Case Report

Adenoid Cystic Carcinoma Metastatic to The Sellar Area: Report of Two Cases

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Summary

Adenoid cystic carcinoma (ACC) is a slow-growing, locally invasive epithelial tumor that is commonly observed in the head and neck. It arises from the minor and major salivary glands. It may metastasize to the intracranial area through the tissue planes along the vascular and neural structures or by way of skull base destruction. In this study, we present two cases of intracranial ACC.

Case 1: A 19-year-old female with ACC arising in the right lacrimal gland, who had a history of operation conducted 3 years ago, presented to our hospital due to reduced vision in the right eye. Upon examination, she was found to have a new solid lesion in the cavernous sinus.

Case 2: A 43-year-old female patient who has been receiving chemotherapy and radiotherapy during the last 4 years due to nasopharyngeal ACC, demonstrated a new lesion in the paraclinoid area. The first case was treated with surgery and gamma knife, while the second case did not receive any additional treatment after the surgery.

Key words: Adenoid cystic carcinoma, Sellae, metastasis

Özet


Anahtar Kelimeler: Adenoid kistik karsinom, sella, metastaz
INTRODUCTION
Adenoid cystic carcinoma (ACC) is a rare and malignant epithelial tumor that commonly arises in the minor and major salivary glands, and less commonly in the lacrimal gland, naso-pharynx\(^{(2)}\). The incidence of intracranial involvement has been reported to vary between 4-22\%\(^{(1,2,4)}\). These tumors are of slow-growing nature, while being locally aggressive and having a tendency for recurrence. Although local spread takes place along the tissue planes or along the vascular and neural structures, perineural route is a common route of spread for the intracranial area. In addition to ACC usually infiltrates and spreads through bone structure. Although intracranial perineural metastasis commonly involves the Gasserian ganglion, hematogenous metastasis has also been reported\(^{(1,3)}\). In this study, we present two cases of metastatic ACC localized in the cavernous sinus and the paraclinoid area.

CASE PRESENTATION
Case 1
A 19-year-old female patient with no prior complaints presented with swelling, intermittent double vision, and sense of pressure in the right eye. The ocular examination showed complete visual acuity and proptosis, with no visual field defects. Orbital MRI demonstrated an extraconal lesion of 28x20x16 mm size. Subsequent to the total excision, the patient received no additional treatment. Following a 3-year postoperative period, the patient presented again with polyuria, polydipsia, and headache. Cranial MRI study revealed a solid lesion of invasive appearance in the cavernous sinus and right sella (Figure 1). The tumor located in the medial wall of the cavernous sinus and sella was resected by endoscopic endonasal approach. For residual tumor, a second surgery was performed via right pterional craniotomy for the tumor over the temporal mesial area. No post-operative complication occurred. The pathologic diagnosis was adenoid cystic carcinoma (Figure 2). The residual tumor was treated by GK (Gamma Knife).

Case 2
A 43-year-old female patient presented to our hospital with a 6-month history of bilateral secretory otitis media. Magnetic resonance images showed an invasive mass located in the nasopharynx, spreading into the parapharyngeal space, cavernous sinus, and the skull base structure. The patient was diagnosed with ACC based on the punch biopsy performed at the ENT clinic. The patient received 64 Gy RT. Three months later, she presented to us with vision blackouts in the right eye and headache. The MRI showed a mass lesion of 16x12 mm size adjacent to the optical nerve in the left paraclinoid area (Figure 3). The patient underwent a craniotomy with left pterional approach to the sellar area. During the surgery, the lesion was observed to be very closely attached to the falciform ligament. The tumor was resected by opening the optic foramen and the distal ring around the anterior clinoid. The patient postoperative course was uneventful. Histopathologic diagnosis of the mass suggested ACC (Figure 4). She was discharged to home on postoperative day 5 in stable condition. The patient underwent follow up MRI without any additional treatment plans.
**Figure 1:** A) Coronal T1-weighted preoperative postcontrast MRI shows right sellar lesion extending medial temporal area, in addition to pituitary gland and stalk displaced contrlaterally. B) Coronal T1 weighted postoperative postcontrast MRI shows subtotal removal of the tumor after transphenoidal and transcranial surgery.

**Figure 2:** Photomicrography showing cribriform pattern; cystic spaces containing eosinophilic material (original magnification x40, HE)
DISCUSSION

In patients with ACC, treatment strategies are biopsy, radical surgery, and supplemental radiotherapy. In many cases, radical surgery is not possible. In such cases with local invasion, biopsy + subtotal excision is coupled with supplemental RT. The most common treatment for intracranial ACC is surgical resection and supplemental RT\textsuperscript{(5,7,12)}. AAC present a limited response to chemotherapy, use for ACC is controversial\textsuperscript{(10)}. Rarely, chemotherapy has been reported to induce tumor shrinkage\textsuperscript{(8,12)}. The relationship between the histologic appearance and prognosis of the tumor has been investigated by many studies. Tumors with a tubular pattern are correlated with better prognosis, while solid pattern is associated with poor prognosis\textsuperscript{(6)}. Spiro et al. did not

Figure 3: A) Coronal postcontrast preoperative T1-weighted MRI shows left nasopharyngeal tumor. Tumor has spread in this area into the ipsilateral cavernous sinus, which is slightly enlarged compared to the right side. Tumor has spread along the carotid artery into the intracranial area. B) Postoperative T1-weighted coronal MRI shows gross total surgical extirpation.

Figure 4: Photomicrography showing multiple lobules of basophilic tumour cells, the lobules contain polls of mucin. (original magnification x40, HE)
find any difference between the histologic subtypes with regard to prognosis(9).

Although there are rare intracranial cases in which the primary site can not be defined(1), ACC may locally expand to the intracranial area through the perineural and hematogenous route. In a review study evaluating the intracranial spread in 53 cases, tumor involvement was determined to be 35.8% for the Gasserian ganglion, 20.7% for middle fossa, 15% for cavernous sinus, and 5.7% for sellar area(1). Vnelick et al. reported the rate of perineural spread as 52.6%. Although hematogenous metastasis is most commonly reported in the lung, bones, and skeleton, there are cases involving brain metastasis, as well(11).

Three years after the operation, the patient diagnosed with lacrimal gland ACC demonstrated ipsilateral metastasis invading CS (cavernous sinus) and spreading to the sella. Since the superior ophtalmic vein, vein of the lacrimal gland, ends in the CS, the spread may occur via hematogenous route. During the surgery, medial wall of the CS was observed to be completely invaded, while its border with the pituitary gland was indistinct. The tumor was determined to spread to the CS via hematogenous route and to the tentorial notch via emissary vein network, whereas sella was observed to push the pituitary gland to the other side. The tumor was spreading to the intracranial area after encapsulating the carotid. Subtotal surgery could be performed owing to the location and spread pattern of the tumor which had recurred during the past 3 years. Postoperatively, gamma knife was added to the treatment. At postoperative 16 months, the patient is still alive. In the second case which was followed up via RTs at every 3 months after the nasopharyngeal ACC, although the tumor was observed to compress the venous compartments within the CS, it was found to spread to the intracranial area through perivascular route by the medial portion of the distal ring, resulting in a compression over the optic nerve. After the oncology committe decided to perform surgery, the tumor was excised up to the CS by opening the distal dural ring after clinoidectomy. The patient is still followed up without any additional treatment and he appears to be stable with no sign of progression.

ACC of the head and neck should be monitored closely for a possible intracranial spread and metastases. The early detection of recurrences bears great importance for the achievement of effective treatment. The adjacent compartments and their vascular and neural patterns of spread should be followed closely.

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