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Recurrent Orbital Cylindroma in a Teenager

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Abstract: *Adenoid cystic carcinoma (ACC), is one of the most common primary malignant tumors of the lacrimal gland. It accounts for less than 4% of all lacrimal gland tumours. Mean age of incidence is 40 years[1]. It has a poor prognosis, in view of its locally invasive and metastatic potential[2-7]. Radical Excision with negative margin is the treatment of choice. However, the complex anatomy of orbital narrows the extent of radical resection in many cases. We report a case of a 16 year old boy in whom ACC of the lacrimal gland recurred locally in a multifocal pattern following a 3 year disease free interval, after initial excision and adjuvant radiation therapy. The case is unique due to its rarity in the age group as well as successful application of conservative approach in the extent of salvage surgery.*

Keywords: *Adenoid Cystic Carcinoma, Cylindroma, Orbital Tumours*

I. Case Report

A 13 years old boy underwent excision of apparently benign left orbital tumor from a local hospital and was subsequently diagnosed as lacrimal gland adenoid cystic carcinoma after the final histopathological analysis, in 2012. He further received adjuvant radiation therapy from our centre in view of close margins. After a 3 year period of disease free interval, he presented with painful swelling of left eye, diminution of vision and ophthalmoplegia of 1 month duration in November 2014. Ophthalmological evaluation revealed lateral gaze restriction in left eye with minimal proptosis and preseptal cellulitis that was treated conservatively. Swelling and pain subsided following spontaneous pus extrusion with improvement in vision and ophthalmoplegia. The magnetic resonance imaging (MRI) showed a homogenously enhancing 2 x 1.4cm mass lesion in superolateral aspect of left orbit in the extraconal space (Figure 1) abutting the globe not separate from lacrimal gland, and another similar sized lesion (Figure 2) in the temporal fossa, in close proximity with lateral orbital wall and zygomatic bone.

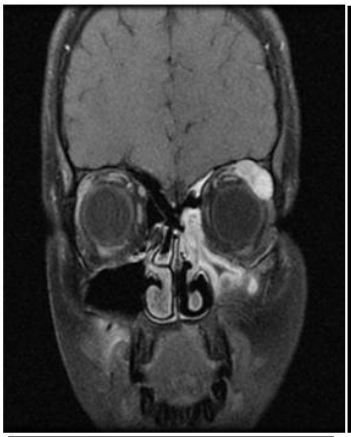

Figure - 1

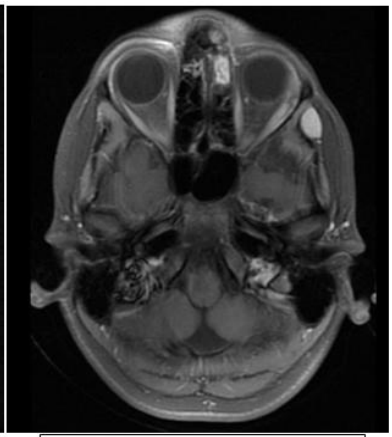

Figure - 2

In view of multifocal recurrence and anticipated margin positivity with a wide local excision of the tumor, he was considered for salvage surgery amounting to orbital exenteration. However, B-mode ultrasonographic scan revealed no globe infiltration. His visual acuity was 6/18. Considering the young boy who is keen on preserving his vision, no eye globe infiltration in the B mode scan and preserved visual aquity, a complete wide local excision was attempted, thus avoiding exentration. However, a consent for exenteration was taken in case the intraoperative findings mandate an exentration. The possibility of an exentration at a later date, in case the final histoopathology report shows margin positivity, was also explained.

Modified Stallard incision was put and muscular flaps elevated. Lateral orbitotomy was done, excising 2cm bone of orbital rim. Tumor was identified medial to periorbita (Figure-3), and was excised in-toto after incising the periosteum just below the swelling. There was no infiltration to globe or extraoccular muscles. A

satellite swelling in the inner aspect of the temporalis muscle at the anterior aspect was noted, for which temporal fossa was cleared (Figure-4). The lateral orbital rim was replaced and fixed with mini-plates and screws. The wound was closed with a drain that was removed on POD-3. Post op ophthalmological evaluation 1 month later revealed normal eye mobility with acuity same as prior to surgery. He is on follow up for last one year and is currently disease free, with good visual acquity.

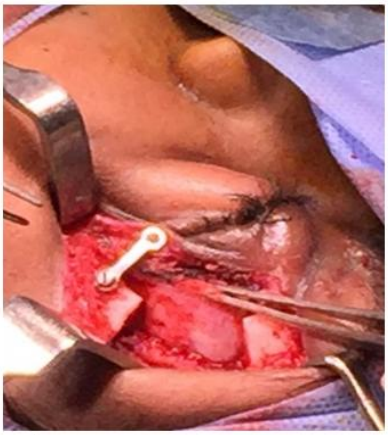

Figure - 3

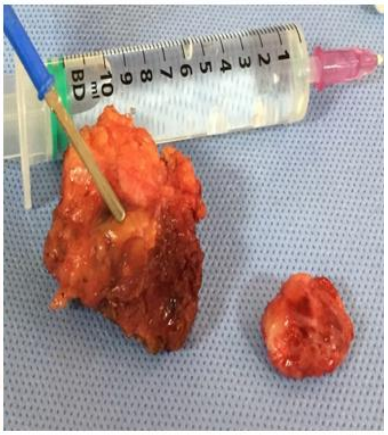

Figure - 4

I. Discussion

ACC, first described by Theodore Billroth, was initially named cylindroma because of its specific histopathologic characteristics[8, 9]. It arises in secretory glands accounting for about 1% of all head and neck malignancies[10-12] .Though rare, ACC is the most common malignant epithelial tumor of the lacrimal gland, representing 11% of epithelial neoplasms of the lacrimal gland [13] and 1.6% of all orbital tumors[14]. Though slow growing, it has poor prognosis with data[1] reporting a survival rate of less than 50% at 5 years and 20% at 10 years regardless of the local treatment regimens. This has been attributed to the complex regional orbital anatomy and the aggressive biological behavior of the tumor causing recurrence. The propensity for perineural invasion results in local invasion and recurrence, especially to skull base. They rarely metastasize to lungs, brain and bone in decreasing order of frequency. The differential diagnoses include inflammatory pseudotumor (dacryoadenitis), pleomorphic adenoma, and adenoid cystic carcinoma of the lacrimal gland.

A multidisciplinary approach is needed to tackle lacrimal gland ACC. Complete surgical resection in lacrimal gland tumour alone, is often difficult to attain, due to adjacent critical structures such as optic nerve, optic chiasm, temporal lobes, brain stem etc. Although most authors advocate the use of surgical excision and postoperative radiotherapy[,3,6,11] some series have found no statistically significant difference when compared with those treated with surgery alone.[4,12]

Adjuvant radiotherapy is advocated in locally advanced ACC of lacrimal gland with incomplete resection, close or positive margins, or peri-neural spread. This has showed improved loco-regional control with acceptable morbidity. Huber et al. reported that fast neutron radiotherapy provided higher local control rates than a mixed beam and photons in advanced, recurrent, or incompletely resected ACC [10].

Careful tumor staging and grading with documentation of perineural invasion and margin status are important for prognostication. Though at experimental stage, Ki-67 and p53 are being analyzed to assess biological behavior of such tumours, to correlate with their aggressiveness and tendency to recur. Few reports favour aggressive approach with exenteration in view of high chance for local recurrences and metastasis occurring years after the initial presentation. The clinical course is heterogeneous, with some patients surviving decades and others surviving only months. In this case report, even though a salvage setting, considering the heterogeneous clinical course, we opted to go ahead with organ preserving approach as the patient was quite young with fully intact vision and no infiltration of neighboring structures intraoperatively.

II. Conclusion

Considering the heterogeneous clinical course, a conservative approach for the management of selected malignant epithelial tumors of the lacrimal gland whenever feasible, to offer the best the quality of life for the patient. Close surveillance is essential to pick up early recurrence and to offer salvage curative treatment.

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