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# Complete Recovery of Visual Disorder Following Surgical Resection of Adenoid Cystic Carcinoma Arising in the Pterygopalatine Fossa

## *Case Report and Review of the Literature*

*Wei Du, MD, Meng Cui, MD, Peng Li, MD, Jiheng Wang, MS, Ruihua Luo, MD, Jinxing Qi, MD, Ming Zhao, MD, and Weihua Lou, MD*

**Abstract:** Adenoid cystic carcinoma (ACC) arising in the pterygopalatine fossa was rare, only 3 cases have been reported. In previous literature, few authors reported whether the visual deficit could be resolved following the resection of the tumor.

One patient with visual dysfunction induced by ACC arising in the pterygopalatine fossa was reported.

Complete visual recovery was achieved following the operation. And the patient was satisfied with the appearance and the functional results in the follow-up.

Visual loss contributed by the tumor in the pterygopalatine fossa could recover in selected patients.

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**Abbreviation**: ACC = adenoid cystic carcinoma.

## INTRODUCTION

Adenoid cystic carcinoma (ACC) is a malignant tumor characterized by perineural invasion, slow growth, and insidious destruction of surrounding tissues. It usually arises in the major salivary gland, but also in minor salivary gland occasionally.[1] ACC arising in the pterygopalatine fossa is extremely rare, only 3 cases have been reported.[2–4] In these cases, 1 patient presented visual deficit because of neural invasion.[2]

Owing to its proximity to the superior orbital fissure and optic canal, neoplasms arising in the pterygopalatine fossa may cause optic neuropathy by direct invasion or oncothlipsis. Current literature has described possible visual recovery following excision of anterior clinoid mucocele,[5] but few authors reported whether the visual deficit could be resolved after the resection of the tumor arising in the pterygopalatine fossa.

In current study, we described 1 patient with visual dysfunction induced by ACC arising in the pterygopalatine fossa.

## CASE REPORT

The Zhengzhou University institutional research committee approved our study, and the participant signed an informed consent agreement.

A 44-year-old Chinese man presented a mass in the palate for 7 months, there was no ulcer or pain. One month ago, the visual acuity of the right eye dropped gradually and then there was no vision at diagnosis. Preoperative radiographs presented a huge mass in the pterygopalatine fossa and infratemporal fossa (Figure 1A), but we could not determine whether the optic nerve was invaded (Figure 1B). After explaining the possible risks including total blindness to the patient, a surgical plan was formulated. A classic Weber-Ferguson incision was performed. Osteotomies were made on the hard palate after the periosteum was elevated. The optic nerve was noted to just be oppressed by the tumor. After carefully distraction, the tumor was resected completely. Intraoperative frozen section and postoperative pathology both showed a diagnosis of ACC. And the patient reported the visual acuity had returned to normal at 10 days after the operation. At 2 months after discharge from the hospital, there was no recurrence of the tumor (Figure 1C), and the patient was satisfied with the appearance and the functional results.

## DISCUSSION

The most distinctive biological behavior of ACC was perineural spread.[1] In previous case reports, facial pain or visual deficit or hypoglossal nerve palsy were the main complaint.[2–4] All these findings supported the above-mentioned viewpoint. In current study, optic dysfunction was also noted, but it would be contributed by external compression rather than direct tumor invasion.

Various surgical approaches for the pterygopalatine fossa and infratemporal fossa have been advocated. It could be divided into 3 groups inferior approach, lateral approach, and anterior approach.[6,7] Inferior approach could not provide adequate exposure of the retromaxillary and the skull base, and it was usually suggested for the tumor restricted in the parapharyngeal space.[6,7] Lateral approach was limited for tumors extending medial to the pterygomaxillary fissure and especially those stretching across the medline.[6,7] The maxillary swing approach was first introduced by Wei et al[8] for recurrent nasopharyngeal carcinoma as a novel anterior approach.

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From the Department of Head and Neck and Thyroid, Affiliated Tumor Hospital of Zhengzhou University, Henan, PR China (WD, MC, PL, JW, RL, JQ, MZ); Department of Head and Neck, The First Affiliated Hospital of Zhengzhou University, Henan, PR China (WD, WL).
Correspondence: Weihua Lou, Department of Head and Neck, The First Affiliated Hospital of Zhengzhou, University, Henan, PR China (e-mail: duweitj@126.com).
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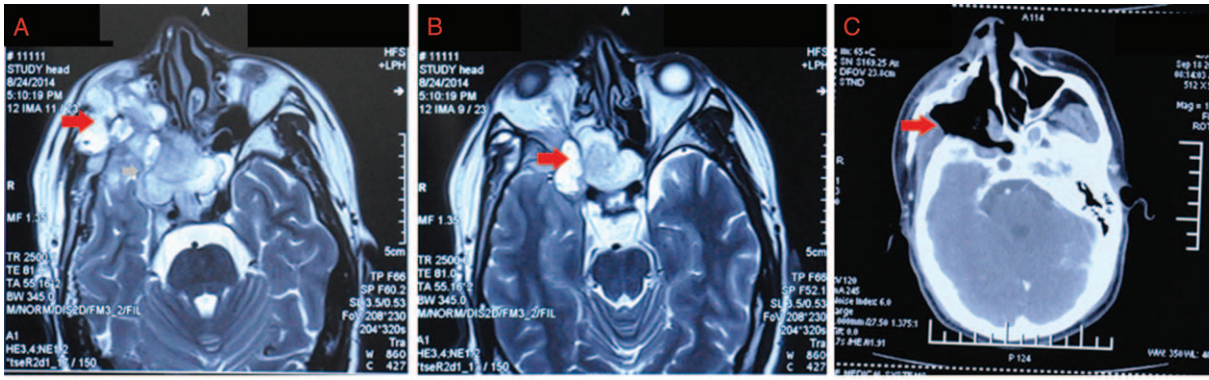


**FIGURE 1.** (A) Huge space-occupying lesion in the pterygopalatine fossa and infratemporal fossa was noted. The surrounding orbital bone was damaged. A malignant tumor tended to be the primary diagnosis. (B) The tumor involved the orbital apex, but the association between the optic nerve and the tumor could not be justified clearly. (C) At 2 months after discharge from the hospital, there was no recurrence of the tumor.

Nowadays, the route has been the preferable approach for tumors arising in the pterygopalatine fossa and infratemporal fossa.

Compared to the facial translocation approach, the maxillary swing approach did not require neurorraphy and could avoid the development of a free facial graft by keeping the anterior maxilla and translocated hard palate attached to the cheek flap. In current study, an adequate exposure of the lesion was achieved and the tumor was resected completely. Possible postoperative complications such as palatal fistula, epiphora, and trismus have been reported. But in current study, no complication was noted.

The most interesting finding in such case was complete recovery of visual disorder following surgical resection of the disease. Previous authors have studied the factors related to vision recovery after optic nerve decompression. Suri et al[9] have studied the visual outcome in patients with suprasellar tumors who experienced preoperative blindness, and the authors found male sex, shorter duration of blindness, operative evidence of hemorrhage in tumor, and soft tumor consistency were significantly associated with the visual outcome; another research conducted by Mathiesen et al[10] presented early optic nerve decompression and primary tumors seemed to predict better visual prognosis. Similarly, Kitano et al[11] compared postoperative improvement of visual function among different approaches in treating suprasellar meningiomas, and the authors found extended transsphenoidal approach might resulted in improvements in visual acuity. Other associated factors, such as the size of mass, the pathology type, tumor consistency and, so on, have also been proven in the literature.[12] The same principle might also be compliant to the present case.

In summary, ACC arising in the pterygopalatine fossa is extremely rare. Visual loss contributed by the tumor in the pterygopalatine fossa could recover in selected patients.

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