CASE REPORT

Adenoid Cystic Carcinoma of Lacrimal Gland

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ABSTRACT

Adenoid cystic carcinoma is a relatively uncommon tumour of salivary glands and is characterised by a prolonged clinical course and a fatal outcome. It was first described as ‘cylindroma’ by Billroth in 1859. Half of these tumors occur in glandular tissues other than the major salivary glands; principally in the hard palate, but they can also arise in the tongue and minor salivary glands. Unusual locations include the external auditory canal, nasopharynx, lacrimal glands, breast, vulva, esophagus, cervix and Cowper glands. The long natural history of this tumor and its tendency for local recurrence are well known.

Key Words: Adenoid cystic carcinoma, lacrimal gland

Case Report

A 43 year old female, known diabetic and hypothyroid was admitted with the complaint of swelling in the left eye which had lead to the protrusion of the eye ball over a period of one year. The patient gave no complaints of diminished vision, diplopia, headache, nasal discharge or nasal obstruction. Clinically visual acuity, visual fields, fundi, and extraocular motility were normal. No abnormality was detected in the ear, nose and throat. The patient had significant proptosis of the left eye, which was pushed downwards. There was no evidence of visual field defects. MRI revealed a mass lesion in the left lacrimal fossa pushing the eye ball downwards and outwards. An eye ball sparing total excision of the tumor was carried out. The final biopsy report was suggestive of an adenocystic carcinoma exhibiting a cribriform pattern with perineural invasion. Patient was planned for intensive modulated radio therapy after a planning CT and MRI, immobilisation was done, the planning was done using Brain Lab Planning system. The therapy was delivered on 600 CD variant linear accelerator. The dose delivered was 54 Gy/30 fractions at 180 cGy/fr to 90% isodose level. The patient is stable and disease free on two years of follow up.

Discussion

Adenoid cystic carcinomas (ACC) of the lacrimal gland are rare malignant tumours accounting for 1.6% of all orbital tumours. Despite their rarity they are the second most frequent epithelial neoplasms occurring in the lacrimal gland after pleomorphic adenomas. It has a slightly higher incidence in females. The peak incidence is in 5th to 7th decade. Blood
borne metastasis in 40 to 60% are common with the lung being the recipient in 40% of the cases. They are slow growing tumours, which tend to spread to adjacent structures and occasionally metastasize via haematogenous spread to lungs, brain and bone in decreasing order of frequency.\(^{1,2}\) ACC in general has a slow biological growth and tends to have a protracted course with a poor outcome, with a reported survival rate of less than 50% at 5 years and 20% at 10 years.\(^{3,4}\) Lacrimal gland ACC displays a similarly aggressive behavior with an overall survival of 20% at 10 years.\(^{1,2}\) A primary Adenoid cystic carcinoma usually is treated with radical surgery. The rarity of lymph node metastasis suggests that radical node dissection is unnecessary. While radiation therapy has a 96% response rate, it also has a 94% incidence of subsequent local recurrence and is thus a poor treatment when used alone. Planned combinations of surgery with preoperative or more commonly postoperative radiotherapy have shown improvement in both local control and survival. The value of adjuvant chemotherapy after surgery has not been explored fully.

References