Rare disease

Adenosquamous carcinoma of the lacrimal gland

Iype Cherian,¹ Sachet Pravat Shrestha,² Moti Lal Panhani,³ O P Talwar,⁴ Nikunja Yogi,⁵ Shiwani Rai,⁶ Subita Lalchan,⁷ Cecilia Fernandes⁵

¹ Department of Neurosurgery, Manipal College of Medical Sciences, Phulbari, Pokhara, Kaski, Nepal

² Ophthalmology Department, Manipal College of Medical Sciences, Phulbari, Pokhara, Kaski, Nepal

³ Plastic and Reconstructive Surgery Department, Manipal College of Medical Sciences, Phulbari, Pokhara, Kaski, Nepal

⁴ Pathology Department, Manipal College of Medical Sciences, Phulbari, Pokhara, Kaski, Nepal

⁵ Department of Surgery, Manipal College of Medical Sciences, Phulbari, Pokhara, Kaski, Nepal

⁶ ENT Department, Manipal College of Medical Sciences, Phulbari, Pokhara, Kaski, Nepal

⁷ Department of Radiodiagnosis, Manipal College of Medical Sciences, Phulbari, Pokhara, Kaski, Nepal

Correspondence to Iype Cherian, driypecherian@gmail.com

Summary

A 48-year-old man presented with proptosis of the left eye, which on excisional biopsy proved to be primary adenosquamous carcinoma of the lacrimal gland. The lesion was excised radically by a limited frontotemporo-orbitozygomatic approach and any further surgeries, such as exenteration, were avoided in view of the patient's wish to preserve the eye and vision. Long-term follow-up is planned to look for any metastasis or recurrence of the tumour. Primary adenosquamous carcinoma of the lacrimal gland is a very rare entity and, following a thorough literature review, only two cases of lacrimal gland adenosquamous carcinoma have been reported so far.

BACKGROUND

Lacrimal gland adenosquamous carcinoma is a very rare entity and has been reported in the literature on only two occasions as far as we know.

CASE PRESENTATION

A 48-year-old man presented with proptosis of the left eye of 5 years' duration with a rapid progression in the last 4 months. This was associated with painful movement of the eyeball, hyperesthesia on left supraorbital region, itching and occasional watery discharge for the last 4 months. There was no diminution of vision, diplopia or visual field defect. The rest of the systemic examination and medical history provided no additional information. There was no relevant past, personal, social or family history.

INVESTIGATIONS

CT scan showed a large intraorbital mass in the superolateral aspect of the left orbit with calcifications. There were no intracranial extensions and there was no extension into the superior orbital fissure.

DIFFERENTIAL DIAGNOSIS

Dermoid tumour and lacrimal sac tumour were considered.

TREATMENT

Excision of the tumour was done by orbitocranial approach using limited frontotemporo-orbitozygomatic craniotomy. The tumour was a 3×3 cm mass, pearly white, cystic in nature and contained pultaceous material mimicking a dermoid tumour.

The mass was sent for histopathology, the tissue was processed and slides were stained with H&E and periodic acid-Schiff. The tumour was a mixed tumour with a glandular and epidermoid component (figure 1). The possibility of

mucoepidermoid carcinoma and adenosquamous carcinoma were considered as differential diagnosis.

Mucoepidermoid is more common than adenosquamous in the lacrimal gland; however, in this case, there were separate areas: glandular and the squamous elements with keratinisation within the epidermoid component. In mucoepidermoid carcinoma these two components are inseparable and present in the same nest of tumour and keratinisation is an uncommon feature and, thus, more in favour of adenosquamous carcinoma of the lacrimal gland.

Postoperative radiotherapy was planned but the patient refused because of the possible effect on vision.

OUTCOME AND FOLLOW-UP

Postoperative CT of the head and orbit showed no residual lesions. The patient has no recurrence of the tumour at 1 month follow-up following the complete excision of the tumour.

DISCUSSION

The lacrimal gland is a minor salivary gland composed of acini with an inner layer of cuboidal to columnar zymogen-bearing cells and an outer layer of myoepithelial cells. The aponeurosis of the levator palpebrae divides the gland into a superficial palpebral lobe and a deep orbital lobe. Lacrimal gland tumour represents 5–25% of the orbital tumour, and the proportion that are epithelial range from 23–70% of biopsied cases in the literature. Most of the tumours of the lacrimal gland are basically epithelial tumour or mixed tumours and others are lymphomas and adenocarcinoma.¹ The most common benign lesions are pleomorphic adenomas and dacryops, whereas, among the malignant lesions, adenoid cystic carcinoma is the most common (66%), followed by carcinoma ex pleomorphic adenoma (18%), primary adenocarcinoma (9%) and mucoepidermoid carcinoma (3%).²

Figure 1 Squamous cell carcinoma with keratinisation (arrow) with cells showing intracytoplasmic mucin (inset) (H&E $\times 100$; $\times$ inset 400).

Adenosquamous carcinoma is a rare neoplasm of the upper aerodigestive tract developing mainly at the nasal, oral and laryngeal cavities with a high propensity to regional lymph node metastasis and a short average survival.³ Histologically, it is characterised by the simultaneous presence of both glandular and squamous components—with the latter arising from the duct epithelium or from squamous metaplasia.

A recent proposed classification of lacrimal epithelial neoplasia suggested by Weis *et al*, based on their series and other recent series, also has not mentioned any incidence of adenosquamous carcinoma of the lacrimal gland.¹ Adenosquamous carcinoma of the lacrimal caruncle are found reported in a few studies,⁴ but only two cases of adenosquamous carcinoma of the lacrimal gland were found reported even on extensive searches using Google Scholar and PubMed search engines. The first case of lacrimal gland adenosquamous carcinoma was described by Forrest *et al*, at the Proceedings of the IXth International Congress of the International Society of Craniofacial Surgery, of a 6-year-old child presenting as unilateral proptosis.⁵ Another case was found in a case series of nine cases of lacrimal gland tumour biopsies, as described by Leeungurasation, where one case of lacrimal gland adenosquamous carcinoma was also found.⁶

Treatment of malignant lesions is controversial. Orbital exenteration or radical exenteration (lateral wall and roof of the orbit are removed down to the level of the dura matter along with orbital soft tissues) is done according to the tumour histotype and grade. In our case, we resorted to an orbitocranial approach for complete excision of the tumour. There is very little evidence supporting low dose postoperative radiotherapy. Histopathology has been shown to be the most significant predictor of patient survival.⁷

The possibility of the tumour being metastatic in this case is less likely because extensive investigations for the sys-

temic malignancy were unfruitful. The patient refused any destructive surgery or postoperative radiotherapy that may lead to diminution of vision. We hope that radical excision of the tumour will avoid recurrence of the tumour for which more follow-up for the patient is required.

Learning points

- Tumors of the orbit and superior orbital fissure can be removed in total by orbito-cranial approach using limited frontotemporo-orbitozygomatic approach.
- Lacrimal gland adenosquamous carcinoma is a rare entity and much discussions are required in this field for better understanding of pathogenesis and management of this disorder.

Competing interests None.

Patient consent Obtained.

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Cherian I, Shrestha SP, Panhani ML, Talwar OP, Yogi N, Rai S, Lalchan S, Fernandes C. Adenosquamous carcinoma of the lacrimal gland. *BMJ Case Reports* 2010; 10.1136/bcr.07.2010.3192, date of publication

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