Case Report
Primary eyelid angiosarcoma in a Chinese patient

Xiaoli Huang, Song Sun

Department of Ophthalmology Wuxi No. 2 Hospital Affiliated to Nanjing Medical University, 68 Zhongshan Rd, Wuxi 214002, P. R. China

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Abstract: Purpose: To describe a case of primary eyelid angiosarcoma and review the literature to emphasize aware of this rare disease. Case Report: We report a further case which is the first Chinese primary eyelid angiosarcoma in the literature. A 76-year-old woman presented with a 6-month history of a painless lesion on her left eyelid. The patient finally proved to be angiosarcoma on histopathologic and be treated with complete surgical excision with a frozen section margin control. With a 6 months follow-up, we haven’t found any symptoms of recurrence or metastasis. Conclusions: Cutaneous angiosarcoma with eyelid is a rare, soft-tissue sarcoma of endothelial cell origin that is aggressive malignancy and has a poor prognosis. In our case report, the patient was treated with complete surgical excision with a frozen section margin control, and a 6 months follow-up, we haven’t found any symptoms of recurrence or metastasis.

Keywords: Eyelid, eye, angiosarcoma, tumor, surgical resection

Introduction
Cutaneous angiosarcoma (AS) with eyelid is a rare, soft-tissue sarcoma of endothelial cell origin that be aggressive malignancy and has a poor prognosis. It’s first described by Livingston and Klemperer in 1926 [1]. Soft tissue sarcomas account for less than 1% of all cancers, and angiosarcoma account for 2% of them, and cutaneous angiosarcomas mostly occur at head and neck are as account for 59%, rarely involve periocular tissue and affect men twice as often as women [2], and results in a terrible 5-year survival rate of 12%–33% [3].

Case report
A 76-year-old woman was referred to our department for a 6-month history of a lesion on her left face. Specialist examination showed an erythematous lesion involving the upper eyelid, inner canthus, lower eyelid, and bridge of the nose. The lesion was approximately 2 cm × 2 cm, without ulceration and without clear boundaries (Figure 1A). The patient had no history of skin cancer. The patient’s family history and past medical history were not significant. Laboratory examination including blood chemistry and blood count could not reveal any specific pathology. An excisional skin biopsy from the center of the lesion was performed. Ocular examination showed serosanguinuous drainage from the biopsy site. Histopathological examination demonstrated a cytologically atypical vascular neoplasm composed of spindle-shaped and epithelioid cells (Figure 2A). The tumour cells were intensely immunopositive for CD31 (Figure 2B) and CD34 (Figure 2C). A diagnosis of eyelid angiosarcoma (AS) was made. The patient underwent complete surgical excision and free flap skin graft under general anaesthesia to achieve complete surgical resection of the tumour. Twelve frozen sections were examined and found to have negative intraoperative margins. The survival of the graft flap was good even after 1 month (Figure 1B). After 12 months of follow-up, there was no evidence of recurrence or metastasis.

Discussion
Cutaneous angiosarcoma is a rare vascular endothelial cells malignant tumour. Panta Rouhani et al [2] reviewed 12, 114 cases of skin and soft tissue sarcoma, and angiosarcoma was found to account for 1.6%. Angiosarcoma has a higher incidence among Caucasians, a male: female incidence ratio of 2.0, and rarely involves the eyelid tissue; in fact,
Primary eyelid angiosarcoma

Figure 1. A: Patient with an isolated unilateral angiosarcoma of left eyelid nasally; B: One month after the surgery.

Figure 2. A: Histopathological examination showed dermal tumor tissue flake, crack-like distribution, some fissures see obesity tumor cells, plasma rich red dye, Cutaneous (hematoxylin and eosin, × 200); B: Immunoreactivity was
the eyelids are involved in only 3% of cases. Most patients were elderly women approximately 70-years-old. Researchers [4] believe that a history of exposure to chemicals, ionizing radiation, chronic lymphedema, sunlight, and trauma, and the presence of other relevant mutations are related to the development of angiosarcoma. Angiosarcomas are rapidly progressing diseases that can spread not only to nearby lymph nodes, but also metastasize through the blood to the lungs, liver, bone, and other locations. Therefore, it is important to make an early diagnosis and to institute the correct treatment.

The diagnosis of angiosarcoma is based on histopathological evaluation. Histologically, angiosarcoma is characterized by abnormal and pleomorphic malignant endothelial cells [5]. Neoplastic cells range from spindle to epithelioid cells with large irregular nuclei, coarse chromatin, and eosinophilic cytoplasm. In addition, angiosarcoma is associated with positive immunohistochemical markers, including CD31, CD34, and VIII factor related antigen. CD31 is one of the most specific and sensitive markers [6]. Our case was considered a well-differentiated angiosarcoma with the expression of immunohistochemical markers of vascular differentiation (CD31 and CD34).

The mainstay treatment for cutaneous angiosarcoma includes surgical excision followed by external beam radiotherapy. Although recent case reports have described approaches for the management and about the outcome of angiosarcoma [6], specific management criteria are absent owing to the lack of large randomized controlled trials of patients with angiosarcoma.

The prognosis of cutaneous angiosarcoma is very poor, with a 5-year survival rate of only 12-33% [7]. The most important factors for improving prognosis are small tumour size, younger age, isolated tumors, negative resection margin, and postoperative radiation therapy [7].

Our case is important for a number of reasons. First, angiosarcoma of the eyelids is rare. We describe the first case of primary eyelid angiosarcoma in a Chinese patient. In addition, successful treatment of this case can provide some suggestions for treatment. Finally, clinicians should be alert that angiosarcoma might present with a wide variety of clinical features.

Disclosure of conflict of interest

None.

Address correspondence to: Dr. Song Sun, Department of Ophthalmology Wuxi No. 2 Hospital Affiliated to Nanjing Medical University, 68 Zhongshan Road, Wuxi 214002, P. R. China. Tel: 86 510 66681222; Fax: 86 510 82754933; E-mail: songsun2000@163.com

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