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Adenoid Cystic Carcinoma Arising in the Sphenoid Sinus Presenting with Headache and Diplopia: A Case Report

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ABSTRACT

Introduction: Adenoid cystic carcinoma (ACC) is a rare, slow-growing malignant tumor, mostly occurs in the minor salivary glands. Moreover, ACC arising in the sphenoid sinus is a rare entity.

Case: Herein, we report a case of ACC in a 69-year-old man who presented with a headache and diplopia. Both computed tomography (CT) and magnetic resonance imaging (MRI) demonstrated a mass lesion in left sphenoid sinus extending to the sellar area and clivus. After resection of the tumor, histopathological examination and immunohistochemical analysis reported "adenoid cystic carcinoma".

Conclusion: We introduced a case of ACC that presented with a headache and diplopia. Although rare, ACC can arise from sphenoid sinus and should be considered as differential diagnosis of sphenoid lesions.

Key words: Adenoid cystic carcinoma, Head and neck, Sphenoid sinus

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INTRODUCTION

Adenoid cystic carcinoma (ACC) is a slowly growing, locally invasive tumor of salivary glands, with high tendency for perineural spread and bony invasion. According to previous reports, ACC accounts for only 1% of all head and neck malignancies [1,2]. In a meta-analysis, sphenoid sinus was involved in only 3% of cases with ACC of sino-nasal tract [3]. Clinical presentation is dependent to the location of tumor that can be in salivary gland, nasal cavity and paranasal sinus. ACC generally presents as an asymptomatic mass. However, numbness, paresthesia, or pain may exist due to high tendency of tumor for neural involvement [4]. Immunohistochemical analysis can be used to differentiate ACC from other salivary gland tumors [5,6]. ACC cells have a positive reaction pattern for Vimentin, pan-cytokeratin, C-kit, p53, Ki67, and also alpha smooth muscle actin in comparison with neoplastic cells in BSSC that have a negative reactive pattern for smooth muscle actin [3-5]. ACC cells reveal compartment staining pattern for p63 and also a strongly positive reactive pattern for Vimentin compared with BSCC. To the best of our knowledge, only a few cases of ACCs arising in the sphenoid sinus have been reported in the literature. Here, we describe a case of ACC of the left sphenoid sinus in which definite diagnosis was achieved by imaging including CT AND MRI with contrast and histopathological examination.

CASE PRESENTATION

A 69-year-old non-smoker male patient presented to the otolaryngology clinic with a history of intermittent right frontal headache and double vision since 3 months. He denied nasal discharge, epistaxis, postnasal drip, allergy, alteration in smell, or other nasal complaints. There was no history of trauma or other illnesses. The past medical history was unremarkable except for a diabetes mellitus and percutaneous coronary intervention (PCI) due to ischemic heart disease.

On ocular examination, a marked right abduction deficit was noted. Both pupils were reactive. Cranial nerve examination revealed a sensory loss in the region of trigeminal nerve on the right side of face. Other cranial nerves were clinically spared. No clinically appreciable lymphadenopathy was present in the head and neck region. On anterior rhinoscopy, he was found to have a septal deviation to the left side. The CT of paranasal

sinuses demonstrated a mass lesion in right sphenoid sinus (Figure 1).

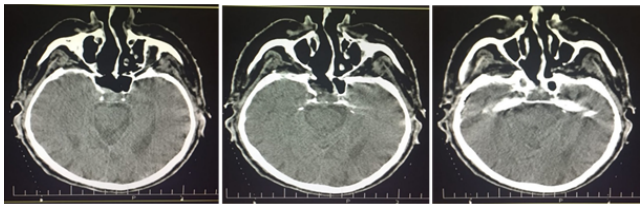

Figure 1: (A) Axial sections of CT show a mass lesion in left sphenoid sinus; (B,C)In upper sections filling of left sphenoid sinus with soft tissue density is seen

Owing to sensory loss in the region of trigeminal nerve on the right side of face, magnetic resonance imaging (MRI) was requested. A contrast-enhanced MRI of the brain revealed a mass in the right sphenoid sinus extending to the sella and clivus. The mass was low-signal in the T1-weighted image and high signal in the T2-weighted image with significant contrast enhancement (Figure 2).

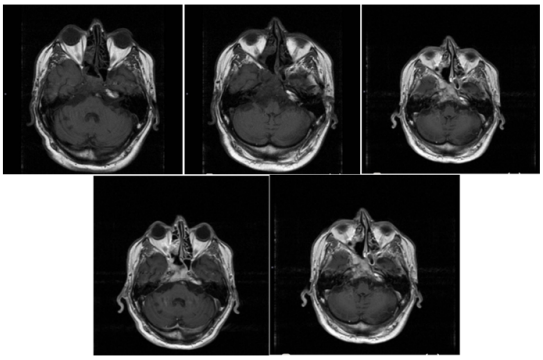

Figure 2: (A,B) MRI,T1 reveals a LOW signal mass in the right sphenoid sinus extending to the sella and clivus; (C,D) The mass was HIGH-signal in the T2-weighted image; (E) The mass has significant contrast enhancement post contrast image

Functional endoscopic sinus surgery (FESS) was planned for achieving tissue diagnosis. An informed consent was taken from the patient. Under general anaesthesia, the middle turbinate was resected partially. On endoscopy, the tumor was brown rubbery in colour, soft in consistency arising from sphenoid sinus. Frozen section specimen was sent to the laboratory which was compatible with ACC/chordoma (Figure 3).

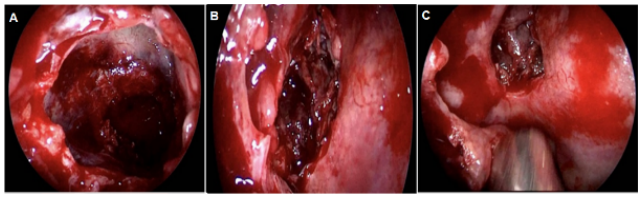

Figure 3: Intra-operative view of the lesion located in the sphenoid sinus, (A ,B)Brown rubbery tumor in sphenoid sinus with ulcerated appearance, (C) Intraoperative image of sphenoid sinus after tumor resection

The permanent pathologic specimens were evaluated in the pathology department. Macroscopically, biopsy specimen was totally 4 × 2 × 2 cm consisting of multiple fragments of brown rubbery tissue.

Histopathological examination of Hematoxylin and Eosin (H & E) stained glass slides revealed a tumoral lesion composed of nest-like structure, columnar cells around the pseudocyst space filled with homogeneous eosinophilic material. Myxoid stroma was present. In addition, collections of mucus were existed that infiltrated with neoplastic cells (Figure 4).

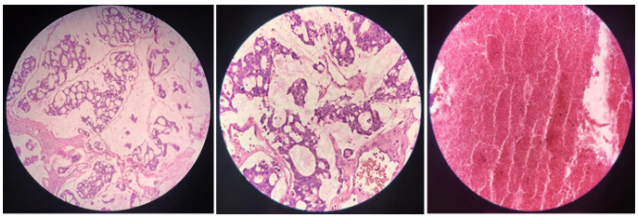

Figure 4: Histopathological examination of nest-like structure, columnar cells around the pseudocyst space filled with homogeneous eosinophilic material

Based on the findings of imaging and histopathology, "adenoid cystic carcinoma" was made as a definite diagnosis.

Abdominal and chest CT and bone scan were done for assessment of metastasis. In our case, the mass was metastatic to the liver, bone and base of skull.

After completion of first course of treatment patient was discharged. Two month later, he was admitted to the hospital because of fever and uremia. Unfortunately, he died following respiratory distress and tachycardia.

DISCUSSION

ACCs occur with equal frequency in male and female patients, especially in fifth to seventh decades of life [7]. Adenoid cystic carcinoma (ACC) has an indolent course and high tendency for perineural spread and bony invasion. According to previous reports, ACC accounts for only 1% of all head and neck malignancies [2,7,8]. Majority of ACCs arise in the salivary glands, with the rest occurring in other sites including the orbit, external auditory canal, lacrimal gland, and rarely in paranasal sinus etc. [8,9]. To the best of our knowledge, only a few cases of ACCs arising in the sphenoid sinus have been reported in the literature. In a meta-analysis conducted on 520 patients suffering from ACCs of sino-nasal tract, sphenoid sinus was involved in only 3% of cases [3].

On microscopic examination, perineural invasion is seen in over 50% of ACCs in head and neck region which it most commonly involves the branches of 5^{th} and 7^{th} cranial nerves [10]. In our patient, 5^{th} and 6^{th} cranial nerves were involved clinically. He presented with headache, diplopia and sensory loss in the region of trigeminal nerve on the right side of face.

Although both CT and MRI have been introduced for diagnosis of perineural spread (PNS), MRI is the modality of choice. The sensitivity and specificity of MRI is higher than CT in detecting ACC of paranasal sinuses with PNS [11].

Currently, two different histopathological systems are used for grading of ACC. In the Perzin/Szanto system, ACC is classified into 3 grades- grade 1: predominantly tubular, no solid component; grade 2: predominantly cribriform, <30% solid; grade 3: Solid component>30%. In the Spiro system, the presence of more than 50% of solid components is considered high grade [12].

Hematoxylin and eosin (H & E) stain shows biphasic cells with cribri forming and basement membrane-like material. Immunohistochemical (IHC) analysis reports pan-cytokeratin, S100 and p63 positive in basal cells and C-kit, CK7 positive in ductal cells [13].

Common sites of distant metastases include lungs and bone [14]. In addition, ACC has a tendency to spread to adjacent sites like the sellar and parasellar areas [15]. In our case, the tumor was extended to the sellar area and clivus.

Although radical surgical resection with free surgical margins followed by radiotherapy is the treatment of choice for ACC [16], other therapeutic approaches such as endoscopic technique have also been described in the literature [15,17].

Giridhar et al. reported a case of ACC of sphenoid sinus who treated by radical radiotherapy [9]. Our patient underwent palliative chemoradiotherapy.

The factors affecting the survival of cases with ACC of head and neck is controversial. A meta-analysis conducted by Amit et al. showed that perineural invasion in ACCs of head and neck region is not associated with prognosis, whereas margin status and tumor site are associated such that positive margins and ACC of the sphenoid or ethmoidal sinuses were significant predictors of outcome [3]. In contrast, some studies encountered perineural invasion as a prognostic factor [2].

ACC needs to be differentiated from basaloid squamous cell carcinoma, small cell neuroendocrine carcinoma, polymorphous low-grade adenocarcinoma, and adenosquamous cell carcinoma can be considered [18]. Regarding its histological features ACC predominantly presents as a mixed tumor, consisting of tubular, cribriform and/or solid growth patterns. The tumor is mostly classified according to the predominant pattern; the solid subtype is considered a high grade tumor with poor prognosis. Compared to cribriform and tubular types, solid type ACC shows a high percentage of loss of heterozygosity , more chromosomal aberrations and somatic mutations and a high expression of p53 [12]. Some authors speculate that the risk of nodal metastases is higher when solid ACC is present [9].

CONCLUSION

We introduced a case of ACC that presented with a headache and diplopia. Although rare, ACC can arise from sphenoid sinus and should be considered as differential diagnosis of sphenoid lesions. It is important to gathering imaging and endoscopic findings and note that histopathological examination is useful to differentiate this tumor from others. Moreover, because of perineural spread and distant metastasis this tumor has a poor prognosis.

CONFLICT OF INTEREST

The authors declared no conflicts of interests.

AUTHORS' CONTRIBUTION

All authors contributed in preparing and writing the manuscript.

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