

Primary lacrimal gland lymphoma.

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Abstract

The authors describe a relatively rare case of a 56-year-old female patient with a two-year history of painless swelling of the outer half of the upper eyelid and small exophthalmos. MRI scan showed a homogeneous tumour swelling of the lacrimal gland, which had the same signal density as ocular muscles on T2-weighted images. After lateral orbitotomy with the removal of the entire tumour, histological examination showed an extranodal marginal zone lymphoma of mucosa-associated lymphoid tissue (MALT lymphoma) affecting the lacrimal gland. After comprehensive oncological, internist, radiological and haematological examinations, findings were concluded as a primary lymphoma of the lacrimal gland. Orbital and systemic findings remain normal one year after the surgery.

Conclusion: At present, it is necessary to consider lymphoma in the case of lacrimal gland tumours. When the diagnosis is uncertain, it is recommended that a complete removal of the tumour is performed and, based on the histological results, that further diagnostic and therapeutic measures are continued.

Keywords: primary lacrimal gland lymphoma, pleomorphic adenoma, MRI

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Introduction

The incidence of lacrimal gland lymphomas in recent decades has increased rapidly. [8] In our work where we followed 100 tumours and pseudotumours of the orbit in the period 1982 – 1994, 6 were lacrimal gland tumours. Adenoid cystic carcinoma 3x, pleomorphic adenoma 2x, solitary fibrous tumour of the lacrimal gland with apparent hemangiopericytoma 1x, but no lymphoma [5].

In an epidemiological study from 2014, Andreoli *et al.* described 702 malignant tumours of the lacrimal gland. Of these, lymphomas formed the largest part (58 %).



Adenoid cystic carcinoma, adenocarcinoma and mucoepidermoid carcinoma accounted for 13.4 %, 3.8 % and 3.6 % respectively. The last three types have poor prognosis. Lymphomas, on the contrary, respond well to treatment and survival is even longer [1].

Lacrimal gland lymphomas are relatively rare, representing 7 % to 26 % of ocular adnexal lymphomas [8].

The most frequent lymphoma subtype was extranodal marginal zone B cell lymphoma (MALT [mucosa-associated lymphoid tissue] lymphoma, 55.5 % [8].

Rasmussen *et al.*, who described 27 lacrimal gland lymphomas in the period of 1 January 1975 to 31 December 2009, determined its primary occurrence in 44 % of cases. In 56 % of cases, the lacrimal gland was affected by secondary infiltration. Histologically, MALT was the most frequent lymphoma, occurring in 37 % of cases [7].

Because this type of primary tumour is not so frequent [9] or easy to diagnose, we would like to present the following case report.

Case report

The 56-year-old and otherwise healthy woman noticed a swelling of the outer half of the left upper eyelid in mid-2012. In February 2014, she came to the office of the first author, where a stiff and painless mass localised in the outer half of the upper eyelid and orbit was found on palpation. Prominence of the left eye was 20-110-23. Eyeball motility was not altered and diplopia was not identified. Ophthalmological findings were normal bilaterally. Visual Acuity: RE = 1.0, LE = 1.0 (-1 dioptre). Figure 1.





Figure 1. Swelling of the outer half of the upper eyelid with a two-year history

The patient was referred for MRI of the orbit, which showed a homogeneous tumour expansion of the left lacrimal gland, which had signal density identical to ocular muscles. Figure 2.

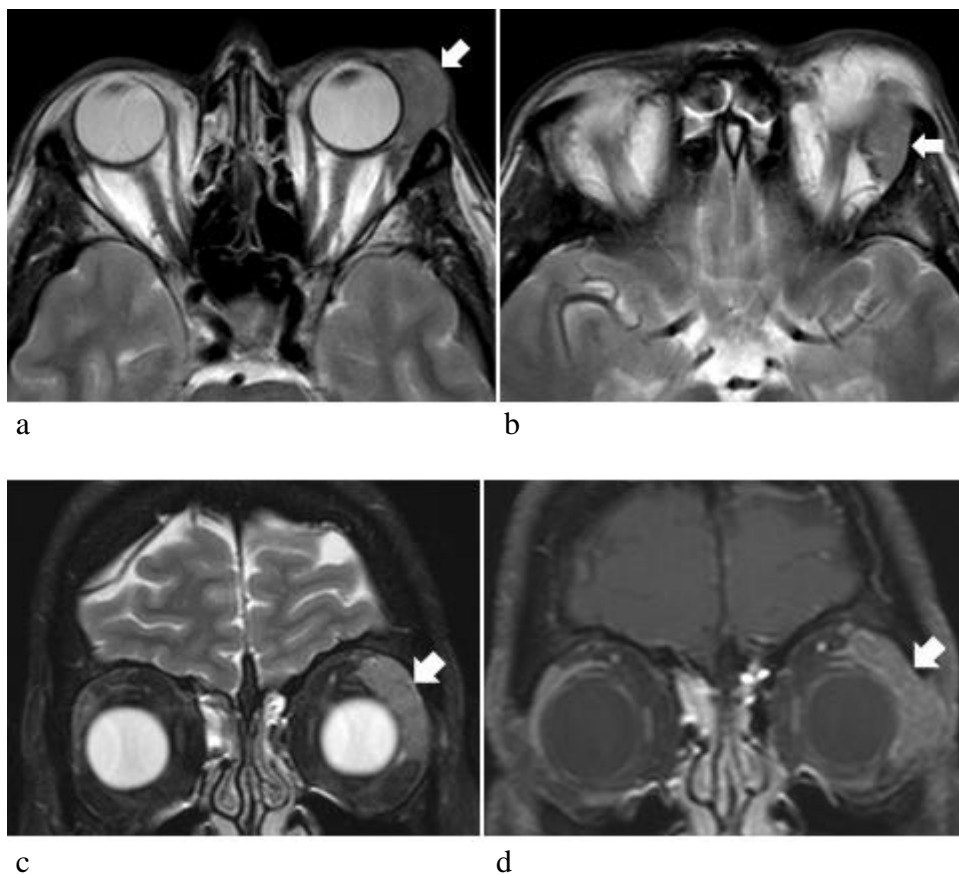


Figure 2. Axial T2-weighted images in the plane of the optic nerve (a) and in the plane of the superior rectus muscle (b). Arrows indicate the homogeneous tumour



expansion of the left lacrimal gland which has signal density equal to that of ocular muscles, does not oppress the left eyeball and does not infiltrate bones of the orbit. Coronal T2-weighted images (c), and contrast-enhanced T1-weighted coronal images (d), both with suppression of adipose tissue signal.

Arrows indicate homogeneous tumour expansion of left lacrimal gland, which does not infiltrate extraocular muscles, does not oppress the left eyeball, does not infiltrate bones of the orbit and shows moderately significantly homogeneous contrast-enhanced saturation (d).



a

b



Figure 3. Intraoperative findings of lymphoma (a) and its size after the extirpation (b).

Based on performed examinations, findings were classified as suspected pleomorphic adenoma of the left lacrimal gland. In May 2014, under general anaesthesia, a complete extirpation of the tumour of the lacrimal gland with the size 14x8 mm was performed through lateral orbitotomy. During the surgery, function of levator palpebrae muscle was monitored. The surgery was uncomplicated. Figures 4, 5.

Histologically, there was a lymphoma with a nodular arrangement. The variably sized tumour nodules were usually centred on residual reactive lymphoid follicles. Focally there were pale tumour infiltrates with either perifollicular or band-like arrangement corresponding to marginal zone pattern of growth (Fig. 4). The tumour was composed of predominantly small lymphocytes, sometimes with centrocytoid nuclear features, without conspicuous plasmacytoid differentiation or significant admixture of large transformed cells (Fig. 5). Mitotical activity was low. In the tumour infiltrate there were residual ductal structures of the lacrimal gland, no typical lymphoepithelial lesion was found (Fig. 6). Immunohistochemically, tumour cells strongly expressed CD20 (Fig. 6) and BCL2, while CD10, CD5 and cyclin D1 were negative. CD23 was positive on organoid follicular dendritic cells highlighting lymphoid follicles effaced by colonisation by tumour cells. The proliferation activity assessed by Ki67 expression was low, approximately 5–10 % of tumour cells. Follicular lymphoma, mantle cell lymphoma, and small lymphocytic B-cell lymphoma were excluded immunohistochemically and the histological picture was consistent with a diagnosis of an extranodal marginal zone lymphoma of mucosa-associated lymphoid tissue (MALT lymphoma) affecting the lacrimal gland.



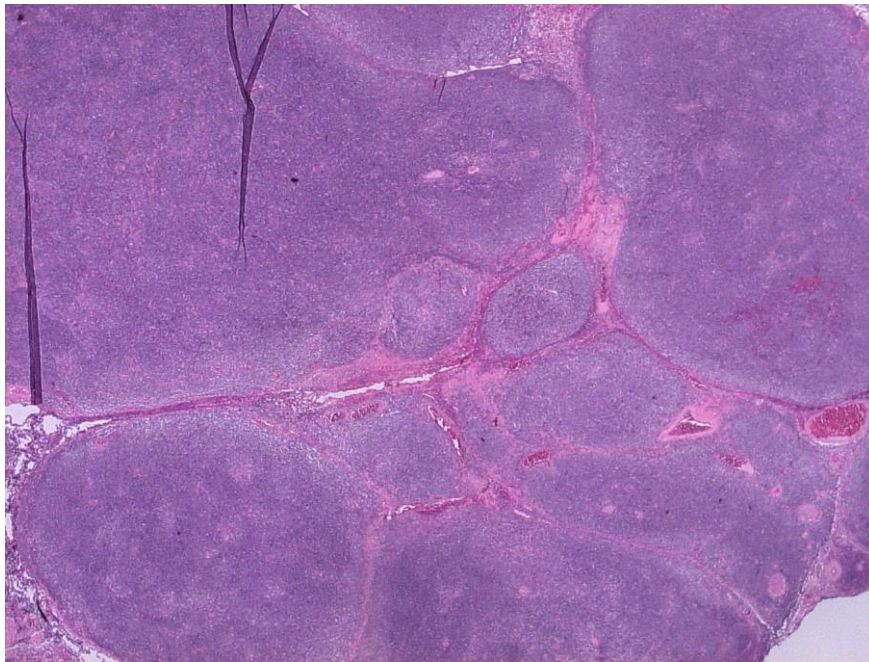


Figure 4. Nodular infiltrate of lymphoma with a marginal zone pattern of growth (HE, original magnification 20x).

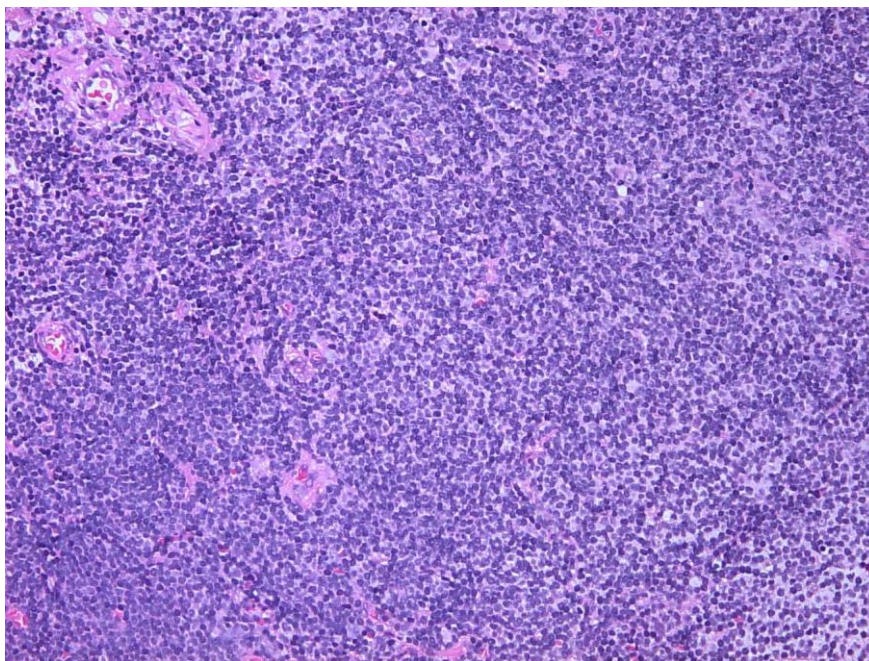


Figure 5. The tumour was composed of small lymphocytes sometimes with centrocytoid nuclear features (HE, original magnification 200x).



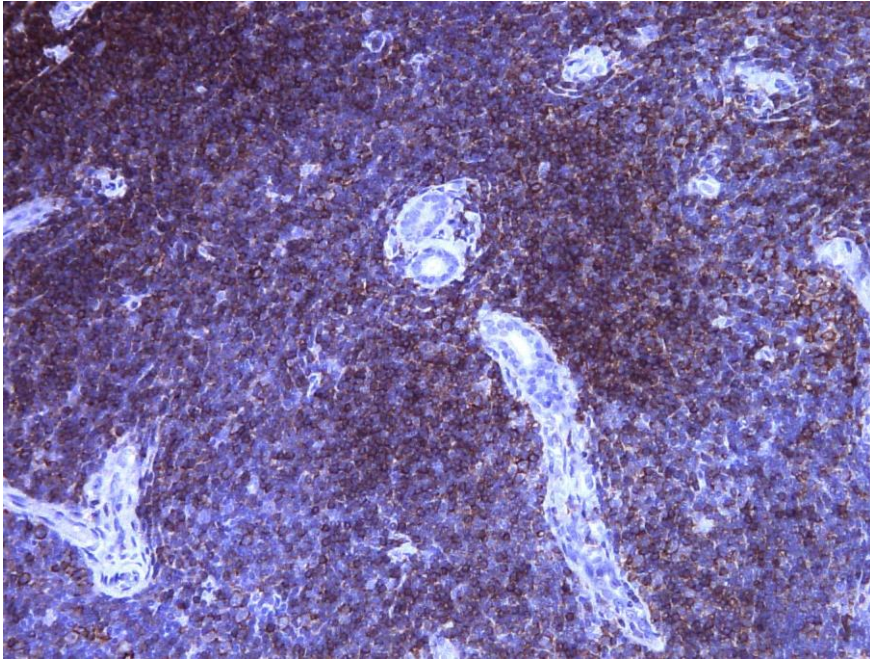


Figure 6. Strong membranous positivity of CD20 on tumour cells. There were no signs of epitheliotropism of tumour cells on residual ductal structures (immunohistochemistry, original magnification 200x).

Gastroscopy, including gastric biopsy, showed the presence of *Helicobacter pylori*. Colonoscopy was negative.

Complete haematological, biochemical and immunological examinations, bone marrow biopsy and CT of bones performed one year after the surgery showed no abnormalities.

Follow-up MRI performed 12 months after the surgery did not show any residual tumour or recurrence of the solid tumour of the lacrimal gland. Figure 7.



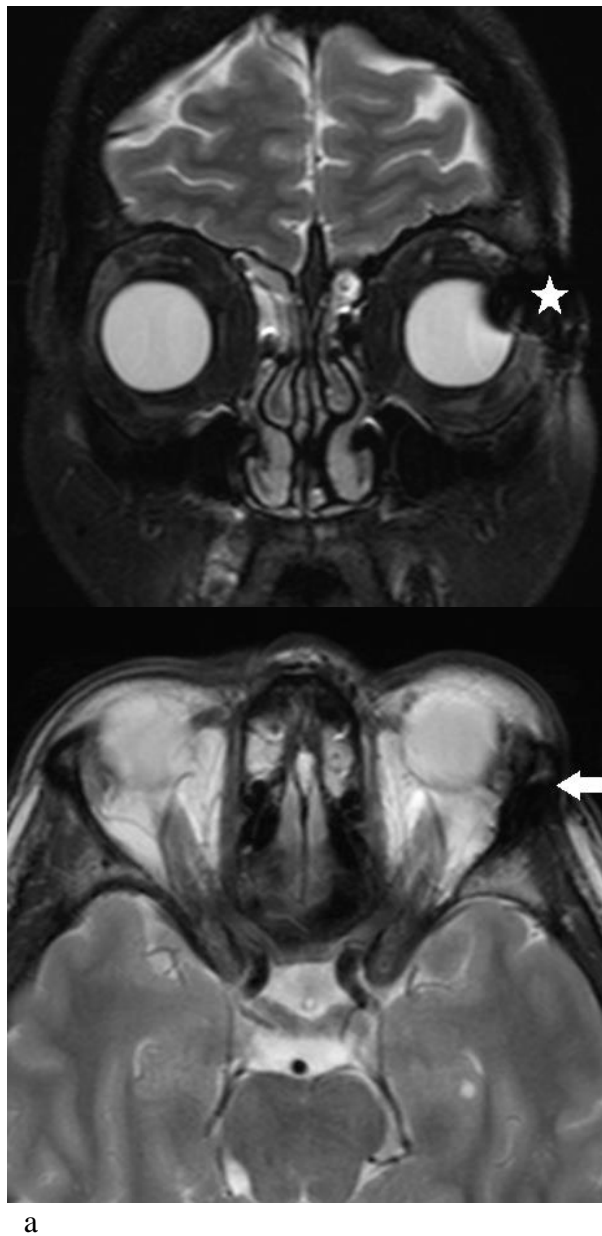


Figure 7. Coronal T2-weighted image 12 months after tumour resection (a), with suppression of adipose tissue signal. Metallic material artefacts after lateral orbitotomy are apparent after the surgery (*).

Axial T2-weighted image in the plane of the superior rectus extraocular muscle (b). The arrow indicates the presence of metallic artefacts of osteosynthetic material after lateral orbitotomy.

Discussion



Although most orbital lymphoid tumours are characterised by a slow, painless onset and a mass that moulds to orbital structures, different presentations may occur. Intensity on T2-weighted MRI is a possible means for differentiating lymphoid tumours (hyperintense). Using MRI, Polito *et al.* described 95 primary and secondary lymphoid tumours of the orbit and found that on T2-weighted MRI, only 35 % of lymphoid tumours were hyperintense [6].

In newer publications by Gunudz *et al.*, 3 lymphoid tumours, and 7 epithelial tumours including pleomorphic adenoma were examined using MRI. The patients with lymphoid tumours demonstrated involvement of the orbital lobe, a moulded configuration with ill-defined margins and sharp angles, lack of bone change, an isointense internal signal on T1-weighted images, an isointense signal on T2-weighted images, and moderate contrast enhancement. All epithelial tumours demonstrated an isointense internal signal on T1-weighted images, a hyperintense signal on T2-weighted images, and moderate contrast enhancement [4].

Recent literature has indicated that orbital lymphoma appeared to be isointense compared with extraocular muscles on T1-weighted and T2-weighted images [3, 10, 11].

With respect to the course of disease of our patient, which was not accompanied by pain and the lymphoma infiltrated only the lacrimal gland, there was not a 100 % certainty that the patient was not suffering from pleomorphic adenoma. Similarly, with respect to the contradiction in the literature regarding MRI evaluation of both possible diagnoses, fine needle biopsy was not performed because of possible malignant transformation of the pleomorphic adenoma or haematological spread of the malignant tumour of the lacrimal gland. Complete surgical removal ensured not only the removal of the tumour itself but also the possibility of accurate diagnosis by histological examination.

Conclusion

At present, it is necessary to consider lymphoma in all cases of lacrimal gland tumours. Whenever the diagnosis is uncertain, it is recommended that complete removal of the tumour is performed and, based on histological results, that further diagnostic and therapeutic algorithms are selected.

Acknowledgments

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Conflicts of Interest

No potential conflict of interest relevant to this article was reported.



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