



Adenoid Cystic Carcinoma of the Nasopharynx—a Rare Entity: Our Institutional Experience and Therapeutic Approach

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Abstract

Adenoid cystic carcinoma of the nasopharynx is a rare, slow growing, and locally aggressive neoplasm. Three cases presented with recurrent epistaxis. Endoscopy-guided biopsy proved the diagnosis of adenoid cystic carcinoma. The location and the extent of the tumor were confirmed on imaging. Surgery followed by radiation therapy was the treatment modality used. All three cases showed good clinical response. The aim is to discuss the surgical approach and review of literature concerning this malignancy.

Keywords Adenoid cystic carcinoma · Nasopharynx · Maxillary swing approach · Radiation therapy · Cribriform pattern

Introduction

Nasopharyngeal adenoid cystic carcinoma is a rare neoplasm, accounting for approximately 0.5% to 4% of all the malignancies of the nasopharynx [1]. Adenoid cystic carcinoma (ACC) has propensity for aggressive local infiltration and perineural spread. They usually extend along the cranial nerve canal, towards the orbital cavity and skull base, making all surgical approaches hard and delicate [2].

ACC seldom metastasizes to regional lymph nodes but can metastasize to distant sites with the lungs being the most com-

mon location [1]. Surgery is the primary treatment for ACC followed by radiation therapy.

Materials and Methods

We report three cases of nasopharyngeal ACC treated with surgery and adjuvant radiotherapy. The neuro-navigation system was used during intracranial resection of the tumor in one case. Due to the retrospective nature of this study, it was granted an exemption in writing by the IRB of Rajiv Gandhi

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Cancer Institute, New Delhi. The radiological image and pathological figure mentioned in the study have been approved by a research ethics committee.

Case 1

A 28-year-old female presented to outpatient department with recurrent epistaxis for 3 weeks. Diagnostic nasal endoscopy (DNE) showed a smooth submucosal bulge of size 2×2 cm in the left nasopharyngeal wall. Biopsy was suggestive of adenoid cystic carcinoma.

A lobulated dumbbell-shaped smooth submucosal mass was noted in the postero-superior part of the nasopharynx with focal dehiscence of the floor of the sphenoid sinus, on magnetic resonance imaging (MRI) of paranasal sinuses (PNS).

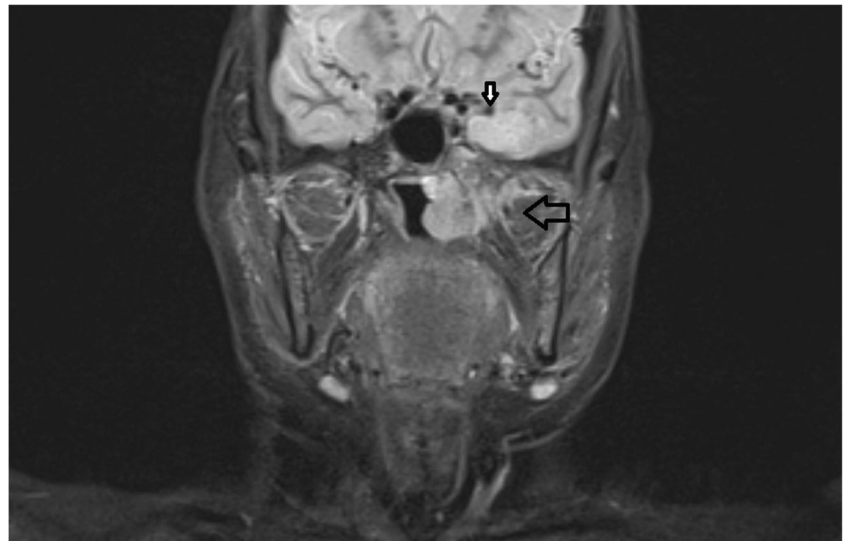
Robot-assisted nasopharyngectomy was performed and the tumor was excised en bloc with negative margins. Histopathological report (HPR) showed features of adenoid cystic carcinoma, predominantly cribriform and focal tubular pattern. She then received adjuvant radiation therapy.

Case 2

A 67-year-old female presented to our institute with history of recurrent episodes of nasal bleeding for 1 month. DNE showed smooth brownish polypoidal mass in the left nasopharynx arising from the fossa of Rosenmuller protruding to the left posterior choanae. Biopsy showed low-grade carcinoma or myoepithelial tumor.

MRI showed $3.1 \times 2.1 \times 3.5$ cm sized mass, involving the left wall and the adjoining roof of the nasopharynx, the left fossa of Rosenmuller, and extending superiorly to involve the left pterygoid plate. The mass was also eroding the floor of the middle cranial fossa with extradural extension towards the anterior temporal lobe (Fig. 1).

Fig. 1 MRI findings suggestive of $3.1 \times 2.1 \times 3.5$ cm sized mass involving the left wall and the adjoining roof of the nasopharynx extending to involve the fossa of Rosenmuller. Laterally, the mass was extending to involve the medial pterygoid muscle (bold arrow), superiorly to the left pterygoid plate, eroding the floor of the middle cranial fossa with extradural extension, and also bulging into the left anterior temporal lobe (white arrow)



The surgery was performed utilizing Weber-Fergusson incision with upper lip split. The maxillary swing approach was combined with left temporal craniotomy using the neuro-navigation system and total excision of tumor was done with negative margins. There were no postoperative complications. HPR was suggestive of adenoid cystic carcinoma with predominant cribriform pattern. She then received postoperative radiation therapy.

Case 3

Another 60-year-old female presented with recurrent epistaxis for 1 month. On examination, a smooth bulge was appreciated over the right side soft palate of size 2×2 cm and no palpable nodes in the neck.

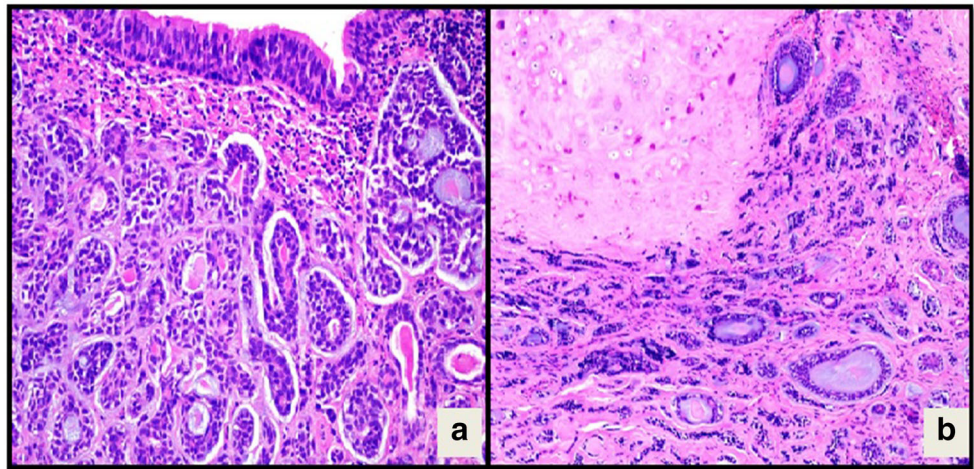
CT scan showed soft tissue mass in the right lateral wall of the nasopharynx of size 3.1×3.3 cm in the region of the Eustachian tube and fossa of Rosenmuller. DNE and biopsy were suggestive of adenoid cystic carcinoma.

En bloc removal of the tumor was done via maxillary swing approach. HPR showed features of adenoid cystic carcinoma with cribriform pattern without perineural invasion (Fig. 2). She was thereafter referred for postoperative radiation therapy.

Results

All three received postoperative radiotherapy. There were no major complications. Cases 2 and 3 developed epiphora postsurgery. A small palatal perforation occurred in Case 3, post radiotherapy for which she was advised obturator plate. We have been following case 1 for 4 years, case 2 for 15 months, and case 3 for 8 months. There is no clinical and radiological evidence of disease in all 3 cases.

Fig. 2 **a** Pseudostratified ciliated columnar epithelial lined tissue with a submucosal adenoid cystic carcinoma arranged in tubules and cribriform pattern with areas showing invasion of tumor into the cartilaginous tissue (**b**)



Discussion

Adenoid cystic carcinomas most frequently arise from major salivary glands, minor salivary glands (usually hard palate), followed by the paranasal sinuses and rarely in the nasopharyngeal area [1].

ACC of the nasopharynx is a slow growing tumor with a long natural history, which is responsible for the delay in diagnosis and management. The interval between onset of the disease and onset of first symptom is estimated to be between 2 and 5 years [1].

Most common symptoms are epistaxis and progressive nasal obstruction. Cranial nerve III, IV, V, and VI involvement leads to ophthalmoplegia, facial pain, and numbness. The pathways of local spread via the Eustachian tube into the temporal bone and cerebellopontine angle whereas via cranial nerves into the cavernous sinus are signs of poor prognosis. [2]

There is no gender predilection but peak incidence occurs predominantly among women, between the 5th and 6th decades of life [3]. Arsenic compounds, nickel, and oak wood dust may be possible etiological factors.

Histologically, three growth patterns can be recognized for ACC: cribriform, tubular, and solid [4]. The cribriform variant is the most common whereas the solid variant is the least common histopathologic subtype. The solid pattern has the worst prognosis and is also associated with the highest incidence of distant metastasis and perineural invasion.

Imaging of ACC is based on computed tomography (CT) scan, particularly helpful in detecting bony erosions of the skull base, and on magnetic resonance imaging (MRI) with gadolinium, effective in demonstrating possible involvement of the cavernous sinus, and perineural or perivascular infiltration [2, 5].

Surgery followed by radiation therapy is the most common treatment modality employed [2]. Owing to the complex

anatomical location of the nasopharynx, getting negative margins with surgery is quite difficult. The maxillary swing approach is quite useful in this regard. One of our cases was a young female, who wished to avoid any external scar. Thus, robot-assisted nasopharyngeal approach was then utilized and we were able to achieve negative margins.

Even after good local control, the presence of distant metastases is possible in 39% of the patients [2]. Regional failure may be due to positive margins, tumor seeding, lymphatic or hematogenous spread, or perineural spread.

Rarely radiation therapy has been used as sole treatment of ACC [5]. Radiation may also provide symptomatic relief from pain, ulceration, bleeding, and pharyngeal obstruction for palliation.

Chemotherapy use for ACC is controversial as it has shown limited and poorly defined role. None of our cases received chemotherapy.

Although the 5-year survival rate is high, the local recurrence rate is equally high [2, 3]. However, prognosis of ACC of the nasopharynx remains favorable.

Conclusion

ACC of the nasopharynx is a rare tumor entity. Owing to the paucity of cases, information regarding appropriate therapy for this malignancy is limited in literature. MRI is the most important tool to examine the local extension of mass. Surgery followed by radiotherapy is the primary treatment in the current era.

Declarations

Conflict of Interest The authors declare no competing interests.

References

1. Lee DJ et al (1985) Adenoid cystic carcinoma of the nasopharynx. Case reports and literature review. *Ann Otol Rhinol Laryngol* 94: 269–272
2. Gormley WB, Sekhar LN, Wright DC, Olding M, Janecka IP, Snyderman CH, Richardson R (1996) Management and long-term outcome of adenoid cystic carcinoma with intracranial extension: a neurosurgical perspective. *Neurosurgery* 38:1105–1112
3. Ellington CL, Goodman M, Kono SA, Grist W, Wadsworth T, Chen AY, Owonikoko T, Ramalingam S, Shin DM, Khuri FR, Beitler JJ, Saba NF (2012) Adenoid cystic carcinoma of the head and neck: incidence and survival trends based on 1973–2007 Surveillance, Epidemiology, and End Results data. *Cancer* 118:4444–4451
4. Batsakis JG, Luna MA, el-Naggar A (1990) Histopathologic grading of salivary gland neoplasms: III. Adenoid cystic carcinomas. *Ann Otol Rhinol Laryngol* 99:1007–1009
5. Vikram B, Strong EW, Shah JP, Spiro RH (1984) Radiation therapy in adenoid-cystic carcinoma. *Int J Radiat Oncol Biol Phys* 10:221–223

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