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Case Report

Skull Base Invasion of Adenoid Cystic Carcinoma of the Lacrimal Gland : A Case Report

Jae Il Lee, M.D.,[1] Young Zoon Kim, M.D.,[1] Eun Hee Lee, M.D.,[2] Kyu Hong Kim, M.D.[1]
Departments of Neurosurgery,[1] Pathology,[2] Masan Samsung Hospital, Sungkyunkwan University School of Medicine, Masan, Korea

Although adenoid cystic carcinoma (ACC) of the lacrimal gland is a rarely encountered orbital tumor, it invades intracranially more frequently than carcinomas of other glands in the head and neck. A 52-year-old man underwent orbital exenteration and resection of intracranially extended tumor via a fronto-orbito-zygomatic approach in combination with a transfacial approach. Histopathologically, the tumor showed perineural, vascular, and lymphatic invasion. Additionally, he received radiotherapy (60 Gy) and adjuvant systemic cisplatin and 5-fluorouracil chemotherapy due to residual tumor in the orbit and systemic metastases (lung, ribs, and spines). He was free of progression and recurrence at 6 months after treatment. The authors report a case of skull base invasion by an ACC of the lacrimal gland to remind neurosurgeons planning intervention that this disease shows a tendency to invade intracranially.

KEY WORDS : Adenoid cystic carcinoma · Skull base · Metastasis · Prognosis.

INTRODUCTION

Adenoid cystic carcinoma (ACC) is an uncommon malignancy that arises in secretory glands, particularly the major and minor salivary glands. It accounts for about 1% of all head and neck malignancies[5,16,17]. Other sites include the tracheobronchial tree, breast, skin, lacrimal gland, female genital tract and prostate. The ACC of lacrimal gland accounts for 8.7% of all ACC[2]. Although ACC of the lacrimal gland is a rare disease, it is the most common malignant epithelial tumor of the lacrimal gland, and represents 11% of epithelial neoplasms of the lacrimal gland[13] and 1.6% of all orbital tumors[10]. ACC was first described in detail by Theodore Billroth, and was initially named cylindroma because of its histopathologic characteristics[3].

ACCs originating in the lacrimal gland invade the intracranial cavity more frequently than carcinomas of the salivary gland. The incidence of intracranial invasion has been reported to range from 4% to 22%. Furthermore, distant metastases are not uncommon[9].

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• Address for reprints : Young Zoon Kim, M.D., M.S.
Department of Neurosurgery, Masan Samsung Hospital, Sungkyunkwan University School of Medicine, 50 Hapseong 2-dong, Hoewon-gu, Masan 630-723, Korea
Tel : +82-55-290-6561, Fax : +82-55-290-6899
E-mail : youngzoon.kim@samsung.com

When ACC of the lacrimal gland invades intracranially, neurosurgeons may be posed with difficult treatment decisions in terms of orbit and eyeball reconstruction, determining the extent of surgical resection, and choosing adjuvant treatment because of the absence of standard therapeutic consensus and risks of recurrence and metastasis. Here, we report the case of a patient with ACC originating in the lacrimal gland, which invaded the skull base, and provide a review of the literature.

CASE REPORT

A 52-year-old man was admitted to our hospital with a complaint of progressive proptosis of the left eye and a medially displaced left globe, which had worsened significantly over the previous month. About one year before visiting our hospital, he started to perceive mild intermittent pain around the left orbit and progressive reductions in visual acuity. A neurological examination conducted at admission revealed visual loss, oculomotor palsy, and trigeminal nerve palsy with absence of corneal reflex on the left side. The left eyelid was tensely swollen and erythematous, and the left conjunctiva was severely chemotic. In addition, a hard, tender, and immobile mass could be palpated around the left orbit.

Computed tomographic (CT) scans of the orbito-facial

area demonstrated a huge solid mass lesion at the left orbital cavity with invasion and destruction of the frontal and temporal skull base and superior maxillary wall. Magnetic resonance (MR) imaging with intravenous gadolinium (Gd) showed moderate and homogenous mass enhancement (Fig. 1). The well-delineated mass extended into the anterior and middle cranial fossa and displaced the left temporal lobe. Left carotid angiography revealed a tumor staining via the ophthalmic artery. Whole body proton emission tomography-computed tomography (PET-CT) revealed multiple metastases to bones (including the spine and ribs) and a lung (Fig. 2).

Three weeks after admission, the patient underwent surgery under a presumptive diagnosis of adenoid cystic carcinoma, a lymphoepithelial tumor (such as lymphoma), or meningioma. Gross total resection of the tumor was done by using a fronto-orbito-zygomatic approach combined with a transfacial approach. The left globe was exenterated and the lateral and superior bony walls were further removed. The tumor had invaded and destroyed the superior and lateral wall of the left orbit, and extended intracranially into the dura of frontal and temporal lobes. The tumor mass in the anterior and middle cranial fossa was completely removed. However, the medial portion of the orbital wall was not completely removed because of the risk of penetrating the sphenoid sinus, which might ascribe to intracranial infection. After resection of the intradural mass, we performed duroplasty with artificial dura. Dead facial space was filled with an abdominal fat graft. Postoperative MR images with Gd enhancement showed small residual mass in the medial part of the left orbital wall (Fig. 3).

The tumor predominantly had a solid pattern and showed focal ductal formation. Perineural and lymphatic invasion and vascular infiltration were suspected at several focal sites.

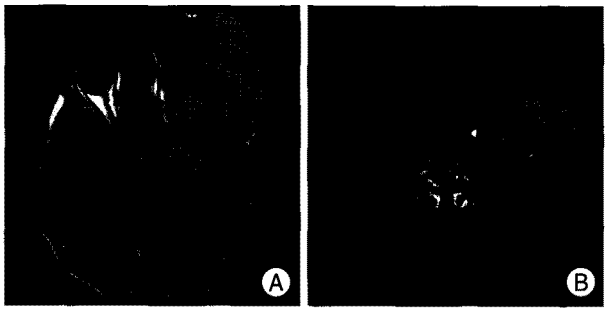

Fig. 1. Preoperative axial (A) and coronal (B) gadolinium-enhanced T1-weighted magnetic resonance image showing a well demarcated moderately enhanced mass in the left orbit. The mass is invading the anterior and middle cranial fossa and causing the left eyeball to deviate medially.

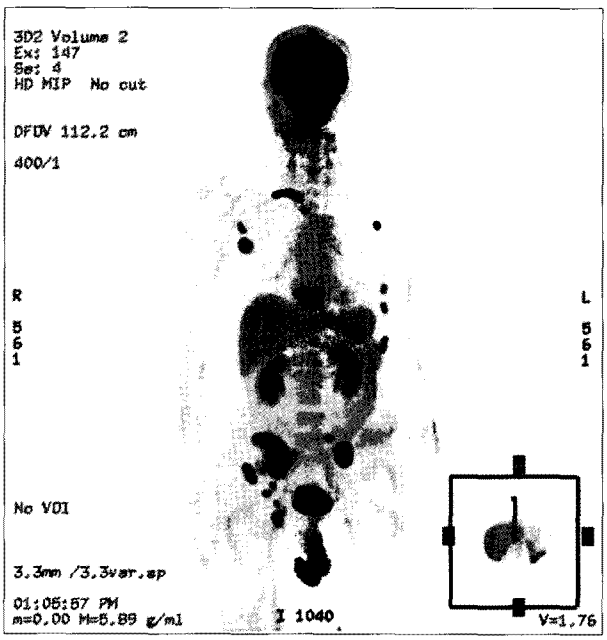

Fig. 2. Whole body proton emission tomography-computed tomography showing multiple metastases with high metabolism in the spine, ribs, and a lung.

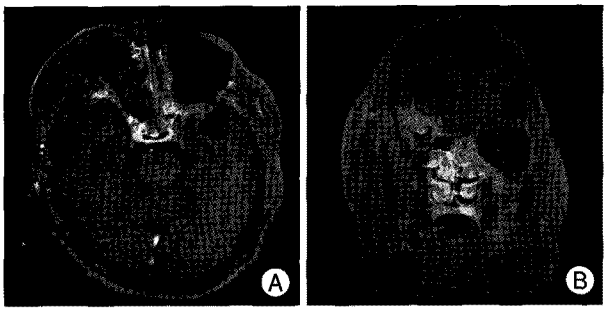

Fig. 3. Postoperative axial (A) and coronal (B) gadolinium-enhanced T1 weighted magnetic resonance (MR) images showing the extent of tumor removal from the left anterior and middle cranial fossa, and the state of the left eyeball after exenteration. The void left after tumor resection is filled with an abdominal fat graft in low signal intensity in T1 weighted MR image.

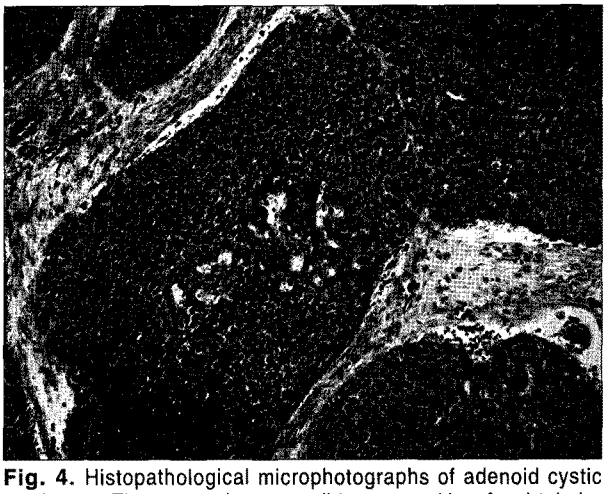

Fig. 4. Histopathological microphotographs of adenoid cystic carcinoma. The tumor shows a solid pattern with a focal tubular appearance (hematoxylin and eosin, original magnification, ×200).

Tumor cells had an abnormally small amount of cytoplasm and exhibited a basaloid appearance with focal necrosis (Fig. 4). The tumor extended into surrounding bony trabeculae, and in these cells, cytoplasm showed positive immunoreactivity for pan-cytokeratin (including low and high molecular cytokeratin), S-100 protein, smooth muscle antigen (SMA), and vimentin. These findings confirmed the diagnosis of ACC. Ki-67 labeling was 20%.

Postoperatively, the patient did well and was relatively free of pain. One month after surgery, he received radiotherapy (a total 6,000 cGy in fractionated doses) because of the known residual tumor in the medial portion of the orbit. Subsequently, he underwent systemic cisplatin and 5-fluorouracil chemotherapy because of the systemic metastases. After 5 cycles of the systemic chemotherapy, follow-up PET-CT showed no progression or recurrence in the tumor bed and in other sites of the body. At 6 months after chemotherapy, he had experienced no further progression or recurrence.

DISCUSSION

Patients with adenoid cystic carcinoma (ACC) usually present with a relatively short symptom duration (less than a year), and follow a painful, rapidly worsening clinical course[1,18-20]. With regard to symptoms, Wright et al. emphasized that pain is an important symptom of lacrimal gland malignancies because it implies perineural infiltration[18]. However, in their series with a primary malignant lacrimal gland neoplasm, it was found that pain was not related to symptom duration, the presence of bone invasion, loss of trigeminal nerve function, or frequency and time of recurrence[19].

Although our patient was 52 years old, ACC patients tend to be younger than those with other lacrimal gland malignancies, for example, in a recent survey of 52 ACC cases, all patients presented at under 30 years of age[19].

ACC is notorious for its slow growth and tendency to recur despite surgical therapy and following radiotherapy and chemotherapy[9]. The appropriate local therapy for ACC of the lacrimal gland remains controversial. Some authorities advocate conservative surgical therapy followed by external-beam radiation therapy or proton-beam therapy, whereas others believe that radical surgery probably results in better local control and possibly better long-term survival[4,18].

The major challenge during the treatment of ACC of the lacrimal gland is the prevention of systemic metastasis. Our case tends to underscore the aggressive nature and likelihood of systemic metastasis from ACC of the lacrimal gland. In keeping with the basic principles of oncology, most authorities would agree that it is desirable to achieve local control by whatever surgical technique or modality that can eliminate the clinically obvious component of any malignancy[7]. Despite the successful systemic control achieved by systemic chemotherapy in our patient, the role of postoperative adjuvant chemotherapy in ACC of the lacrimal gland remains unclear. Meldrum et al. reported promising results in 2 patients who underwent intra-arterial chemotherapy followed by orbital exenteration, and postoperative adjuvant radiation therapy[12]. However, some authorities believe that adjuvant chemotherapy offers little, because conventional chemotherapy has been demonstrated to be only marginally effective in cases of clinically metastatic ACC of the lacrimal gland[6,15].

Perineural invasion is considered an indicator of poor prognosis, because of the inherent risks of spread to the skull base and local recurrence[7,8]. ACC of the lacrimal gland has a greater likelihood of invading intracranially for the following reasons : more neural and vascular structures exist in the orbit; the bones of the orbit are connected directly to the intracranial cavity; and the peri-orbit and nerve sheath are closely connected[14]. These factors increase the risk that an ACC of the lacrimal gland will invade intracranially via perineural, vascular, intraosseous, and leptomeningeal routes and via a nerve sheath. Therefore, neurosurgeons need to be familiar with the tendency of ACC to extend intracranially. In the described case, histopathologic findings showed perineural, vascular, and lymphatic invasions.

From the neurosurgical perspective, Gormley WB et al.[11] summarized strategies for the management of ACC with intracranial extension : 1) when the carotid artery is involved by ACC, a preliminary cerebral revascularization procedure with a cervical carotid to middle cerebral artery vein bypass graft should be performed before tumor resection; 2) when the cavernous sinus is involved, the tumor should be removed as fully as possible, but every effort should be made to preserve the 3rd and 6th cranial nerves to achieve optimal functional and cosmetic results; and 3) when the orbit is involved, it should be exenterated. In terms of palliative surgery, he suggested that it could be considered even in patients with a local disease extent that precludes surgical cure. In the present case, although there was no involvement of ACC in the cavernous sinus fortunately, the orbit was involved, and which made the orbit be exenterated. Until this presenting report, the patient still survives 14 month after surgical resection of ACC in the lacrimal gland with skull base invasion, and adjuvant radiotherapy and chemotherapy were beneficial for preventing progression of systemic disease.

CONCLUSION

The authors report a rare case of ACC of the lacrimal gland that invaded the skull base. Despite its rarity, ACC of the lacrimal gland frequently involves the intracranial region. When establishing a therapeutic plan for ACC with intracranial invasion, careful consideration for surgical extent can prevent recurrence. In view of the poor prognosis and refractory nature of ACC of the lacrimal gland after perineural invasion, optimal adjuvant treatment can be helpful.

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