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Adenoid Cystic Carcinoma of Paranasal Sinuses – Review from Ophthalmic Standpoint

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ABSTRACT

Adenoid cystic carcinomas (ACC) comprise 5% of sino-nasal malignancies. Histologically, the cribriform pattern is the most common variant while the solid type confers a poor prognosis. A non-specific clinical picture in the initial stages of tumor growth necessitates a high index of clinical suspicion from the ophthalmologist upon discerning subtle clinical clues to guide the radiologist in identifying foci of tumor cell activity. With routes of tumor spread being myriad, it is imperative that patient is monitored for signs of tumor recurrence as recurrences are frequent, often present late, years after treatment of primary tumor complex.

Keywords: Adenoid cystic carcinomas, Paranasal sinuses, Sino-nasal malignancies

INTRODUCTION

Adenoid cystic carcinomas (ACC) comprise 5% of sino-nasal malignancies. They are neoplasms of epithelial cell origin that characteristically originate from maxillary sinus (48-54.7%) and less commonly from ethmoid sinus (4.2-16%) and sphenoid sinus (3-12%) [1-3]. They have a unique female predilection and present clinically, often, from the fourth decade to the sixth decade [4].

HISTOLOGY

The different histological subtypes of ACC include tubular, cribriform and solid pattern. The solid variant is a red flag sign, being frequently equated to an aggressive behavior and a poor prognosis while the cribriform pattern is the most frequently reported histological subtype in literature. Identification of c-kit in immunohistochemical assessment of tumor confers poorer prognosis in tubular and solid ACC subtypes while tumors with epidermal growth factor receptor (EGFR) expression are associated with better 3 year survival rate [3,5].

CLINICAL PICTURE

ACC is characterized by non-specific clinical picture in the initial stages of tumor growth- nasal congestion, subtle discomfort, facial pain and altered sensation in the trigeminal distribution. Non-specific symptomatology coupled with slow, insidious tumor growth leads to diagnostic delay. Other common presenting features include recurrent epistaxis, maxillary pain and unilateral nasal obstruction. Neuro-ophthalmic manifestations either due to primary tumor growth or its recurrence include diplopia and movement limitation secondary to involvement of cranial nerves (III, IV and VI), ptosis, pupillary abnormalities (Horner's syndrome, Adie's pupil) and reduced corneal sensation. The time between onset of initial symptoms and partial cavernous sinus syndrome ranged between 6 months and 6 years. It is imperative that patient is monitored for signs of tumor recurrence as recurrences are frequent, often present late, years after treatment of primary tumor complex [3,5,6].

PATHOPHYSIOLOGY AND DIAGNOSTIC CLINICAL SIGNS

Adie's pupil is often noticed due to ipsilateral metastatic involvement of neurons in the ciliary ganglion. It is characterized by anisocoria that increases in light, vermiform constriction of the affected larger pupil coupled with an absent ipsilateral consensual reflex and noticeable lag in the near response (slow redilatation). Super sensitivity to 0.125% pilocarpine constricts the affected pupil.

Diplopia may occur secondary to movement limitation either due to perineural tumor spread, direct invasion of cavernous sinus by tumor cells or intracranial invasion [5]. Corneal

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sensations may be reduced (using aesthesiometer or wisp of cotton) probably due to direct involvement of second division of trigeminal nerve that innervates the maxillary sinus [5].

EXAMINATION OF VISUAL PSYCHOPHYSICS

Measuring visual acuity is of paramount importance in addition to documenting the presence of RAPD (Relative Afferent Pupillary Defect) and any color vision defect (Ishihara color vision chart) or diminished contrast sensitivity response (Pelli-Robson chart) to rule out an intracranial invasion of the optic nerve. It is relevant to measure the near visual acuity using patient's own spectacles or reading glasses as patients with Adie's pupil have blurred near vision due to a noticeable lag in shifting fixation from distance to near or vice-versa. Examination of pupillary reflex (direct, consensual and near reflex) will help identify any defect of the afferent or efferent pupillary pathway due to tumor spread. Ocular motility testing should be done to assess individual eye muscles (ductions) and conjugate eye movements (versions). It is important to examine the patient in primary position and then in all cardinal positions of gaze.

Evaluation of head posture and its correlation with movement limitation is critical to identify involvement of corresponding cranial nerves (for example, right face turn is usually associated with limitation of abduction in right eye). Diplopia and Hess charting are done to confirm the nerve palsy (restrictive or paralytic), document the baseline for future comparison and assess for improvement with simultaneous appropriate cancer therapy. Confrontation fields using static images may point to involvement of visual pathway.

ROUTES OF TUMOR SPREAD

The peculiarity of ACC lies in their local invasiveness, perineural spread (50%) and delayed recurrence consequent to their proximity to adjacent anatomical structures and resultant incomplete surgical tumor clearance despite palliative radiotherapy and chemotherapy [2,7,8,9]. Routes of spread are myriad- perineural, hematogenous (to lungs, bone, liver and brain), direct invasion of cavernous sinus or intracranial involvement [5,10-14]. Subtle neuro-ophthalmic signs like Adie's pupil (due to perineural tumour seeds from cavernous sinus along first division of trigeminal nerve, coursing along nasociliary nerve to ciliary ganglion through Annulus of Zinn) are essential pointers to site of tumor cell activity and recurrence [5,15] Skull base extension, an adverse prognostic factor for survival, may occur along Eustachian tube, internal carotid artery, maxillary and mandibular nerve [5,16]. Gandour-Edwards et al. [17] have reported that tumors with dural invasion express neural cell adhesion molecules increasing their affinity for the dura [17].

NEUROIMAGING

While CT imaging reveals bony destruction and extension to skull base [5], contrast enhanced MRI with fat suppression is key to identify perineural enhancement [6]. Radiologic signs that outline cavernous sinus involvement are sinus enlargement, convexity of lateral dural margins and replacement of trigeminal cistern with soft tissue [18].

TREATMENT

Surgical resection, if possible, is the most effective treatment modality [19] with adjuvant radiotherapy and chemotherapy in cases of residual tumor activity, positive surgical margins on frozen section and advanced tumor stage. Signs of perineural spread may necessitate shift in treatment goal from cure to palliation [2,6].

PREDICTORS OF OUTCOME/SURVIVAL RATES

Prognosis in ACC is determined by histological subtype (poor with solid pattern) [3], tumor location (better with maxillary sinus compared to ethmoid or sphenoid sinus) [2] and tumor stage (poorer with advanced TNM stage) [14]. Prognosis is circumspect in tumors with perineural invasion [20], extension to skull base and positive surgical margins on frozen section [5,20]. No difference in survival rate was observed between open and endoscopic surgical approach [3].

CONCLUSION

Clinical behavior of ACC necessitates a high index of clinical suspicion from the ophthalmologist, possible only on discerning subtle clues like Adie's pupil or loss of corneal sensation to help the radiologist pinpoint signs of tumor activity through relevant and accurate imaging techniques. A multidisciplinary collaboration is of paramount importance in facilitating early tumor identification and monitoring clinical course for a considerable impact on long-term survival by negating any diagnostic or therapeutic challenge.

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