing nature of the disease and also the patient factors like time of presentation to the hospital after the initial symptom. Because of the complex anatomy of the rare sites like the nasal cavity and paranasal sinuses, the disease may be concealed for an extended period without any presenting symptoms. [10]

Since it is a locally aggressive tumor, surgery remains the mainstay of treatment. Complete surgical resection of the tumor with clear margins is the goal. Surgery to remove the main tumor was performed on all our patients. Clinically negative margins were achieved in all the patients. However, a positive microscopic margin was present in only one patient. Paranasal sinus tumors are known to have positive surgical margins due to their proximity to vital structures like the orbit, dura, brain, and cranial nerves. In our case of ACC of the right maxilla, all the margins were negative except for the posterior. Perineural spread which is unpredictable makes clearance with negative surgical margins even more difficult. Microscopic perineural invasion was seen in 2 of our patients, that is, in the case of the right maxilla (perineural invasion of the infraorbital nerve and pterygopalatine ganglion was noted) and in the case of the right parotid gland.

ACC is considered to be a radiosensitive tumor but not a radio-curable tumor. Radiotherapy is to be instituted in all patients with advanced disease with positive margins and histopathologic evidence of perineural invasion. Post-operative radiotherapy delays rather than preventing local recurrence. [11]

The follow-up period was variable in our study. One of the patients with the left parotid gland ACC who was lost for follow-up came back with recurrence after 4 years of primary surgery. All other patients are on regular follow-ups to date. In addition, ACC in the Head and Neck region has a high recurrence rate with a tendency for distant metastasis. [12] In our study, two patients had recurrence one with the left parotid tumor who later underwent radical neck dissection along with revision surgery at the primary site, and another with the sphenoid sinus tumor underwent surgical debridement. Other patients were free of disease with no evidence of recurrence till now. Among our patients, no one had distant metastasis.

The 5- year survival rate is 75 %, and the 10-year survival rate is only 20% (because of perineural invasion). [4,7] It is reported that compared to major salivary glands ACC of minor salivary glands tends to have poorer prognosis. [13] Even though ACC has a bad prognosis, all our reported patients had a good prognosis.

CONCLUSION

Adenoid cystic carcinoma is a slow-growing aggressive tumor with a high chance of locoregional recurrence. Primary treatment is surgery in combination with radiotherapy. ACC of minor salivary glands has a worse prognosis than those of major salivary glands. It frequently develops delayed distant metastasis occurring as late as 10-20 years and needs long-term follow-up. Postoperative radiation is a must to delay rather than prevent local recurrence.

Declaration by Authors

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Conflict of Interest: The authors declare no conflict of interest.

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