Institute of Experimental Morphology, Pathology and Anthropology with Museum
Bulgarian Anatomical Society

Acta Morphologica et Anthropologica, 30 (1-2)
Sofia ● 2023

Retrobulbar Pleomorphic Adenoma of Ectopic Lacrimal Gland – Case Report

Angelina Mollova[1,2], Iveta Hubavenska[1], Maria Stoilova[2], Emral Kyosebekirov[2,3], Maria Koleva[1,2]*

[1] Department of General and Clinical Pathology, Medical University of Plovdiv; St. George University Hospital, Plovdiv, Bulgaria
[2] Department of General and Clinical Pathology, Medical University – Plovdiv, Plovdiv, Bulgaria
[3] PhD Student, Department of Anesthesiology and Intensive Care, Medical University of Plovdiv, University Hospital "St. George", Plovdiv, Bulgaria

*Corresponding author e-mail: angelinamollovakyosebekirova@gmail.com

Retrobulbar ectopic lacrimal gland is a rare choristoma with a risk of neoplastic transformation. This report describes a case of a 56-year-old female presented with left eye proptosis, paradoxal sensation, and paresthesia on the left side of the face along with persistent headache and decreasement of vision. Magnetic resonance imaging revealed a formation in the left retrobulbar area. An excision biopsy was performed and a fragmented mass, away from the lacrimal fossa, was removed. The microscopical evaluation of the specimen showed features of pleomorphic adenoma.

Key words: retrobulbar, pleomorphic adenoma, lacrimal gland

Introduction

Choristomas are congenital lesions where histologically normal tissues with little or no growth potential are present at abnormal locations [1]. Lacrimal gland choristomas are benign lesions formed by normal lacrimal gland tissue present outside the lacrimal fossa [3]. We report a case of unilateral proptosis with diminished vision and paradoxical sensation, secondary to retrobulbar pleomorphic adenoma arising from the ectopic lacrimal gland.

Case report

A 56-year-old female complained of pain in the left eye, persistent headache, and diminished vision. The examination revealed proptosis of the left eye with paradoxal sensation in the area. Magnetic resonance found a left-situated retrobulbar mass with well-circumscribed borders. Consultation with a neurosurgeon was performed and the neurological status assessment reported amblyopia of the left eye, paresis of nervus facialis, and nervus oculomotorius on the same side with clinical signs of increased intracranial pressure. Based on these observations, the patient was admitted into neurosurgery for an operation.

There were no data for biopsy evaluation, before the operation. The formation was excised and sent for a frozen section evaluation. Grossly the specimen was fragmented with greyish-pinkish color and soft consistency. The rapid microscopical analysis suggested a wide spectrum of differential diagnoses with a final word on the permanent slide.

The biopsy specimen was fixed in formalin and embedded in paraffin, and 5-µm-thick tissue sections were used for staining with hematoxylin and eosin. Written informed consent was obtained from the patient.

Macroscopically, the specimen showed a fragmented mass measuring 1 cm in diameter, with a soft consistency. The microscopical evaluation showed tumor composed of tubules and cysts lined by epithelial cells and surrounded by myoepithelial cells with myxoid stromal component (**Figs**. **1, 2**). The full excision of the tumor can not be assessed, because of the lack of a capsule.

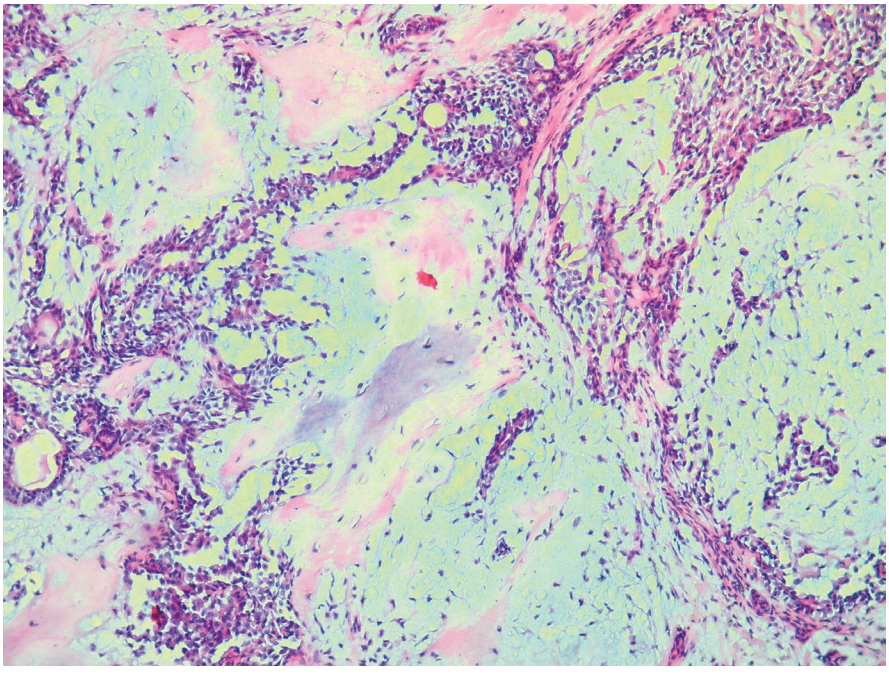

Fig. 1. Lesion characterized by tubules and cysts lined by epithelial cells and surrounded by myoepithelial cells with myxoid stromal component. HE, ×50.

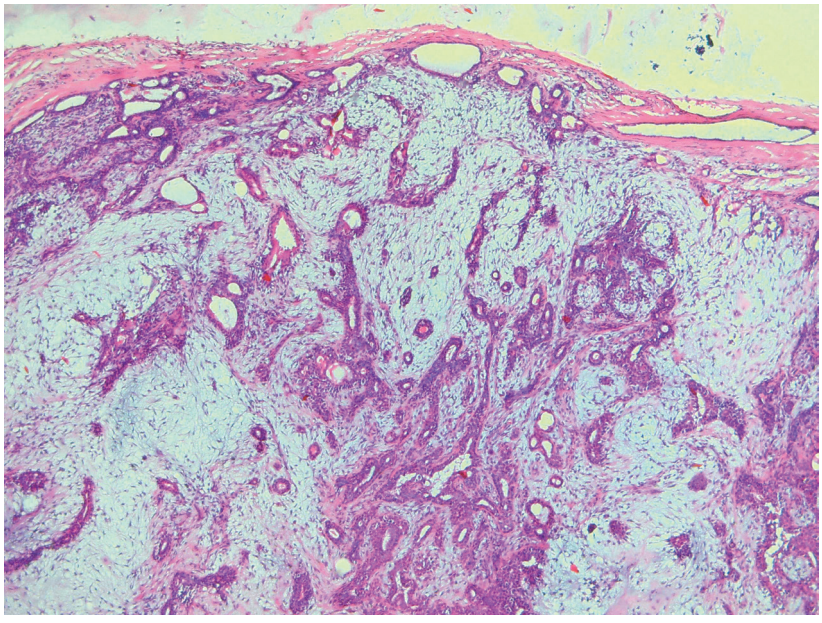

Fig. 2. Tumor, presented by tubules and cysts. HE, ×200.

Discussion

The secretory lacrimal apparatus is composed of the main lacrimal gland located in the orbital lacrimal fossa, with its palpebral lobe at the temporal side of the superior fornix, and two further sets of accessory lacrimal glands, the glands of Krause and the glands of Wolfring (or Ciaccio). Lacrimal gland tissue located at any other site is considered ectopic [2]. Ectopic lacrimal gland tissue is found most commonly in the bulbar conjunctiva and unusually in the retrobulbar region. It can also be observed in the caruncle, outer canthus, lower lid, and intraocular regions. Pleomorphic adenoma is the most common epithelial tumor of the lacrimal gland and has a high tendency to occur in the orbital lobe of the lacrimal gland and rarely occurs in the accessory lacrimal gland. In the literature, six patients have been reported for pleomorphic adenoma arising from the ectopic lacrimal gland [4]. In our patient, pleomorphic adenoma originated from an ectopic lacrimal gland located in the retrobulbar region, causing significant displacement and compression of the globe, visual dysfunction, and symptoms of increased intracranial pressure. Pleomorphic adenoma presents an excellent prognosis when the lesion is completely excised [6]. The difficulty with calling the final diagnosis on frozen section evaluation in our case came from the extensive differential diagnosis and absence of previous histopathological assessment. A wide variety of processes can produce space-occupying lesions in and around the orbit. These include benign neoplasms, malignant neoplasms, vascular lesions, inflammatory disease, congenital lesions, and infection, among other causes [5].

Conclusion

Even it is rare, retrobulbar pleomorphic adenoma arising from ectopic lacrimal gland should be considered in the differential diagnosis in retro-orbital tumors.

References

1. **Alia, M. J., D. K. Mishrab, M. N. Naika**. Ectopic lacrimal gland in a complex choristoma involving the lacrimal sac fossa. – *Saudi Journal of Ophthalmology*, **31**, 2017, 162-164.
2. **Alyahya, G. A., R. Bangsgaard, J. U. Prause, S. Heegaard.** Occurrence of lacrimal gland tissue outside the lacrimal fossa: comparison of clinical and histopathological findings. – *Acta Ophthalmol. Scand*., **83**(1), 2005, 100-103.
3. **Cruz, A. A., R. M. Limongi, E. D. Feijó, T. J. Enz**. Lacrimal gland choristomas. – *Arq. Bras. Oftalmol*., **85**(2), 2021, 190-199.
4. **Htoon, Z. M., K. K. Thant, L. S. Zin, T. Naing**. Pleomorphic adenoma of orbital ectopic lacrimal gland associated with swollen optic disc. – *J. Case Rep. Images Opthalmol*., **2,** 2019: 100011Z17ZH2019
5. **Khan, S. N., A. R. Sepahdari**. Orbital masses: CT and MRI of common vascular lesions, benign tumors, and malignancies. - *Saudi Journal of Ophthalmology*, **26** (4), 2012, 373-383.
6. **Sharma, A. K., R. Sharma, S. Misra, N. Misra, R. Ashik, N. Hussain, N. Nayak, P. R. Singh**. Pleomorphic adenoma of the orbital ectopicl acrimal gland: An extremely rare occurrence with review of literature. – *Neurol. India*, **70**(1), 2022, 296-299.