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Sir,

Localized peripheral primitive neuroectodermal tumor of the conjunctiva: a rare presentation

We report a rare presentation of peripheral primitive neuroectodermal tumor (pPNET) as an isolated conjunctival lesion.

Case report

A 23-year-old man presented to our clinic with a 1-month history of a painless, enlarging conjunctival lesion in his right eye (Figure 1a). The lesion was located at the lower temporal bulbar conjunctiva and was round, pinkish, and well-circumscribed on slit lamp biomicroscopy. He was diagnosed with left frontoparietal glioblastoma multiforme and had received subtotal tumor excision with radiochemotherapy as well as adjuvant Temozolomide two years ago. We excised the lesion and the histopathological report revealed conjunctival tissue with subepithelial diffuse infiltrates of tumor cells that possess medium-sized nuclei, fine chromatin, small nucleoli, and inconspicuous cytoplasm (Figure 2). Morphologically, it was a malignant tumor different from the previously diagnosed glioblastoma of the brain. Further immunohistochemical study showed positivity for CD99, CD56, neuron-specific enolase, and S-100 protein. Some were strongly positive for vimentin. All were negative for glial fibrillary acidic protein (GFAP, 6F2) and synaptophysin (SY-38). Re-excision was done weeks later, with no evidence of residual tumor histologically. Systemic survey showed no sign of metastasis. The oncologist suggested total surgical resection without adjuvant radiochemotherapy. The patient was followed for 18 months without local tumor recurrence (Figure 1b) or systemic metastasis.

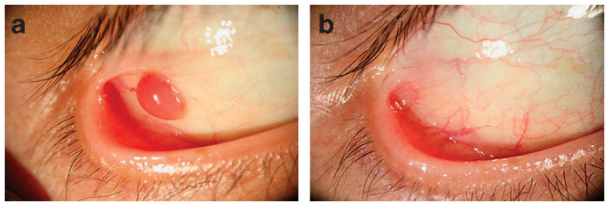

Figure 1 (a) Right eye external eye photo revealed a conjunctival mass located at the temporal side of the lower bulbar conjunctiva initially. (b) No sign of local tumor recurrence after excision with 18 months follow-up.

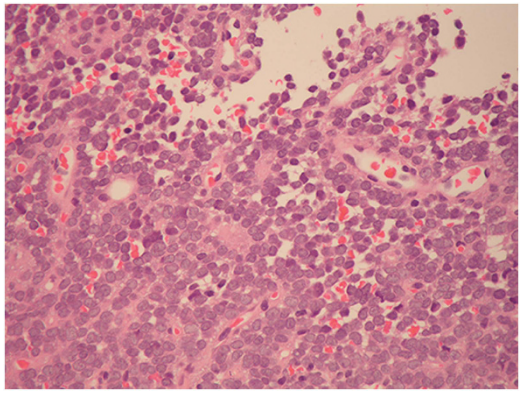

Figure 2 High-power magnification (× 400) shows medium-sized nuclei, fine chromatin, small nucleoli, and inconspicuous cytoplasm. The tumor cells are focally arranged in rosette-like structures.

Comment

PNET is a group of high malignant small round cell tumors arising from primitive neuroectodermal progenitor cells, which was classified as central PNET and peripheral PNET (pPNET). Batsakis *et al*[1] divided PNET into the following three groups based on the tissue of origin: central nervous system PNET (tumors derived from the central nervous system), neuroblastoma (tumors derived from the autonomic nervous system) and pPNET (tumors derived from tissues outside the central and autonomic nervous system). pPNET was reported in a variety of locations, including the orbit and conjunctiva,[2,3] and has been considered as a highly aggressive tumor that typically metastasizes.[1] A less propensity of metastasis and better prognosis for orbital pPNET was considered to be related to symptomatic disease, which led to early medical care seeking and early diagnosis.[3] The only conjunctival pPNET case reported was a 16-year-old Caucasian boy who was treated with local wide excision and chemotherapy. No distant metastasis and no recurrence of tumor both clinically and radiologically for 29 months was reported.[2] Here we reported the second case of conjunctival pPNET treated with only local excision, who has been in complete tumor remission for 18 months. Conjunctival pPNET as secondary malignancy was less likely because the secondary malignancy after Temozolomide treatment was mostly hematogenic.[4] The oncologist suggested surgical resection only and close follow-up, which was similar to Chow's experience for localized cutaneous and subcutaneous Ewing's sarcoma family tumors.[5]

We reported a rare case of conjunctival pPNET who remained in tumor remission for 18 months following surgical excision only. A standard treatment strategy cannot be concluded from a single case report. Nevertheless, collaboration with specialists and timely diagnosis and treatment may be beneficial for a better outcome.

Conflict of interest

The authors declare no conflict of interest.

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Sir,
Re: Spontaneously resolved exudative retinal detachment caused by orbital cellulitis in an immune compromised adult

We thank the authors for presenting a very interesting case of orbital cellulitis and exudative serous retinal detachment with excellent MRI scans showing orbital pathology.[1] The authors suggest orbital cellulitis as a possible cause of the retinal detachment. The hematological tests suggest that the patient may have had a septicemia, with elevated ESR, C-reactive protein level, and a neutrophilic leukocytosis. Septicemia is well known to cause serous retinal detachments.

We wonder if the serous retinal detachment described in this case report was caused by the septicemia resulting from the primary infection, and if blood cultures were done to exclude this possibility.

Conflict of interest

The authors declare no conflict of interest.

Reference

1 Farah E, Kalantzis G, Papaefthimiou I, Koutsandrea C, Georgalas I. Spontaneously resolved exudative retinal detachment caused by orbital cellulitis in an immunocompromised adult. *Eye* 2014; **28**: 109–110.

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Sir,
Response to Perera and Ali

We would like to thank Perera and Ali[1] for their interest in our case report and their useful comments. It is true that serous retinal detachments can occur in patients suffering from septicemia or disseminated intravascular coagulation (DIC).[2,3]

In our case,[4] an 89-year-old immunocompromised patient presented with fever and elevated inflammatory markers (that is, raised ESR, C-reactive protein and neutrophilic leukocytosis), but the blood cultures that were performed were negative for any microorganism. Additionally, culture of nasal aspirates revealed methycillin-resistant *Staphylococcus aureus* and Warneri-Staphylococcus, which were consistent with ethmoid sinusitis leading to orbital cellulitis and subsequent exudative retinal detachment.

Conflict of interest

The authors declare no conflict of interest.

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Sir,
Maculopathy following extended usage of Clomiphene citrate

Clomiphene citrate (CC) is a selective estrogen receptor modulator mostly used for treatment of infertility associated with polycystic ovarian disease.[1] Unlike Tamoxifen, which is also a selective estrogen receptor