Pleomorphic adenoma of orbital ectopic lacrimal gland associated with swollen optic disc

Zin May Htoon, Ko Ko Thant, Le’ Shwe Zin, Thet Naing

ABSTRACT

Introduction: Pleomorphic adenoma is the most common benign epithelial neoplasm of the lacrimal gland. This is the case report of a 63-year-old lady with pleomorphic adenoma of ectopic lacrimal gland in the superomedial part of orbit. Case Report: A 63-year-old lady presented with gradually increasing non-axial proptosis of left eye for two years duration. There was downwards and outwards displacement of left eye ball by a hard, palpable mass in the superomedial part of left orbit. The left eye showed relative afferent pupillary defect and swollen optic disc. Computerized tomography demonstrated an extraconal, well-circumscribed mass lesion occupying superomedial part of the left orbit. Complete surgical excision of tumor was performed through anterior orbitotomy. The histopathology showed an encapsulated tumor composed of nests and strands of cuboidal-shaped epithelial cells and myoepithelial cells supported by loose myxoid and hyalinized calcified stroma. The mass was diagnosed “a pleomorphic adenoma arising from ectopic lacrimal gland.” Conclusion: To our knowledge, this is the first case of extraconal orbital pleomorphic adenoma arising from ectopic lacrimal gland associated with signs of optic neuropathy.

Keywords: Ectopic lacrimal gland, Optic disc swelling, Orbital tumor, Pleomorphic adenoma

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INTRODUCTION

Ectopic lacrimal gland is a rare condition which is one of the choristomata of lacrimal gland tissue outside the lacrimal gland fossa. In the literature review, ectopic lacrimal gland is most commonly seen in bulbar conjunctiva of 115 cases, 35 cases as orbital lesion, 8 cases in eyelid and canthal region, and 11 intraocular lesions [1–5]. There was also reported of three cases of ectopic lacrimal in lacrimal sac causing nasolacrimal duct blockage, one case in intramuscular of inferior rectus muscle [6–9]. Pleomorphic adenoma is the most common benign epithelial neoplasm of the lacrimal gland. There were six reported cases of pleomorphic adenoma arising from ectopic lacrimal gland. This is the case report of a 63-year-old lady with pleomorphic adenoma of ectopic lacrimal gland in the superomedial part of orbit.
CASE REPORT

A 63-year-old lady presented with gradually increasing non-axial proptosis of left eye for two years duration. There was downwards and outwards displacement of left eye ball by a hard, palpable mass in the superomedial part of left orbit (Figure 1).

Examination of left eye showed, corrected visual acuity of counting fingers 3 feet, relative afferent pupillary defect, superior choroidal folds, and optic disc swelling. Examination of right eye was normal. Computerized tomography demonstrated an extraconal, well-circumscribed mass lesion occupying superomedial part of the left orbit 2.3×2.3×3.7 cm in greatest dimensions with some areas of calcification and mild heterogeneous contrast enhancement. There was bony excavation of roof and medial wall of left orbit and indentation of superomedial globe (Figure 2A–C).

Complete surgical excision of tumor with its capsule was performed through anterior orbitotomy. No connection between the tumor and lacrimal gland was noted intraoperatively (Figure 3).

The histopathology showed an encapsulated tumor composed of nests and strands of cuboidal-shaped epithelial cells and myoepithelial cells supported by loose myxoid and hyalinized calcified stroma. The mass was diagnosed “a pleomorphic adenoma arising from ectopic lacrimal gland” (Figure 4).

DISCUSSION

Ectopic lacrimal gland tissue is the presence of lacrimal gland tissue outside the lacrimal gland fossa excluding the accessory glands of Krause and Wolfring [5, 9–11]. Ectopic lacrimal gland tissue is found most commonly in bulbar conjunctiva. It is unusual to find in
retrobulbar region. It can also be found in caruncle, outer canthus, lower lid, and intraocular regions [10–12].

At the two months of embryonic period, basal conjunctiva cells mass give rise to lacrimal gland. These cells mass gradually developed to gain final size and position in the lacrimal gland fossa. This tissue transition was completed at three years of age. During the development, part of the lacrimal gland developed separately as ectopic lacrimal gland. Most of the ectopic lacrimal glands did not connect to main lacrimal gland [10, 12, 13].

Pleomorphic adenoma is the commonest epithelial tumor of the lacrimal gland and has a high tendency to occur in the orbital lobe of lacrimal gland and rarely occur in accessory lacrimal gland. The clinical presentation of ectopic lacrimal gland varies depending on the location of the ectopic tissue [10, 12, 13]. In literature six patients have been reported for pleomorphic adenoma arising from ectopic lacrimal gland. One pleomorphic adenoma was detected in the upper eyelid, one in the subconjunctival space of the lateral fornix, one in the sub-brow position, and three within the orbit [10–15]. Complete surgical excision with intact capsule is necessary to decrease the chance of recurrence [11, 15]. There are no reported cases of pleomorphic adenoma arising from ectopic lacrimal gland affecting the optic nerve.

In our patient, an extracanal, well-circumscribed lesion at supero-medial part of left orbit caused indentation of the left globe, choroidal fold, and optic disc swelling. The tumor was excised completely within its capsule and no connection was noted with the lacrimal gland during surgery. Histologically the mass was confirmed as pleomorphic adenoma arising from ectopic lacrimal gland.

CONCLUSION

To our knowledge, this is the first case of extracanal orbital pleomorphic adenoma arising from ectopic lacrimal gland associated with signs of optic neuropathy.

REFERENCES


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Authors declare no conflict of interest.

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