Importance of correlating radiohistopathologic features in lacrimal gland pleomorphic adenoma

Lacrimal gland pleomorphic adenoma (LGPA) is a common benign epithelial tumour of the lacrimal gland that frequently appears on imaging as a well-circumscribed lesion with no bony erosion.[1] In some cases, it can present with atypical radiologic features and then needs to be correlated with histopathologic findings to ascertain a diagnosis.

We present a case of LGPA with cystic changes associated with coagulative necrosis. A 71-year-old asymptomatic man was noted to have incidental left proptosis during diabetic retinopathy screening. Clinically, his best-corrected visual acuity was 6/4.5 OD and 6/45 OS eye. There was no relative afferent pupillary defect, and colour vision was preserved. The patient had a palpable left superotemporal mass. There was 5 mm of nonaxial proptosis and 4 mm of inferior dystopia (see Supplementary Fig. 1, available online). The patient had a limitation of left abduction and upgaze (−1) without diplopia. Moreover, he had a left hemi−retinal vein occlusion with macular edema secondary to diabetic retinopathy, which accounted for the reduced left vision and lack of diplopia. The patient was referred to the retinal team for further management of the retinal findings.

Magnetic resonance imaging with gadolinium demonstrated a well-defined mass (22 × 19 × 19 mm) in the left superotemporal extraconal space showing T_1 isointensity and mixed T_2 intensity (Fig. 1A). The mass showed heterogeneous contrast enhancement centrally with prominent peripheral contrast enhancement (Fig. 1B).

The patient underwent an excisional biopsy. Histopathology showed a circumscribed LGPA with epithelial and myoepithelial populations accompanied by chondromyxoid stroma (Fig. 2A). The lesion was centrally acellular and hyalinized, with a region showing coagulative-type necrosis. The necrotic area had regions of hemorrhage and inflammatory cells without any tumour cells (Fig. 2B). There was no evidence of perineural or vascular invasion. PLAG1 fluorescence in situ hybridization testing was negative. The histopathology was reviewed by 2 senior consultant histopathologists, who confirmed the diagnosis of LGPA.

On magnetic resonance imaging, most LGPAs are isointense (97%) on T_1-weighted images and hyperintense (86%) on T_2-weighted images.[2] An enhancing rim can be seen on T_1 fat-suppressed postcontrast images in 27% of LGPAs, with cystic spaces reported less frequently.[3] Nonenhancing cystic lesions that are bright on T_2-images, as seen in our patient, can be found in up to 19% LGPAs.[3] They may be consistent with a cyst filled with serous, hemorrhagic, or myxomatous findings on histopathology.[3] It is important to correlate both findings because variable materials on histology may explain the mixture of T_2 intensities on radiology.[3]

Necrosis in LGPA is uncommonly reported, and although it can be associated with malignancy, this finding must not be used alone as a sign of malignant change.[3] Necrosis in these benign lesions may be secondary to iatrogenic causes such as fine-needle aspiration or incisional biopsy. In the absence of previous surgery, necrosis also may occur spontaneously owing to the growing tumour itself exerting a mass effect on the surrounding tissue or interrupting the vascular supply, causing infarction or ischemia.[4] Thus, correspondence with the histopathologist is required to request examination of the lesion at all levels to exclude malignant cells.

Where there is histologic uncertainty, PLAG1 testing can be used in conjunction with the histopathology to distinguish LGPAs from other malignant lacrimal gland tumours. However, PLAG1 fluorescence in situ hybridization testing can be negative in up to 10% of LGPAs, as seen in our patient.[5,6] No evidence of adenocarcinoma was found on careful sectioning through our specimen, ruling out the

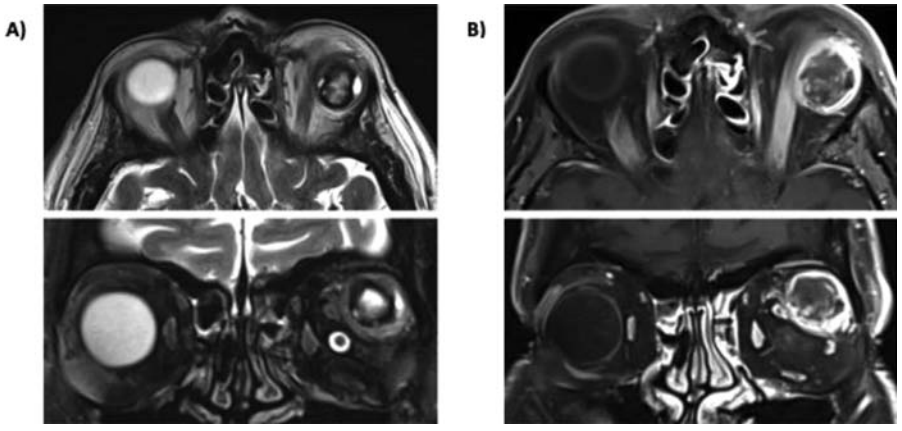

Fig. 1—Radiologic findings of the LGPA with coagulative necrosis. (A) Magnetic resonance imaging axial (top) and coronal (bottom) T_2 images show the orbital mass in the left superolateral extraconal space. The lesion is heterogeneous with mixed regions of peripheral hypointensity and central hyperintensity. (B) Magnetic resonance imaging axial (top) and coronal (bottom) T_1, fat-suppressed, postcontrast images show prominent peripheral contrast enhancement with heterogeneous internal enhancement

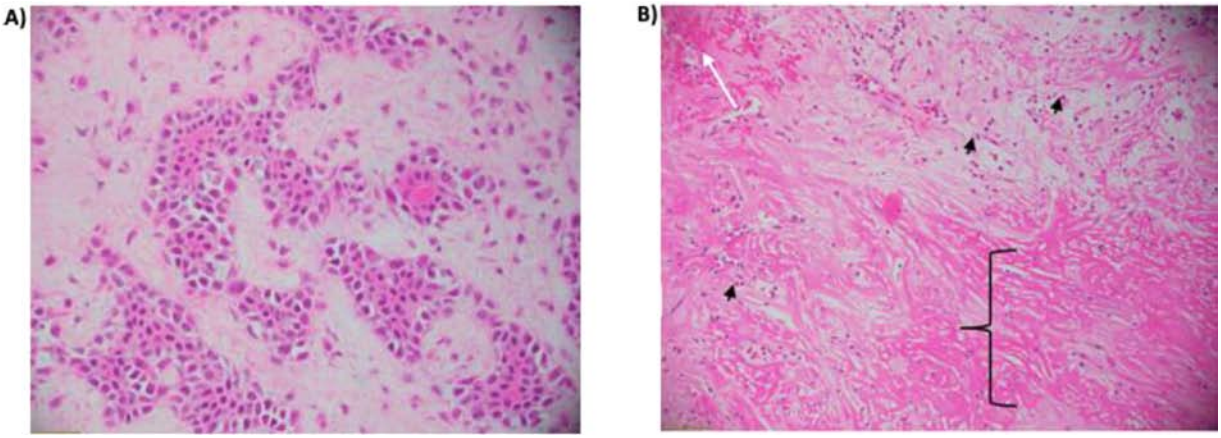

Fig. 2—Histologic sections of the LGPA. (A) Section of pleomorphic adenoma (high-power field, × 40 magnification) displaying epithelial and myoepithelial populations suspended in a myxoid stromal matrix giving the appearance of a biphasic tumour. (B) Section of coagulative necrosis (high-power field, × 20 magnification) showing hyalinization area (black bracket) and areas of hemorrhage (white arrow) and monocytes (black arrows).

diagnosis of carcinoma ex pleomorphic adenoma. The incidental presentation of our case is in keeping with the benign diagnosis.

In summary, we present a patient with cystic LGPA showing coagulative necrosis, further adding to the growing evidence that coagulative necrosis and cystic changes are not always associated with malignancy and that careful correlation of radiohistopathologic features is crucial.

Supplementary Materials

Supplementary material associated with this article can be found in the online version at doi:10.1016/j.jcjo.2021.09.020.

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Originally received Jul. 31, 2021. Final revision Sep. 21, 2021. Accepted Sep. 27, 2021.

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References

1. Bernardini FP, Devoto MH, Croxatto JO. Epithelial tumors of the lacrimal gland: an update. Curr Opin Ophthalmol 2008;9:409–13.
2. Young SM, Kim Y-D, Shin HJ, Imagawa Y, Lang SS, Woo KI. Lacrimal gland pleomorphic adenoma and malignant epithelial tumours: clinical and imaging differences. Br J Ophthalmol 2019;103:264–8.
3. Watanabe A, Andrew NH, Ueda K, et al. Clinico-radiological features of primary lacrimal gland pleomorphic adenoma: an analysis of 37 cases. Jpn J Ophthalmol 2016;60:286–93.
4. Allen CM, Damm D, Neville B, Rodu B, Page D, Weathers DR. Necrosis in benign salivary gland neoplasms: not necessarily a sign of malignant transformation. Oral Surg Oral Med Oral Pathol 1994;78:455–61.
5. Martins C, Fonseca I, Roque L, et al. PLAG1 gene alterations in salivary gland pleomorphic adenoma and carcinoma ex-pleomorphic adenoma: a combined study using chromosome banding, in situ hybridization and immunocytochemistry. Mod Pathol 2005;18:1048–55.
6. Andreasen S, von Holstein SL, Homøe P, Heegaard S. Recurrent rearrangements of the PLAG1 and HMGA2 genes in lacrimal gland pleomorphic adenoma and carcinoma ex pleomorphic adenoma. Acta Ophthalmol 2018;96:e768–71.

Footnotes and Disclosure

The authors have no proprietary or commercial interest in any materials discussed in this correspondence.

Treatment of an orbital pseudomeningocele through an eyelid incision

Pseudomeningoceles (PMs) occur from extravasation of cerebrospinal fluid (CSF) into soft tissue secondary to a dural tear.[1–3] Unlike a true meningocele, which is lined by arachnoid tissue, PMs are associated with the formation of a fibrous capsule.[2] Although PMs can form due to surgical insults to the dural covering of the brain or spine, they can also occur secondary to trauma or congenital abnormalities.[1,2] Clinical symptoms usually arise when there is mass effect on the