An Adenoid Cystic Carcinoma of Nasal Septum Origin

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Abstract

Background: Adenoid cystic carcinoma (ACC) of the nasal septum is a rare disease of the head and neck. This elusive cancer is despite its locoregional control improvement over the years may cause distant metastasis years after definitive treatment. Reliable prognostic factors were identified over the years and their implications are immense especially with regards counseling and therapeutic planning.

Case Report: We present a case of nasal septum ACC with perineural invasion. A 54 years old male presented with recurrent mild epistaxis for two months duration with associated right nasal obstruction. Endoscopic excision of the tumor was performed. The patient received adjuvant radiotherapy due to involved resection margins and perineural invasion.

Conclusion: Nasal septal ACC is a rare entity. Adjuvant radiotherapy is advocated as a part of treatment due to its characteristic perineural spread. Lifelong follow up is necessary due to its high recurrence rate.

Keywords: Adenoid Cystic Carcinoma; Septum; Epistaxis

Introduction

Adenoid cystic carcinoma (ACC) was first described as cylindroma by Bilroth in 1856 [1]. ACC is a rare form of malignancy representing about 5% of all head and neck carcinomas [2]. Sino nasal ACC accounts for 10-20% of all head and neck ACC [3]. The most common site of the disease is the maxillary sinus followed by the nasal cavity, nasopharynx and ethmoid sinus [4]. It has a tendency to progress slowly with a characteristic perineural invasion. We present a rare case of nasal septum ACC.

Case Report

A 54 years old male presented with recurrent mild epistaxis for two months duration with associated right nasal obstruction. He is a smoker with a background history of hypertension, benign prostatic hyperplasia and prolapsed intervertebral disc at the level between the fourth and fifth lumbar vertebrae. The patient had no family history of malignancy.

Naso-endoscopic examination revealed a friable mass originating from the posterior part of the right nasal septum extending to the floor of the nose. The ostiomeatal complex, sphenoid-ethmoidal recess and nasopharynx were normal. There were no palpable cervical lymph nodes. Per oral and otoscopic examinations were unremarkable.

Discussion

Adenoid cystic carcinoma (ACC) is a rare malignant neoplasm representing about 5% of all head and neck carcinomas [2]. Sinonasal ACC can affect any sites within the nasal cavity. The commonest site of sinonasal ACC was the ethmoid sinus [4]. However, other studies found that the most common sinonasal ACC was in the maxillary sinus [3]. In this case, the presence of ACC arising from the septum made it more interesting. Six reports of ACC of the nasal septum had been available in the literature rendering the rarity of the disease [5].

Histologically, ACC is classified into the commonest cribriform type, tubular type and the least common solid type. The classification correlates with the disease prognosis. The cribriform type reported to have the best outcome and the solid type had the worst prognosis [6, 7]. In addition, the staging of the tumour, involvement of perineural invasion and margin of resection also play an important role in the
prognosis of the disease. T3 and T4 tumours are associated with much higher incidence of perineural invasion compared to T1 and T2 tumours (60% vs. 23.5%) [8].

Perineural invasion maybe demonstrated on MRI scan as fat planes obliteration, enlargement or destruction of foramina, enlargement or enhancement of nerves, neuropathic atrophy and convexity of the lateral sinus wall and replacement of the trigeminal subarachnoid cistern with soft tissue [9]. However, the imaging studies in our patient revealed the mass confined to the nasal cavity without perineural invasion. Endoscopic sinus surgery and wide local excision of the mass on the posterior part of the septum was performed.

The nasal septum cartilage in our case was not involved. Should the nasal septum cartilage be involved, there are several approaches to gain margin clearance depending on the localization of the lesion on the septum. If the anterior part of the septum is involved, lateral rhinotomy would be the option of choice. Mid-facial degloving approach can be utilized for lower nasal septum involvement with good cosmesis. If the posterior nasal septal area is involved, adequate exposure is allowed by using lateral rhinotomy with sublabial incision. To resect tumor of the nasal septum and floor of the nose, lateral rhinotomy with lip-splitting incision is the choice of approach [5].

Due to the neurotropism nature of the ACC, current treatment recommendation is complete surgical resection with post-operative radiotherapy (RT) [10]. This was supported by the findings by a study by Garden et al demonstrating local control rates of 95%, 86% and 79% at 5, 10, and 15 years in 198 patients with ACC of the head and neck treated with surgery and then followed by RT [11]. Total removal of the tumour with negative tissue margin is vital to a successful combined therapy. Adjuvant RT was indicated in our patient in view of positive resection margin and the presence of perineural invasion on histopathological examination.

Recurrence rate of ACC proved to be a notorious. It typically recurs locally. Recurrence rate is reported to be as high as 65% in sinonasal ACC despite surgical excision followed by RT [12]. Combined treatment for ACC of head and neck origin achieved 85% freedom from relapse and disease-specific survival at 5, 10, and 15 years was 89%, 67.4% and 39.6% respectively. Distant failure occurs predominantly within 5 years but it was registered even after 15 years [8].

Our patient whom unfortunately had T3 tumour, perineural invasion and a positive margin may have an unfavorable prognosis. He predictably developed distant failure 15 months after the initial treatment.

Conclusion

Nasal septal ACC is a rare entity. Adjuvant radiotherapy is advocated as a part of treatment due to its characteristic perineural spread. Lifelong follow up is necessary due to its high recurrence rate.

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