

Adenocarcinoma of lacrimal gland

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Section: Head & neck imaging

Area of Interest: Head and neck

Procedure: Education

Imaging Technique: CT

Imaging Technique: MR

Imaging Technique: MR-Diffusion/Perfusion

Special Focus: Pathology Case Type: Clinical Cases

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Patient: 41 years, female

Clinical History:

A 41-year-old female patient presented to the hospital with a one year history of migrainous headaches, getting progressively worse. These headaches were intermittent during the day with pain located on the right side, sometimes sharp, sometimes pressure behind the right eye. No clinical signs of an orbital mass were found.

Imaging Findings:

Contrast CT head images showed a rounded extraconal lateral orbital mass centred on the right lacrimal gland with strong peripheral and mild central heterogeneous enhancement. No bony changes were present (Fig. 1).

Orbital MRI showed a T1W hypointense and T2W hyperintense mass with heterogeneous enhancement, and selective restricted diffusion of the peripherally enhancing component (Fig. 2 and 3).

Excisional biopsy revealed primary ductal adenocarcinoma of the lacrimal gland.

Discussion:

The lacrimal glands occupy the superior temporal quadrant of the orbit, adjacent to superior and lateral recti [1]. Since they are usually symmetric, organ asymmetry is an important indicator of disease. Normal lacrimal glands enhance to match the MR signal of extraocular muscles, a useful guide to judge if the degree of enhancement is excessive or not.

Although they are anatomically related to the orbits, lacrimal glands are embryologically, functionally, and pathologically similar to major salivary glands. In consequence, the World Health Organization (WHO) classification of lacrimal gland tumours has been adapted from salivary pathology. Because they contain epithelial and lymphoid cells, they are affected by infectious, infiltrative, inflammatory and neoplastic pathologies of these cell lines. About 50% of lacrimal gland masses are inflammatory, 25% are salivary type tumours and 25% are lymphoid lesions or lymphoma [2].

Mucosa-associated lymphoid tissue type (MALT) lymphomas, also known as eye-associated lymphoid tissue (EALT), are the commonest form of lymphoma affecting lacrimal glands. On DWI, lymphoma demonstrates lower mean ADC values ($< 0.7 \times 10^3 \text{ mm}^2/\text{sec}$) than other less cellular tumours such as adenoid cyst carcinoma, which may help differentiate them.

They clinically present as unilateral gland enlargement with globe displacement. Epithelial tumours arise mainly from

the orbital lobe (posterior), with < 20% involving the palpebral lobe (anterior). 50% are benign and 50% malignant. Benign tumours include pleomorphic adenoma and oncocytoma, while malignant tumours include adenoid cystic carcinoma, mucoepidermoid carcinoma, and adenocarcinoma.

Primary ductal adenocarcinoma of the lacrimal gland (PDACL) is rare [3, 4], with few cases reported in the literature since first described in 1996 [3]. They are characterized by their aggressive nature and poor prognosis [5, 6].

PDACL can be classified into low- and high-grade, being histologically and immunohistochemically similar to highly aggressive salivary ductal carcinoma [4]. The lacrimal gland is composed of lobular acini and ducts, similar to the major salivary glands [5].

PDACL is treated with complete gland excision, lymph node dissection and adjuvant radiotherapy. Its long-term prognosis and response to various treatment regimens is hampered by the few reported cases in the literature [6, 7].

Orbital MRI is the diagnostic method of choice for visualization and evaluation of lacrimal gland tumours. Its superior soft tissue resolution permits better assessment of glandular, peri-glandular and perineural tumour spread. CT is mainly used to discern the presence of bone erosion. PET/CT is used for more sensitive detection of distant metastatic disease.

Differential Diagnosis List: Primary adenocarcinoma of lacrimal gland (low grade), Lacrimal gland sarcoidosis, Orbital inflammatory pseudotumour, Benign reactive lymphoid hyperplasia, Lacrimal gland pleomorphic adenoma, Lacrimal gland oncocytoma, Lacrimal gland adenoid cystic carcinoma, Lacrimal gland malignant lymphoma

Final Diagnosis: Primary adenocarcinoma of lacrimal gland (low grade)

References:

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Figure 1

a



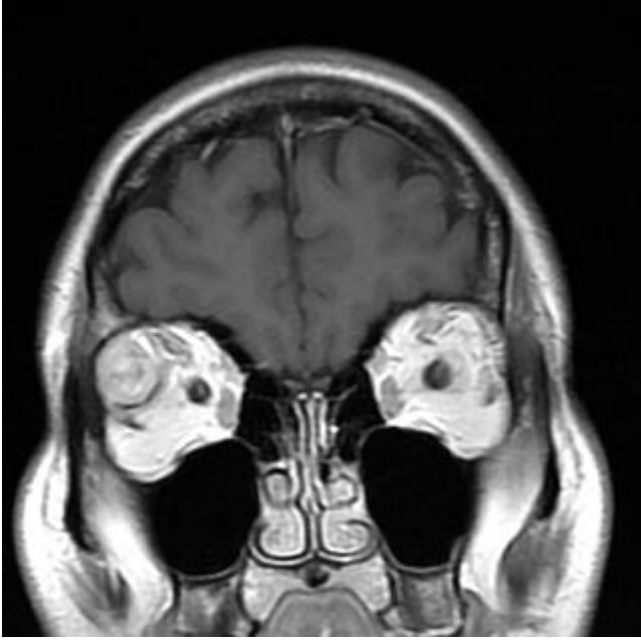
Description: T1W coronal MRI shows an intermediate intensity right lacrimal fossa mass. **Origin:** Augusta University

b



Description: T2W coronal MRI shows a hyperintense right lacrimal fossa mass. **Origin:** Augusta University

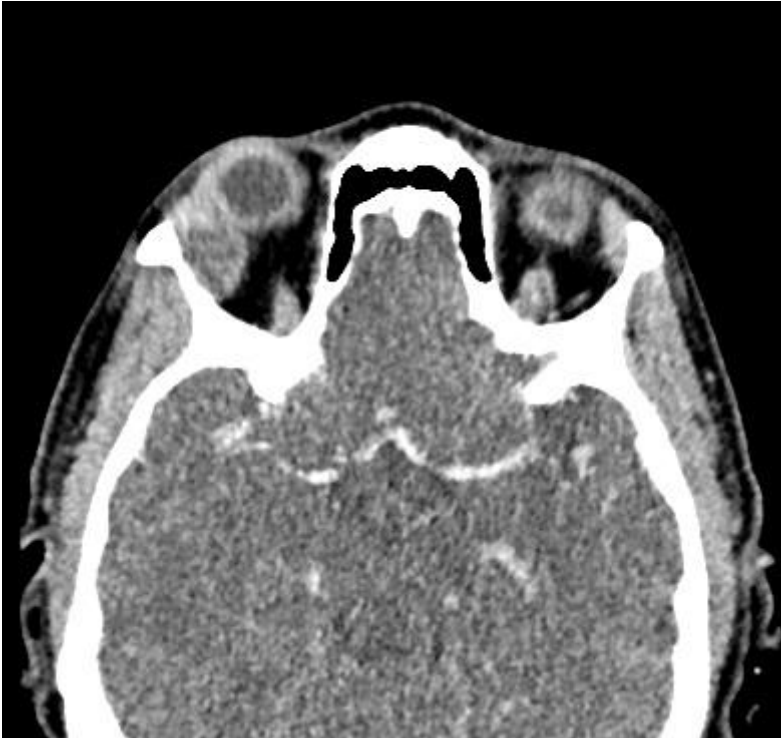
c



Description: Post-contrast T1W coronal MRI shows a right lacrimal fossa mass with heterogeneous post-gadolinium enhancement. **Origin:** Augusta University

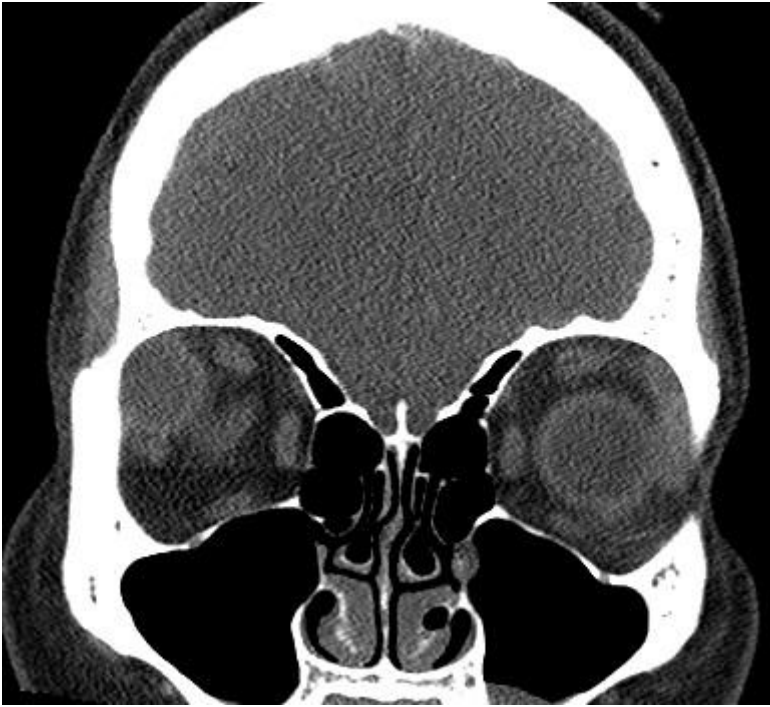
Figure 2

a



Description: Images show a rounded extraconal mass centred on the right lacrimal gland at the superolateral aspect of the right orbit, with strong peripheral and mild central heterogeneous enhancement. **Origin:** Augusta University

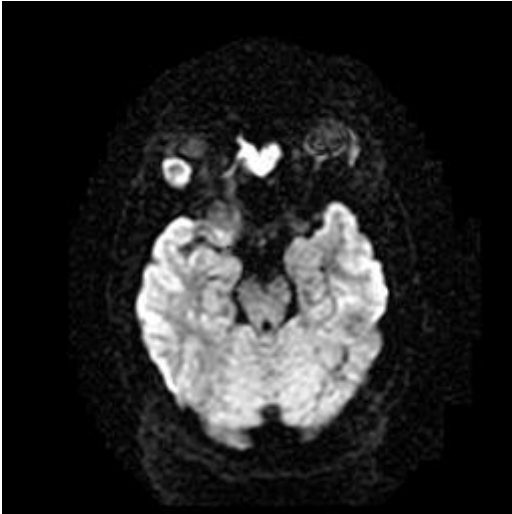
b



Description: Images show a rounded extraconal mass centered on the right lacrimal gland along the superolateral aspect of the right orbit, with strong peripheral and mild central heterogeneous enhancement. **Origin:** Augusta University

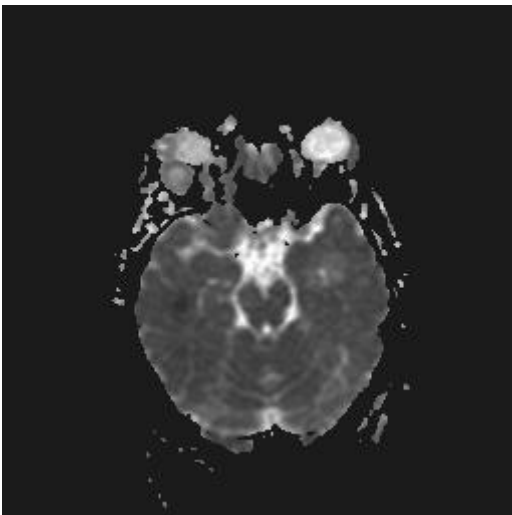
Figure 3

a



Description: B1000 diffusion weighted image (DWI) showing partially hyperintense right lacrimal fossa lesion. **Origin:** Augusta University

b



Description: Apparent diffusion coefficient (ADC) map showing areas of reduced signal within the right lacrimal fossa lesion, consistent with restricted diffusion. **Origin:** Augusta University