

## Adenoid cystic carcinoma of the hard palate presenting as ipsilateral sixth nerve palsy

Akshay Gopinathan Nair<sup>1,2</sup>, Indumati Gopinathan<sup>3</sup>,  
Rima Singh<sup>4</sup>, Rima S Pathak<sup>5</sup>

Adenoid cystic carcinoma (ACC) is an uncommon malignant neoplasm composed of basaloid epithelial and myoepithelial cells. The palate is the most commonly involved intraoral site for ACC. Here, we document the case of an advanced ACC arising from the hard palate that presented with right-sided sixth nerve palsy in a 75-year-old male with no other systemic illnesses. ACC of the head and neck involving the cavernous sinus and presenting as isolated sixth nerve palsy is exceedingly rare. In the absence of vasculopathic or ischemic risk factors, regardless of the age of the patient; neuroimaging should be performed in cases of isolated nontraumatic sixth nerve palsy.

**Key words:** Abducens palsy, cavernous sinus, diplopia, lateral rectus palsy, metastasis, salivary gland tumor

Adenoid cystic carcinoma (ACC) comprises about 1% of all malignant tumors of the oral and maxillofacial region.<sup>[1,2]</sup> The most common clinical presentation of ACC is a low-grade, dull pain that gradually increases in intensity over time. Due to the propensity of ACC to spread through perineural invasion; the tumor, sometimes can extend far beyond the obvious tumor margin. Here, we present a case where isolated nontraumatic sixth nerve palsy was the sole presenting symptom of ACC of the hard palate with contiguous perineural spread to the cavernous sinus.

### Case Report

A 75-year-old male presented with a 1-week old history of sudden onset binocular diplopia. He had been diagnosed by his primary ophthalmologist, on day one, as right sixth nerve palsy of presumed ischemic origin and had been advised prism glasses. On examination, both eyes had visual acuities of 6/6 N6. Intraocular pressures, pupillary reactions, and dilated fundus examination of both eyes were normal. Ocular motility evaluation

showed that the patient had an isolated right-sided abduction deficit [Fig. 1a-c]. There was no proptosis or enophthalmos. All other cranial nerves were normal. Systemically, the patient had no vasculopathic risk factors or other illnesses. On eliciting a detailed systemic history with pointed questions, the patient disclosed that he had been suffering from a dull toothache in the region of his right upper molars and had difficulty in chewing for the last one month; for which, he had not sought any treatment. Magnetic resonance imaging (MRI) of the head was performed. A mass measuring 4.2 cm × 3 cm × 2.2 cm was seen arising from the right half of the hard palate with solid contents eroding into the right maxillary sinus through the floor along with destruction of the right maxillary-alveolar ridge in the region of the molar teeth [Fig. 2a]. Posteriorly, it was seen extending up to and involving the right anterior cavernous sinus close to the right superior orbital fissure [Fig. 2b and c]. A small portion of the right anterior Meckel's cave was also invaded by the tumor. An incisional biopsy of the hard palate was performed; the histopathological examination of which, showed basoloid tumor cells in a cribriform pattern with eosinophilic material-consistent with ACC. A positron emission tomography-computed tomography scan showed high uptake in the region of the primary malignancy as well as multiple cervical lymph nodes. All treatment options were explained to the patient who chose to undergo palliative radiation and received 30 cGy in 10 fractions. He was prescribed a Fresnel "press-on" prism for the diplopia. The patient succumbed to the disease 4 months after presentation.

### Discussion

This case highlights two issues: first is the unusual perineural spread that led to abducens palsy being the sole presenting feature of a hard palate ACC; the second is the role of neuroimaging while evaluating a patient with nontraumatic isolated sixth nerve palsy.

High-resolution, fat-suppressed, and contrast-enhanced MRI is the investigation of choice for detection of abnormal perineural enhancement.<sup>[3]</sup> The Gasserian ganglion is the most frequently involved whereas the cavernous sinus is less frequently affected by ACC.<sup>[4]</sup> Presenting signs and symptoms are related to the anatomical site of the lesion and ACCs involving the cavernous sinus can be asymptomatic or present with involvement of either 3<sup>rd</sup>, 4<sup>th</sup>, 5<sup>th</sup>, and 6<sup>th</sup> cranial nerves and the internal carotid artery.<sup>[5]</sup> The perineural spread of the tumor from the hard palate to the cavernous sinus follows the course of the maxillary division of the trigeminal nerve: sensory fibers of the greater and lesser palatine nerves, respectively, pass through the greater or lesser palatine foramen and then go upward along the descending pterygoid canal to enter the pterygopalatine fossa. From here, the fibers join the main trunk

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<sup>1</sup>Ophthalmic Plastic Surgery and Ocular Oncology Services, Aditya Jyot Eye Hospital, <sup>2</sup>Advanced Eye Hospital and Institute, <sup>3</sup>Clinico-Path Labs, <sup>4</sup>Dr. Mukherji's Dental Centre, <sup>5</sup>Department of Radiation Oncology, Tata Memorial Hospital, Mumbai, Maharashtra, India

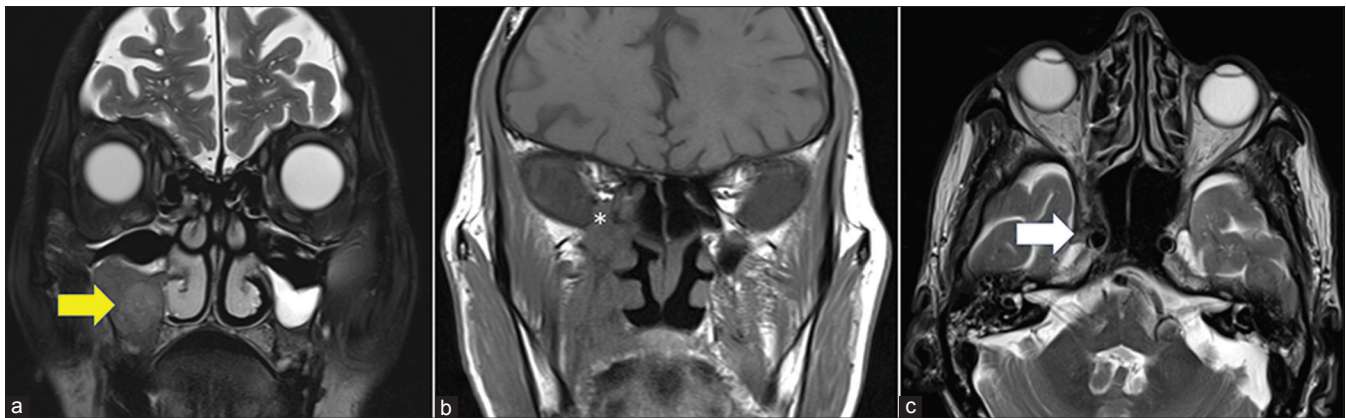
**Correspondence to:** Dr. Akshay Gopinathan Nair, Aditya Jyot Eye Hospital, 153, Major Parmeshwaran Road, Wadala, Mumbai - 400 031, Maharashtra, India. E-mail: [akshaygn@gmail.com](mailto:akshaygn@gmail.com)

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**Figure 1:** Clinical photograph of the patient demonstrating right abducens palsy



**Figure 2:** A coronal T2-weighted fat-saturated fast spin-echo sequence image (a) showing a large mass (yellow arrow) arising from the right half of the hard palate with solid contents eroding into the right maxillary sinus through the floor. (b) Is a T1-weighted turbo spin echo coronal slice showing the posterior extent of the tumor (white asterisk). The bony erosion is evident on comparing the normal anatomy on the contralateral side. (c) Is a T2-weighted turbo spin echo axial image showing the tumor involving the cavernous sinus and almost encasing the internal carotid artery (white arrow)

V2 then course posteriorly through the foramen rotundum to reach the cavernous sinus.<sup>[6]</sup>

Dumitrascu *et al.* compiled a comprehensive review of all reported cases of histologically confirmed ACC with cavernous sinus involvement identified in the literature.<sup>[7]</sup> They identified 27 articles that reported 32 individual cases of ACC with infiltration of the cavernous sinus. Of these 32 cases, only 8 of them originated from the palate. However, all of these eight cases had multiple cranial nerve palsies with none of them presenting as isolated abducens nerve palsy.

In our case, retrospectively, prompt neuroimaging had yielded a diagnosis. However, one can argue that it may have acceptable to defer immediate imaging in this patient with a presumed diagnosis of ischemic sixth nerve palsy.<sup>[8]</sup> Literature shows that there is no clear consensus on the timing of neuroimaging in isolated sixth nerve palsy. Patel *et al.* on the basis of their population-based study suggested that patients with nontraumatic neurologically isolated sixth nerve palsy may undergo a focused medical evaluation followed by close observation, whereas nonneurologically isolated cases warrant a full neurologic evaluation including neuroimaging.<sup>[8]</sup> With this recommendation, by delaying an MRI in an isolated nontraumatic sixth nerve palsy, the risk of not diagnosing an intracranial tumor exists— as would have been the case in our patient had imaging been delayed. Conversely, Bendszus *et al.* have recommended that MRI should routinely be performed in patients presenting with an acute sixth nerve palsy, even those with evidence of a vasculopathy.<sup>[9]</sup> This, however, may not be feasible in every case. The middle path, which was proposed by Nair *et al.* recommends that in patients >50 years, with the

presence of vasculopathic risk factors or multiple sclerosis, may be treated conservatively with focused management of the underlying systemic condition. However, worsening, nonimprovement, and progressive involvement of other cranial nerves are indications for neuroimaging. In patients over 50 years of age, isolated sixth nerve palsy in the absence of vasculopathic risk factors requires neuroimaging.<sup>[10]</sup>

## Conclusion

ACCs of the hard palate have a high propensity for perineural spread, which has considerable implications for prognosis and treatment. Involvement of the cavernous sinus is relatively uncommon for ACCs and in rare cases it may present with isolated cranial nerve palsy. In the absence of vasculopathic risk factors, regardless of the age of the patient; neuroimaging should be performed in cases of isolated nontraumatic sixth nerve palsy.

## Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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## Conflicts of interest

There are no conflicts of interest.

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