



Exophthalmos revealing a pleomorphic adenoma of the lacrimal gland

Z.Hazil, N.Tebay, I.Hasnaoui A.Krichen, S.Hassina, Y.Akannou, L.Serghini, Z.Hajji, B.Ouazzani, E.abdellah¹

Department of Ophthalmology B,
Rabat Specialty Hospital, CHU ibn Sina,
Mohammed V Souissi University Rabat, Morocco

Abstract: Pleomorphic adenoma is a heterogeneous benign tumor of the salivary glands, composed of several epithelial and myoepithelial tissue elements. It frequently occurs in the main salivary glands, but exceptionally in other sites, particularly the orbital cavity. We report a clinical case of pleomorphic adenoma of the orbital cavity.

Index Terms - exophthalmos, lacrimal gland, pleomorphic adenoma.

I. INTRODUCTION

Introduction:

Epithelial tumors of the lacrimal gland account for 40-50% of all lacrimal gland lesions, 50% of which are malignant. Other lesions are non-epithelial tumors such as lymphomas, dermoid cysts, metastases, inflammatory pseudotumors or dacryops.

Epithelial tumours of the lacrimal gland include :

- Benign tumours: pleiomorphic adenomas, oncocytomas.
- Malignant tumors: mixed malignant tumors, adenoid cystic carcinomas, adenocarcinomas, mucoepidermoid carcinomas;
- More rarely: squamous cell carcinomas, acinar cell carcinomas, myoepitheliomas, malignant oncocytomas.

Clinical case:

This is a 65-year-old female patient with no previous pathological history of any kind, who presented with a palpebral swelling over her left eye that had been present for over a year.

Clinical examination revealed a solid-looking, painless, irreducible palpebral mass, with no inflammatory signs opposite, pushing the globe downwards and inwards, causing exophthalmos and partial ptosis of the left eye (Figure 1). Oculomotor examination revealed limited abduction, and the rest of the examination was unremarkable.

Magnetic resonance imaging showed a left orbital expansive process with heterogeneous tissue signal, a fleshy component with discrete T2, FLAIR, diffusion hypersignal signal, intensely enhanced after injection of contrast medium, and a cystic component with T2 hypersignal, diffusion hyposignal signal, peripherally enhanced after injection, measuring 17*34*23mm long axis, located in the superior-external quadrant of the orbit at the expense of the lacrimal gland, coming into contact with the superior rectus and lateral rectus muscles, which are suppressed, without bone lysis of the opposite orbital wall, suggestive of a pleomorphic adenoma (Figure 2).

Anatomopathological study after surgical excision confirmed the diagnosis.

Discussion:

Pleomorphic adenoma is a frequent tumor of the salivary glands, generally developing at the expense of the parotid gland in 80% of cases [1]. Its intra-orbital location is rare, accounting for only 3-5% of cases [2].

According to Garrity, the average age at diagnosis is 42 (range 16 to 81). Exceptional pediatric forms have been reported.

Clinical features: exophthalmos is always unilateral, painless, irreducible and slow to progress over several years. The eyeball is pushed inwards and downwards, sometimes causing vertical diplopia. Partial ptosis is a classic sign. As the tumor progresses, a mobile mass may be found under the skin in the superior-external angle of the orbit, with or without local edema.

Reduced visual acuity due to astigmatism or acquired hyperopia, lagophthalmos due to keratitis, visual field impairment due to posterior pole compression, scotomas or fundus abnormalities are also possible.

This slowly progressing tumor is generally painless; this sign appears much more frequently in malignant tumors. Nevertheless, atypical forms including inflammatory presentation, painful subcutaneous nodule, calcification and bone erosion have been reported.

cerebro-orbital scanner is sufficient for diagnosis, and is superior to magnetic resonance imaging in its analysis of bone wall changes. Treatment is based on total removal of the lesion and its capsule, to prevent local or regional recurrence and malignant transformation [3, 4]. The appearance of the tumor was typically that of a pleomorphic adenoma. It comprised the two usual components of this type of tumor: on the one hand, a glandular component with cords and tubes or other cavities filled with a mucoid substance, and on the other, a connective tissue component of variable density, here without a chondroid territory. The lesion was entirely encapsulated, but this capsule split to surround an aberrant tumor island.

Conclusion:

Pleomorphic adenoma is a benign tumor whose surgical excision must be total, encompassing the capsule and imposing an approach allowing en bloc removal. Clinical and radiological monitoring for several years should be systematic.

Figures



Figure1: photo showing exophthalmos with ptosis on an upper external palpebral mass pushing the globe inwards and downwards

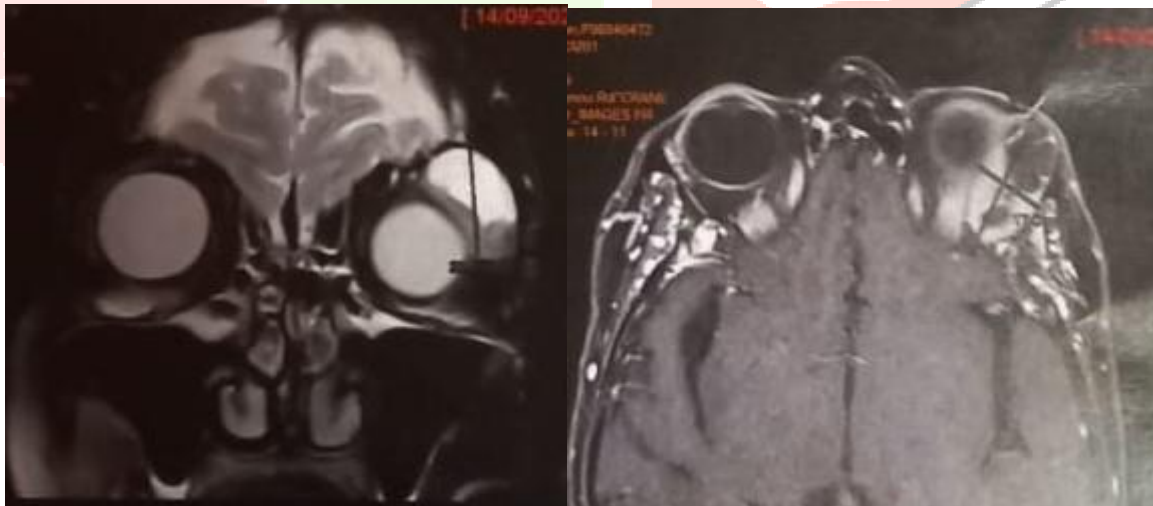


Figure2: magnetic resonance imaging of a tumoral process in the left lacrimal gland suggestive of a pleomorphic adenoma

Conflicts of interest:

All authors declare that they have no conflicts of interest.

References:

- 1] MOURTADA FAISSAL, A. SAKALI LEILA, EL KOHEN ASMA, JAMAL HICHAM, Adénome pléomorphe géant des fosses Nasales. Fr ORL 2007, 92, 309 —11 .
- 2] M. HALHAL, A. MEYER, X. MOREL, C. COCHARD-MARIANOWSKI, F. BEHARCOHEN, A. BLANQUET, J. COPHIGNON, Adénome pléomorphe simulant une dacryo-adénite A propos d'une observation anatomo-clinique. Journal Français d'Ophtalmologie 2004, 27, 10, 1200-1203.
- 3] VAGEFI MR, HONG JE, ZWICK OM, BEDROSSIAN EH JR, SEIFF SR, Atypical presentations of pleomorphic adenoma of the lacrimal gland. Ophthal Plast Reconstr Surg. 2007, 23, 4, 272-4 .
- 4] MENSINK HW, MOOY CM, PARIDAENS D. In situ denocarcinoma ex pleomorphic adenoma of the lacrimal gland. Clin Experiment Ophthalmol. 2005, 33, 6, 669-71.

