

CASE REPORT

Open Access



CT and MRI findings in relapsing primary malignant melanoma of the lacrimal sac: a case report and brief literature review

Ju-Wei Shao^{1†}, Jian-Hua Yin^{2†}, Shu-Tian Xiang¹, Qian He¹, Hong Zhou³ and Wei Su^{1*}

Abstract

Background: Primary lacrimal sac melanoma is an extremely rare condition with fewer than 50 cases reported so far. Clinically, its symptoms resemble those of dacryocystitis, leading to frequent misdiagnosis. During diagnosis, imaging examination is often performed to differentiate tumors from inflammation. In this report, we present a case of primary lacrimal sac melanoma and summarize the CT and MRI characteristics of lacrimal sac melanoma.

Case presentation: We report a 50-year-old female patient who had undergone a dacryocystectomy for the left lacrimal sac mass. Postoperative pathological examination confirmed the presence of primary malignant melanoma. Three months later, a lump in the lacrimal sac area was found. The patient underwent CT and MR examinations. CT scan demonstrated a partially well-defined soft mass in the fossa of left lacrimal sac extending into the nasolacrimal duct and anterior ethmoid sinus. MRI revealed an intermediate signal intensity on T1 and iso- or hyper-signal on T2 weighted images. Histopathological examination on biopsy confirmed recurrence of primary lacrimal sac melanoma.

Discussion and conclusions: None has described the CT and MR findings of primary lacrimal sac melanoma so far. Typically, MR images show hyperintensity signal on T1-weighted images and hypointense signal on T2-weighted images owing to the paramagnetic properties of melanin. In contrast to previous reports and the present case, most cases do not present these typical signals. Thus, reporting such radiological findings is important to create awareness on variant images of primary lacrimal sac melanoma. This will reduce misdiagnosis and mistreatment.

Background

Primary malignant melanoma of the lacrimal sac is an extremely rare condition. Yet, the lesion is highly malignant, can metastasize at a relatively early stage and patients with such lesions have poor prognosis. Early diagnosis is therefore essential. Notably, this condition is often misdiagnosed as chronic dacryocystitis. For accurate diagnosis, imaging examination is recommended to assess the severity of the disease, differentiate between tumors and inflammation and characterize tumor

features. Here, we present a case of primary malignant melanoma of the lacrimal sac. We provide a summarize of the computed tomography (CT) and magnetic resonance imaging (MRI) findings and review previous literature on this condition.

Case presentation

A 50-year-old female patient who had a mass in the lacrimal sac area for more than 3 months is presented in this report. The mass exhibited slow but progressive swelling. She experienced occasional epiphora and pain, without blood discharge or impaired vision. After 6 months, the patient underwent dacryocystectomy for the left lacrimal sac mass. Postoperative pathological examination confirmed the presence of malignant melanoma.

* Correspondence: suwei1970@yeah.net

[†]Ju-Wei Shao and Jian-Hua Ying are Co-first author

¹Department of Radiology, the Fourth Affiliated Hospital of Kunming Medical University, 176 Qing Nian Street, Kunming, People's Republic of China
Full list of author information is available at the end of the article



© The Author(s). 2020 **Open Access** This article is licensed under a Creative Commons Attribution 4.0 International License, which permits use, sharing, adaptation, distribution and reproduction in any medium or format, as long as you give appropriate credit to the original author(s) and the source, provide a link to the Creative Commons licence, and indicate if changes were made. The images or other third party material in this article are included in the article's Creative Commons licence, unless indicated otherwise in a credit line to the material. If material is not included in the article's Creative Commons licence and your intended use is not permitted by statutory regulation or exceeds the permitted use, you will need to obtain permission directly from the copyright holder. To view a copy of this licence, visit <http://creativecommons.org/licenses/by/4.0/>. The Creative Commons Public Domain Dedication waiver (<http://creativecommons.org/publicdomain/zero/1.0/>) applies to the data made available in this article, unless otherwise stated in a credit line to the data.

Three months after the operation, a lump in the lacrimal sac area had gradually increased. Local recurrence was suspected and she was referred to ophthalmology department for further treatment.

A physical examination revealed a scar from the previous operation. In addition, a 1.5 cm × 1.5 cm firm, localized and relatively clearly defined mass was observed in the left medial canthus. There was no cervical lymphadenopathy. A CT scan demonstrated a partially well-defined soft mass in the fossa of left lacrimal sac extending into the nasolacrimal duct and anterior ethmoid sinus (Fig. 1a). CT plain scan revealed that the lesion had a CT value of 49HU. The CT value of the tumor was similar to that of the extraocular muscle and wall of eyeball. Enhanced CT scan detected a CT value of 103HU for the arterial phase and 95HU for the venous phase (Fig. 1b). There was no eyeball involvement. Bone windows showed that the left nasolacrimal duct was enlarged and partial bone destruction of the nasolacrimal duct was confirmed. There was no regional or distant metastasis.

MRI plain scan demonstrated a partly well-demarcated mass of 1.2 cm × 1.3 cm × 2.3 cm in the lacrimal sac lesion. The tumors had infiltrated the septa orbital and subcutaneous adipose tissue. MRI revealed intermediate signal intensity on T1 and iso-or hyper-signal on T2

weighted images (Fig. 1c, d). The lesions displayed slight hyperintense signal on DWI. A dynamically enhanced MRI scan showed intense enhancement of the lesion margin and inhomogeneous enhancement of the lesion center (Fig. 1e).

A biopsy was obtained from the patient before the operation. Histopathological examination on the biopsy confirmed fusiform malignant tumoral cells with hyperchromatic nuclei. The tumor had high number of pigmented cells, inhomogeneous pigment granules and massive hemorrhage (Fig. 1f, g). Immunoreactivity to the S-100 protein, Melan-A, HMB-45, Nestin, and CD56 was positive (Fig. 1h, i). Therefore, a diagnosis of malignant melanoma was confirmed. Further examination of the whole body, including computed tomography scanning and B ultrasound revealed that there was no metastasis in the head and neck, chest, abdomen, and pelvis regions. Thus, total excision of tumor was performed. The patient refused to undergo radiotherapy and chemotherapy.

Discussion and conclusions

Primary malignant melanoma of the lacrimal sac is a rare condition, with less than 50 cases reported over the past 90 years worldwide. The first case of this disease was published in 1926 in Russia by Muravleskin [1]. The

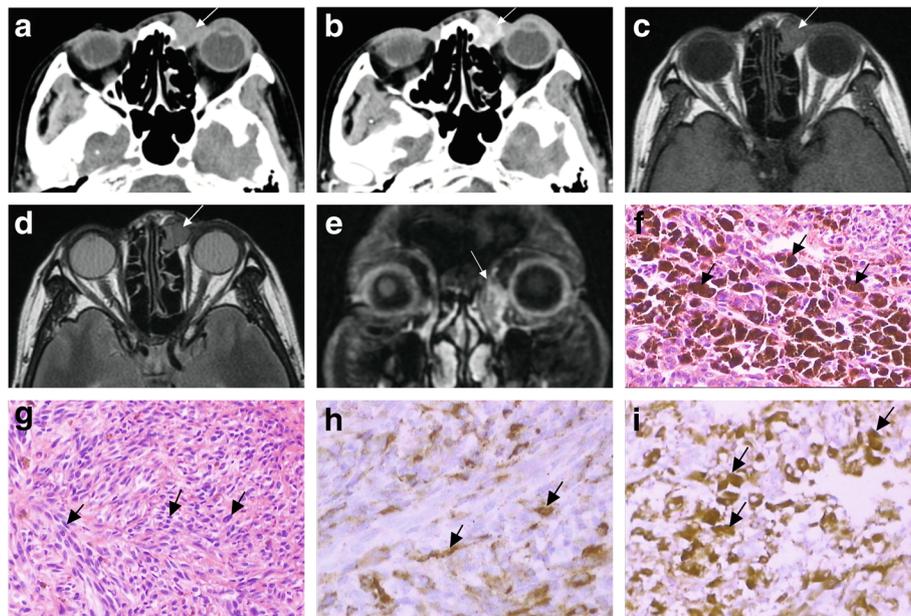


Fig. 1 **a** Axial CT shows a partly well-defined soft mass in the fossa of left lacrimal sac. **b** An enhanced CT scan shows marked enhancement. **c** T1-weighted axial MRI shows an intermediate signal intensity mass. **d** T2-weighted axial MRI shows a iso-or hyper-signal mass. **e** Gadolinium-enhanced T1-weighted coronal MRI demonstrates a hyperintense, well-defined enhancing mass, extending into the nasolacrimal duct and anterior ethmoid sinus. **f** Photomicrography of the lacrimal sac melanoma showing heavy melanin pigmentation produced by tumor cells (hematoxylin-eosin, × 40). **g** These tumour cells producing higher magnification showing spindle and ovoid shaped tumor cells with inhomogeneous pigment granules (hematoxylin-eosin, × 40). **h** Immunohistochemical staining with HMB-45 showing positive staining of the tumor cells (immunohistochemical staining, × 40). **i** The tumour cells showing strong immunoreactivity to melanoma-specific antibody, Melan-A in the immunohistochemical examination. (immunohistochemical staining, × 20)

incidence of primary lacrimal sac melanoma in lacrimal sac tumors ranges between 4 and 13% and it accounts for 0.7% of all ocular melanomas [2]. Melanoma cells are neuroectodermal tumors that originate from the neural crest. However, lacrimal sac epithelium does not contain melanocytic cells. It is speculated that malignant melanoma develops from melanin cells left in the epithelium of the lacrimal sac during the development of the embryonic system [3]. Based on a review of previous reports, the average age of patients at diagnosis is 59 years old (range of 27–81) with no significant sex predilection [1]. Primary melanomas of the lacrimal sac have a poor prognosis [2]. Clinically, its symptoms resemble those of dacryocystitis, leading to frequent misdiagnosis. Blood discharge (44%) and painless swelling (42%) are the most frequent symptoms of this condition. Due to its insidious onset, understanding the extent of invasion and early diagnosis are critical to facilitate timely intervention and improve the survival outcomes [1]. Although pathology and immunohistochemistry tests are needed to make a definitive diagnosis, CT and MRI imaging are also effective diagnostic tools for this condition [4]. Several reports on CT-scan for lacrimal sac melanoma show the presence of a soft lesion in the lacrimal sac fossa (Table 1). Similarly, our case presented isodensity and

slightly high density on CT images, without calcification. Cystic change is uncommon.

In the early stage, imaging examination showed round, well-defined and homogeneous density lesions without orbital or nasal sinus involvement. Nasolacrimal duct is easily involved [4], according to previous literatures on CT images, the lesions extend into the nasolacrimal duct without nasolacrimal duct bone destruction, accounting for 50% (10/20) of CT images (Table 1), after which they invade the surrounding bone and soft tissue. In the late stage, the lesions tended to be irregular and ill-defined masses with invasion of nasolacrimal duct, orbital and sinus [22]. CT is more sensitive to bone destruction than MRI [4]. Non-epithelial lesions appear to cause lower bone destruction than epithelial tumors. In accordance with previous imaging findings, only five cases mentioned bone destruction on CT images (5/20) (Table 1). Because our case is a relapsing primary malignant melanoma of the lacrimal sac after surgery, bone destruction may result from the previous dacryorhynchostomy. The operation was done in other hospitals. There was no CT before the operation. Unfortunately, we don't know the exact cause of bone destruction.

Of note, CT is not sufficient for tumor diagnosis because CT values do not show the number of melanin

Table 1 Description of CT findings in primary malignant melanoma of the lacrimal sac

case	references	CT density	extending area	bone destruction
1	Lloyd and Leone (1984) [5]	STM	a	a
2	Eide et al. (1993) [6]	STM	a	no
3	Owens et al. (1995) [7]	STM	ND and IM	a
4	Levine et al. (1996) [8]	STM	a	no
5	TY Malik et al. (1997) [9]	STM	a	no
6	Fishman and Ophir (1999) [10]	STM	ND, IM and NC	a
7	Lee et al. (2001)[11]	STM	ND and IM	a
8	Billing et al. (2003) [12]	STM	ND	no
9	Tello et al. (2004)[13]	STM	ND	no
10	Gleizal et al. (2005) [4]	STM	a	no
11	Nam et al. (2006)[14]	STM	ND and MOW	a
12	Lewis et al. (2006)[15]	STM	ND	no
13	Sitole et al. (2007)[16]	STM	a	posterior lacrimal duct wall
14	Heindl et al. (2008)[17]	STM	ND and NC	no
15	Li et al. (2012) [18]	STM	MOW	nasolacrimal canal and inferior turbinate
16	Maegawa et al. (2014) [19]	STM	ND and IM	a
17	Ren et al. (2014) [20]	STM	a	no
18	Pujari et al. (2014) [21]	isodense	a	anterior lacrimal crests infiltration
19	McGrath et al. (2016) [1]	STM	a	the lateral wall of upper nasal canal and anterior ethmoidal air cells
20	Kavoussi et al. (2016) [22]	STM	a	ethmoid sinuses
21	Present case	Isodense, slightly high	ND and AES	ND

STM soft tissue mass, ND nasolacrimal duct, IM inferior meatus, NC nasal cavity, MOW medial orbital wall, AES anterior ethmoid sinus, a: not mentioned

Table 2 Description of MR findings in primary malignant melanoma of the lacrimal sac

case	references	MR features
1	Billing et al. (2003) [12]	intermediate signal intensity on T1 and T2 weighted images, enhanced with intravenous gadolinium
2	Tello et al. (2004) [13]	intermediate signal intensity on T1 weighted images and hyperintensity on T2 weighted images
3	Lewis et al. (2006) [15]	iso-or hyper-signal on T1 weighted images
4	Li et al. (2012) [18]	lower intensity on T1 weighted images and high intensity on T2 weighted images, gadolinium-enhanced T1-weighted high intensity
5	Maegawa et al. (2014) [19]	hypointense on T2
6	Kavoussi et al. (2016) [22]	intermediate signal intensity on T1 and T2 weighted images
7	Present case	intermediate signal intensity on T1 weighted images and iso-or hyper-signal on T2 weighted images

granules. In contrast, MRI images provide effective diagnosis for malignant melanoma. MRI has a high soft-tissue resolution, making it the best imaging technique for evaluating malignant melanoma currently. Typically, MR images show the presence of hyperintensity signal on T1-weighted images and hypointense signal on T2-weighted images owing to the paramagnetic properties of melanin in most other parts of malignant melanomas. But this condition is rare in malignant melanoma of lacrimal sac. To the best of our knowledge, only six case reports have been published describing the MRI radiological features of this condition. Four cases (4/7) (including our case) show intermediate signal intensity on T1 weighted image, and only one case presented with typical signals on T2 weighted images (Table 2).

The characteristic MR signals are based on histologic features. The higher the melanin content, the more typical the signal is. In our case, massive hemorrhage and inhomogeneous pigmentation may precipitate the atypical signals, and since after only 3 months thereafter tumor blood supply and structure could still be some post-surgical changes in MR signals. This atypical signal may lead to misdiagnosis of amelanotic melanoma, which accounts for 2% of all cases of melanoma [23]. The malignant melanoma of the lacrimal sac contains fewer melanocytes than uveal melanoma. Compared with other parts of orbital malignant melanoma images, fewer cases present with the typical paramagnetic signals. Less pigmentation and intratumoral hemorrhage may cause atypical MR signals in the lacrimal sac, and therefore they should be considered during diagnosis. Malignant melanoma of the lacrimal sac may present intermediate signal intensity on T1 and iso-or hyper-signal on T2 weighted images in most cases. Intravenous gadolinium provides more information on tumor blood supply and structure [22]. However, only one case mentioned moderated image enhancement of lesions excluding our case. The patient in this report exhibited marked enhancement of lesions, suggesting hypervascular neoplasms. According to previous studies, many orbital

melanomas contain moderate and intense enhancement lesions following the administration of contrast material [24]. Contrast enhancement may improve the detection. Unfortunately, contrast enhanced cases are few and thus the characteristics of melanoma of lacrimal sac cannot be summarized.

The differential diagnosis of melanoma in the lacrimal sac considers primary epithelial neoplasms (85.6%), malignant lymphoma (7.8%), neural tumors, inflammatory lesions and metastatic tumor [25]. Cases with well-defined lesions extending into the nasolacrimal duct with minimal bone destruction are likely to be non-epithelial neoplasms. Most lacrimal sac tumors present intermediate signal intensity on T1 and T2 weighted images. Typical signals of malignant melanoma of lacrimal sac are rare, so high scrutiny is required to identify them. Imaging can diagnose cystic mass [26]. Imaging examination can effectively identify the location, size, extent of lesions, and invasion of adjacent soft tissue. The CT and MRI examinations are suitable for determining tumor recurrence during patient follow-up. Imaging examination provides more reliable information to guide decision-making in the choice of clinical treatment. Definite diagnosis should be made by pathologic evaluation and immunohistochemistry. A limitation of this case report is that imaging was only performed at the relapsing stage, and thus bone destruction may result from tumor development or from the initial surgery (or both).

To date, none has summarized the CT and MR findings on primary lacrimal sac melanoma. Based on previous reports and our case, most cases do not present typical signals. Thus, reporting such radiological findings is important to create awareness on variant images of primary lacrimal sac melanoma. This will reduce misdiagnosis and mistreatment.

Abbreviations

AES: Anterior ethmoid sinus; CT: Computed tomography; IM: Inferior meatus; MOW: Medial orbital wall; MRI: Magnetic resonance imaging; NC: Nasal cavity; ND: Nasolacrimal duct; STM: Soft tissue mass

Acknowledgements

We thank each author who contributed to this paper, especially thank Dr. Hong Zhou for her excellent assistance in the histopathologic procedures.

Authors' contributions

JWS and JHY collected the data, wrote the manuscript and reviewed the literatures. STX collected the data. WS and QH corrected the data and critically reviewed the manuscript. HZ participated in the histopathologic procedures. All authors have read and approved the final manuscript.

Funding

This work was supported by Special Joint Fund from Yunnan Provincial Department of Science and Technology and Kunming Medical University, Kunming, Yunnan, China. [Grant number 2018FE001(-267)]. The founder is Ju-wei Shao (first author) who has collected the data, written the manuscript and provide financial support.

Availability of data and materials

Not applicable.

Ethics approval and consent to participate

Not applicable.

Consent for publication

The patient clarified and gave written consent to use of the clinical information, CT and MR images for publication in the BMC Ophthalmology.

Competing interests

The authors declare that they have no competing interests.

Author details

¹Department of Radiology, the Fourth Affiliated Hospital of Kunming Medical University, 176 Qing Nian Street, Kunming, People's Republic of China.

²Department of Radiology, the First Hospital of Kunming, Kunming, People's Republic of China. ³Department of Pathology, the Fourth Affiliated Hospital of Kunming Medical University, Kunming, People's Republic of China.

Received: 30 August 2019 Accepted: 26 February 2020

Published online: 14 May 2020

References

- McGrath LA, O'Hagan SB. Malignant melanoma of the lacrimal sac: case report and major review with treatment update. *J Ophthalmol Vis Neurosci*. 2016;1(3):1–4. <https://pdfs.semanticscholar.org/60b8/a5fe52ab8930f84846e786ba85ce7415ba55.pdf>.
- Subramaniam SS, Anand R, et al. Primary lacrimal sac melanoma with metastatic cervical disease: a review of the literature and case report. *J Oral Maxil Surg*. 2017;75:1438–41.
- Heindl LM, Jünemann AG, Kruse FE, et al. Tumors of the lacrimal drainage system. *Orbit*. 2010;29(5):298–306.
- Gleizal A, Kodjikian L, Lebreton F, et al. Early CT-scan for chronic lacrimal duct symptoms-case report of a malignant melanoma of the lacrimal sac and review of the literature. *J Cranio Maxill Surg*. 2005;33:201–4.
- W. C. Lloyd, C. R. Leone, (1984) Malignant Melanoma of the Lacrimal Sac. *Archives of Ophthalmology* 102 (1):104-107.
- Eide N, Refsum SB, Bakke S, Primary malignant melanoma of the lacrimal sac. *Acta Ophthalmol (Copenh)* .1993;71(2):273-6.
- Owens RM, Wax MK, Kostik D, et al. Malignant melanoma of the lacrimal sac. *Otolaryngol Head Neck Surg*.1995;113(5):634-40.
- Levine MR, Dinar Y, Davies R, et al. Malignant melanoma of the lacrimal sac. *Ophthalmic Surg Lasers*.1996;27(4):318-20.
- TY Malik, R Sanders, JDH Young, et al. Malignant melanoma of the lacrimal sac. *Eye*.1997; 11,935-945.
- Fishman G, Ophir D. Malignant melanoma of the lacrimal sac: a case study. *Am J Otolaryngol*.1999 ;20(5):336-9.
- Lee HM, Kang HJ, Choi G, et al. Two cases of primary malignant melanoma of the lacrimal sac. *Head Neck*.2001,23(9):809-13.
- Billing K, Malhotra R, Selva D, et al. Magnetic resonance imaging findings in malignant melanoma of the lacrimal sac. *Br J Ophthalmol*. 2003; 87:1187-8.
- Tello JS, Campillo NG, Rodriguez JL, et al. Malignant melanoma of the lacrimal sac. *Otolaryngol Head Neck Surg*. 2004;131:334-6.
- Nam JH, Kim SM, Cho JH, et al. Primary malignant melanoma of the lacrimal sac: a case report. *Korean J Intern Med*. 2006;21(4):248-51.
- AM Lewis, PM Clarke, JM Olver. Primary nasolacrimal duct malignant mucosal melanoma. *Br J Ophthalmol*.2006;90:114-125. .
- Sitole S, Zender CA, Ahmad AZ, et al. Lacrimal sac melanoma. *Ophthal Plast Reconstr Surg*.2007;23(5):417-9.
- Heindl LM, Schick B, Kämpgen E, et al. Malignant melanoma of the lacrimal sac. *Ophthalmologie*.2008;105(12):1146-9.
- Li YJ, Zhu SJ, Yan H, et al. Primary malignant melanoma of the lacrimal sac. *BMJ Case Rep*.2012;13:2012.
- Maegawa J, Yasumura K, Iwai T, et al. Malignant melanoma of the lacrimal sac: a case report. *Int J Dermatol*.2014;53(2):243-5.
- Ren M, Zeng JH, Luo QL, et al. Primary malignant melanoma of lacrimal sac. *Int J Ophthalmol*.2014;7(6):1069-70.
- Pujari A, Ali MJ, Mulay K, et al. The black lacrimal sac: a clinicopathological correlation of a malignant melanoma with anterior lacrimal crest infiltration. *Int Ophthalmol*.2014;34(1):111-5.
- Kavoussi SC, Levin F, Servat JJ. Orbital Extension of Untreated Lacrimal Sac Melanoma Following Dacryocystorhinostomy. *Ophthal Plast Reconstr Surg*. 2016; 32: e76.
- Lorenzano D, Miszkiel K, Rose GE. Orbital melanoma masquerading as a "Gallop haemangioma". *Orbit*. 2017; 36:81-3.
- Purohit BS, Vargas MI, Ailianou A, Becker, M; Orbital tumours and tumour-like lesions: exploring the armamentarium of multiparametric imaging. *Insights Imaging*. 2016;7(1):43-68.
- Song XM, Wang J, Wang S, et al. Clinical analysis of 90 cases of malignant lacrimal sac tumor. *Graefes Arch Clin Exp Ophthalmol*. 2018;256(7):1333-1338. .
- Deep P, Rand R, Sylvia K. Primary lacrimal sac diffuse large B-cell lymphoma in a child[J]. *Journal of AAPOS: the official publication of the American Association for Pediatric Ophthalmology and Strabismus*. 2019, 23(1):53-55.

Publisher's Note

Springer Nature remains neutral with regard to jurisdictional claims in published maps and institutional affiliations.

Ready to submit your research? Choose BMC and benefit from:

- fast, convenient online submission
- thorough peer review by experienced researchers in your field
- rapid publication on acceptance
- support for research data, including large and complex data types
- gold Open Access which fosters wider collaboration and increased citations
- maximum visibility for your research: over 100M website views per year

At BMC, research is always in progress.

Learn more biomedcentral.com/submissions

