Primary Conjunctival Rhabdomyosarcoma Successfully Treated with Surgery and Chemotherapy

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Abstract

***Purpose*:** To report the outcome of a case of primary conjunctival rhabdomyosarcoma treated by surgical excision combined with chemotherapy

***Case report*:** A 4-year-old boy presented with a visible recurrence of conjunctival mass in the left eye. The orbital magnetic resonance imaging (MRI) showed the lesion was confined to the conjunctiva without orbital infiltration. Histology and immunophenotype were consistent with an embryonal rhabdomyosarcoma. A surgical excision was performed followed by intensive chemotherapy. The patient remains clinically tumor-free during 4 years of follow-up, with $^{20}/_{20}$ vision in both eyes and no treatment complications.

***Conclusion*:** The treatment by surgical excision combined with chemotherapy was justified in this case of primary rhabdomyosarcoma, which eliminated the potential complications of radiotherapy.

***Keywords*:** Conjunctival Tumor, Embryonal Rhabdomyosarcoma, Pediatrics, Surgery, Chemotherapy

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Introduction

Rhabdomyosarcoma is the most common primary malignancy of the orbit in children.[1] However, primary conjunctival rhabdomyosarcoma is rarely reported.[2-5] We report herein the case of a 4-year-old boy with primary embryonal rhabdomyosarcoma confined to conjunctiva, treated by surgical excision combined with chemotherapy. The child remained clinically tumor-free during 4 years' follow-up.

Case report

A 4-year-old boy was referred to Zhongshan Ophthalmic Center, Sun Yat-Sen University by a local ophthalmologist because of a recurrent conjunctival granuloma in the left eye in April 2007.

Eight months earlier, a red mass was noted in the superior bulbar conjunctiva, extending slowly during one month, and the patient was seen by a local ophthalmologist for excision without histopathological evaluation. In January 2007, there was a visible recurrence of the lesion in the superotemporal conjunctiva. At 3-month follow-up examination, the mass appeared slightly enlarged.

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Therefore, an excisional biopsy was scheduled by the local ophthalmologist, and then the patient was referred to Zhongshan Ophthalmic Center.

There was no history of eye trauma, and no family history of similar ocular disease. The patient had $^{20}/_{20}$ vision in both eyes. The lesion appeared as a fleshy pink mass in the superior conjunctiva of the left eye (Figure 1-A). There was no proptosis, eyelid edema or limitation in eye movements. The orbital magnetic resonance imaging (MRI) showed the lesion was confined to the conjunctiva without orbital infiltration.

Histopathologic examination of biopsy specimens showed a tumor composed of primitive mesenchymal cells (Figure 1-B). The mesenchymal cells were elongated shapes with cytoplasmic eosinophilia, and the immunohistochemical staining for markers of desmin (Figure 1-C), actin (Figure 1-D) and vimentin were positive. Histology and immunophenotype were analogous to embryonic skeletal muscle, and consistent with an embryonal rhabdomyosarcoma.[6,7] Systemic workup for extension of tumor was negative.

Considering that the rhabdomyosarcoma was confined to the conjunctiva, a surgical excision completed with cryotherapy was performed with the aim to prevent the recurrences. However, there were still microscopic residual neoplastic cells after surgery. Chemotherapy was then initiated by our pediatric oncologists: alternating the use of ifosfamide, vincristine (IV) and cyclophosphamide, actinomycin D, vincristine (CAV). Six cycles were repeated with an interval of 3 weeks, followed by two cycles with an interval of 3 months, and then two cycles with an interval of 6 months. After one-year of chemotherapy, biopsies showed no microscopic neoplastic elements. The patient remained clinically tumor-free during 4 years of follow-up, with $^{20}/_{20}$ vision in both eyes and no treatment complications (Figure 2).

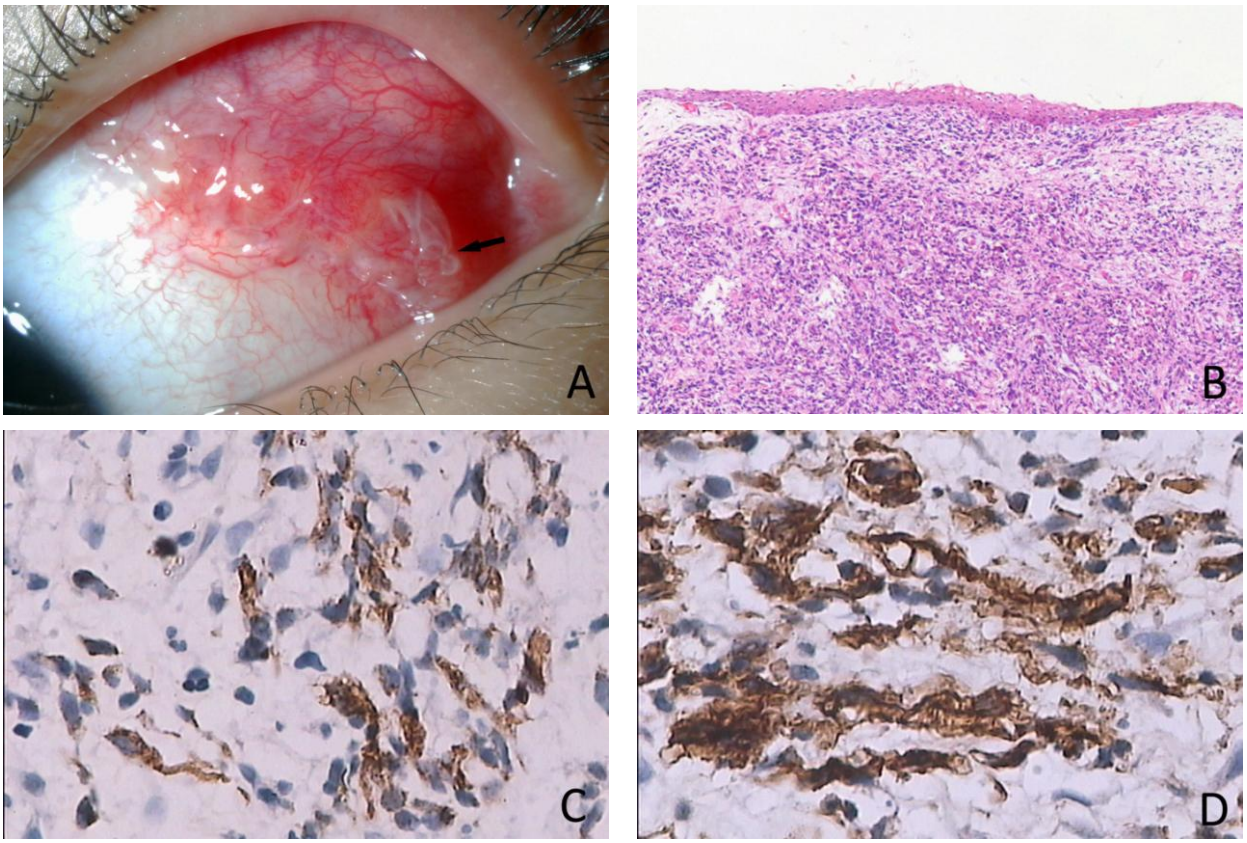

Figure 1. A 4-year-old boy with primary embryonal rhabdomyosarcoma confined to conjunctiva. (A) Conjunctival rhabdomyosarcoma appears as a fleshy pink mass in the superotemporal fornix. Conjunctival sutures (arrow) remains after the first surgical excision. (B) Histopathologic examination shows a tumor composed of primitive mesenchymal cells, acquired cytoplasmic eosinophilia and elongated shapes (hematoxylin-eosin x100). The immunohistochemical stainings for markers of desmin (C) and actin (D) were positive (x400).

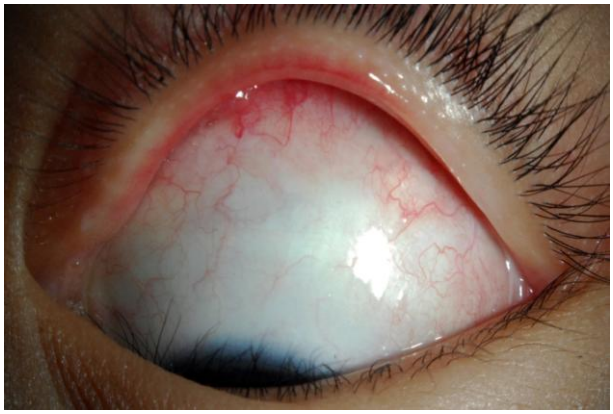

Figure 2. Photograph shows clinically tumor-free after 4 years' follow-up, only a small amount of conjunctival scar observed in the superior fornix.

Discussion

Rhabdomyosarcoma is the most common soft tissue sarcoma in the pediatric population. The primary sites include head and neck (45%), trunk (40%), and extremities (15%). About 25-35% of head and neck rhabdomyosarcoma arise in the orbit. However, the occurrence of this tumor in the conjunctiva is rare. Generally, rhabdomyosarcoma is classified into four morphologic categories, including embryonal, alveolar, botryoid, and pleomorphic. The favorable prognosis may be related to the tumor site, earlier detection and embryonal cell type.[8]

In the current case, the diagnosis was difficult because the lesion was confined to the conjunctiva, with no orbital involvement. The initial diagnosis of conjunctival granuloma was made, and the lesion was initially excised without biopsy. It should be emphasized that histopathological investigation of all enlarging conjunctival masses is mandatory for a prompt diagnosis and treatment. Furthermore, surgical removal of a mass without further investigations could lead to the eventual recurrence of the tumor.

The tumor was staged as group IIa according to the International Rhabdomyosarcoma Study Group (IRSG) clinical grouping classification, and the current recommendations for group II were chemotherapy, Vincristine and Dactinomycin (VA) and radiotherapy.[1] However, the patient's parents disagreed with radiotherapy considering the side effect of radiation, although severe complications are rare.[8] Since the conjunctival rhabdomyosarcoma was anteriorly located, there was the possibility of complete removal of the mass. On the other hand, results of the International Society of Pediatric Oncology Malignant Mesenchymal Tumor (MMT-89) trial suggested that avoidance of radiotherapy was justified for some patients if treated according to IRSG guidelines.[9] Brichard and coworkers have reported a case of primary conjunctival rhabdomyosarcoma treated by chemotherapy alone (stage II nonalveolar orbital tumors).[10] In our case intensive chemotherapy was administered without radiotherapy after surgery. The results were encouraging after 4 years of follow-up; the patient remained clinically tumor-free, with no complications or recurrences.

Conclusion

Our case emphasizes the importance of careful histopathological review of all enlarging conjunctival masses in children and adult for a prompt diagnosis and treatment. The treatment by surgical excision combined with chemotherapy was justified in this case of primary conjunctival rhabdomyosarcoma, which eliminated the potential complications of radiotherapy.

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