

CASE REPORT

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Nasal septum and sphenoid sinus located adenoid cystic carcinoma

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Abstract

Adenoid cystic carcinoma (ACC) is a rare tumor comprising 1% of malignant tumors of head and neck. The most common localization for ACC is minor salivary glands and involvement of sphenoid sinus is very rare. It grows slowly and has a high tendency of local recurrence, and perineural invasion. Surgery and postoperative radiotherapy is considered as an optimal treatment. We present a case of 50-year-old female with complaint of nasal obstruction. The computed tomography imaging showed the presence of nasal mass arising from the posterior part of the nasal septum and the adjoining sphenoid sinus anterior wall. The mass was excised and the pathologic specimens revealed ACC with perineural invasion, positive surgical margin and bone destruction. The patient received postoperative radiotherapy. Improved treatment outcomes led the investigators to recommend the postoperative radiation as the routine treatment approach for most patients with the ACC of the head and neck to improve local control rates.

Keywords: Adenoid cystic carcinoma, sphenoid sinus, radiotherapy

Introduction

Adenoid cystic carcinoma (ACC) is a rare tumor of epithelial cell origin, comprising 1% of malignant tumors of head and neck and 10% of all salivary gland neoplasms [1]. The most common localization for ACC is minor salivary glands (30%). ACC of sino-nasal tract is a rare tumour constituting 10% of malignancies in this region. Sphenoid sinus ACC is very rare lesions. Patients of ACC of sino-nasal tract, sphenoid sinus constituted 3% of cases [2,3]. Treatment of ACC pose challenges due to its biological behavior of slow growth, high tendency of local recurrence, and distant metastases [4]. Surgery is the mainstay treatment but its high propensity to spread locally through bony destruction and perineural and perivascular invasion make difficult to achieve clear surgical margins [1]. To this respect radiotherapy was considered as an adjuvant treatment option after surgery to improve local control.

We present a rare case of ACC of sphenoid sinus treated with adjuvant radiotherapy after surgical treatment in current literatur light.

Case

A 50-year-old female presented to the department of otolaryngology and head and neck surgery with a history of progressively increasing nasal obstruction since 6 months. The nasal obstruction was due to the mass

originating from the nasal cavity and expanding to sphenoid sinus anterior wall bilaterally (Figure 1). There was no history of nasal discharge, postnasal drip, allergy, epistaxis, headache, facial fullness, alteration in smell, or other nasal complaints. The non-contrast computed tomography (CT) of head confirmed the presence of nasal mass arising from the posterior part of the nasal septum and the adjoining sphenoid sinus anterior wall which was obstructing the bilateral nasal cavity. Aeration of the associated sinuses were clear. Prior to surgical intervention an informed consent was taken. The mass was excised under general anaesthesia using endoscopic guidance. The excision was started with the posterior of the nasal septum and extending to the anterior wall of the sphenoid sinus. Excision margins enlarged till torus tubarius bilaterally (Figure 2).

The pathologic specimens were evaluated in the pathology department and reported as adenoid cystic carcinoma. Macroscopically biopsy spesimen was totally 3x3x1 cm consisting of irregular-looking pieces in different sizes. In microscopic examination tubules infiltrating hyalinized stroma were observed and the cribriform pattern was seen in some parts (Figure 3). The spaces contain a mildly basophilic mucoid basal lamina material. Futhermore perineurial and bone invasion was determined (Figure 4). Solid component was absent. Surgical margins were infiltrated with tumor. Immunohistochemically the tumor cells was positively stained with pancytokeratin, c-kit, bcl-2, S-100. In addition the myoepithelial cells were stained with p63 and actin positively.

The patient was referred for radiotherapy. Radiation was delivered 5 days a week, once a day to a total dose of 66

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Gy in 33 fractions with simultaneous integrated boost technique by intensity modulated radiotherapy (IMRT).

The patient finished the treatment without complications and there is no recurrence during one year follow-up

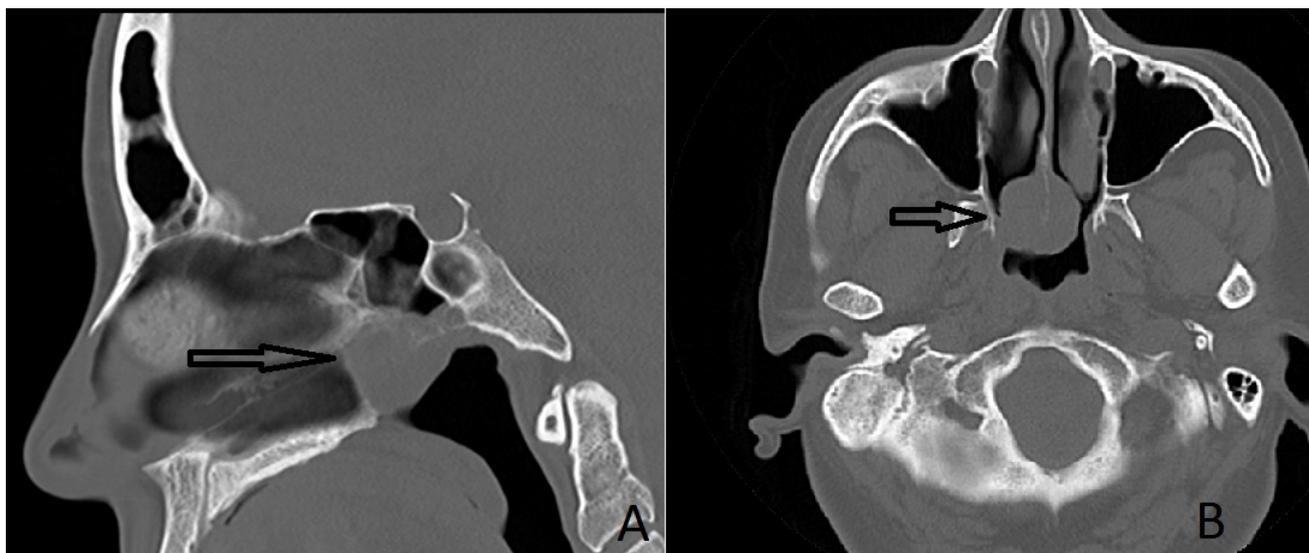


Figure 1. Computer tomography imaging of lesion (A: saggital image, B: Axial image)

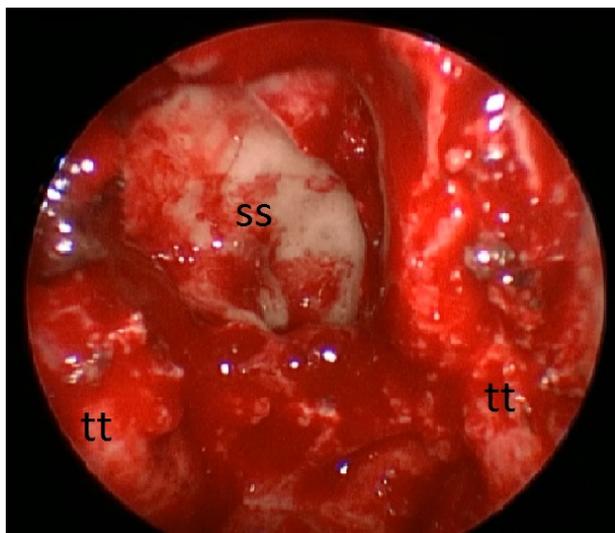


Figure 2. This figure show that after lesion excision (ss: sphenoid sinus, tt: torus tubarius)

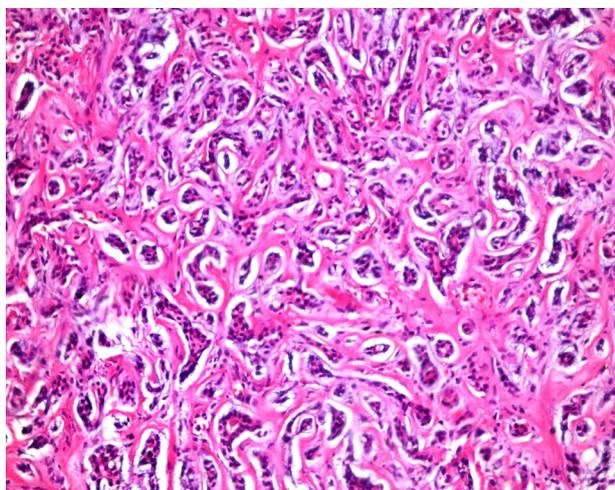


Figure 4. Tubular structures with inner layer of duct-lining cells and an outer layer of clear cells (HE, x200).

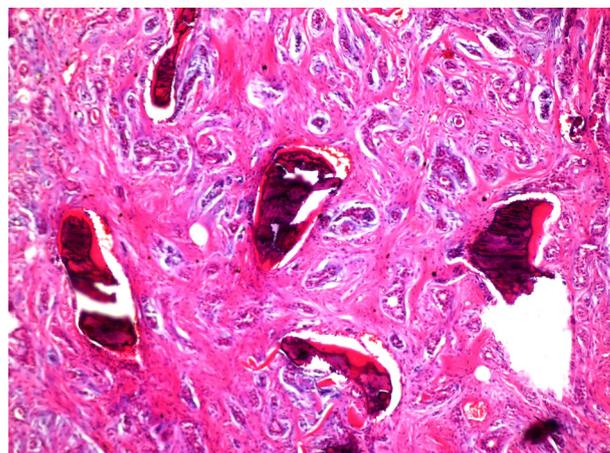


Figure 5. Tumor infiltrating bone trabecules (HE, x100)

Discussion

Neoplasms of the paranasal sinuses account for 0.2 to 0.8% of all cancers and for 2 to 3% of head and neck cancers. Primary involvement of the sphenoidal sinus has been reported to occur in only 1 to 2% of all paranasal sinus tumors. The most common malignant tumors were squamous cell carcinoma and ACC rarely seen in this region. ACC is a relatively rare tumor of epithelial cell origin, most commonly arising from major or minor salivary glands, and comprises 3 to 5% of all head and neck malignancies [5]. It usually presents as a slowly growing, firm mass. Presence of lymphadenopathy is uncommon since ACCs do not usually spread to regional lymph nodes. Unlike ACC of the salivary glands, ACC can rarely occur in other areas of the head and neck region, and present as nasal congestion or obstruction as in our case [6].

ACC of the head and neck, and specifically of the nasal cavity and paranasal sinuses, has many treatment challenges for several reasons as it has a high propensity

for local invasion to adjacent structures, making resection more difficult [5]. Treatment approach for ACC with adverse parameters such as advanced tumor lesions, positive surgical margin and perineural invasion appears to be best achieved with combined surgical resection and radiotherapy [7-9] This was corroborated by Zhang et al.'s study of 88 patients with ACC in the nasal cavity and paranasal sinuses looking at 5-year and 10-year survival rates. The 5-year and 10-year survival rates of 76% and 41% were obtained in patients who received surgery combined with radiotherapy. The 5-year and 10-year survival rates were 75% and 37% for those treated by surgery alone and 29% and 14% for those treated by radiotherapy alone [10]. Another study by Liu et al. of 42 patients with ACC of the nasal cavity concluded that surgery combined with high-dose postoperative radiation improves the local control and survival in patients with positive margins, no sufficient margins, or advanced disease [11]. Contrary to these reports, other studies by Ellington et al. have reported unclear benefit of adjuvant radiotherapy and stated the mainstay treatment as surgical resection [12].

Chemotherapy has unclear benefit in ACC and these tumours are considered as chemo-resistant and there is no accepted standard systemic chemotherapy for patients with ACC [13].

In our patient, due to the unfavorable prognostic factors such as presence of perineural invasion and bone destruction we added radiotherapy after surgery to improve local control rates.

In conclusion, sinonasal tract ACC is rare lesions and it may directly arise from the sphenoid sinus, present as a large mass and affect the nasal airway. For these lesions surgery is mainstay treatment and the postoperative radiation is considered as the routine treatment approach for improved treatment outcomes.

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