Case Report

Rhabdomyosarcoma of the Conjunctiva: A Case Report

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ABSTRACT

Rhabdomyosarcoma is an uncommon, rapidly growing soft-tissue sarcoma that primarily affects the paediatric population. The most common sites of involvement are the head, neck, and orbit. Conjunctival involvement is rare. We report a 9-year-old boy who presented with a conjunctival lesion in his right eye. The lesion was surgically removed, and histopathological examination of the excisional biopsy specimen confirmed embryonal rhabdomyosarcoma. The patient subsequently received systemic chemotherapy locally and abroad. At 24 months follow-up, he showed good clinical recovery with scar tissue formation at the excision site and no evidence of recurrence. This case highlights the importance of considering rhabdomyosarcoma in the differential diagnosis of atypical conjunctival lesions in children, as prompt diagnosis and treatment may improve prognosis.

Keywords: Rhabdomyosarcoma, Conjunctival Neoplasms, Eye Neoplasms, Soft Tissue Neoplasms, Orbital Neoplasms.

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INTRODUCTION

Rhabdomyosarcoma (RMS) is the most common soft-tissue sarcoma in the pediatric population and adolescents, arising from primitive mesenchymal cells capable of differentiating into striated muscle tissue.[1] It most commonly involves the head and neck region, genitourinary tract, and extremities, with the orbit representing a recognized ophthalmic site. Approximately one-fifth of RMS cases originate in the orbit, accounting for nearly one-third of head and neck cases.[1] In contrast, conjunctival involvement is exceedingly rare, reported in only about 12% of orbital cases.[2] A retrospective case series from Khyber Teaching Hospital, Peshawar, Pakistan, reported 12 patients with orbital rhabdomyosarcoma between 2015 and 2019.[3] However, primary conjunctival rhabdomyosarcoma remains exceedingly rare.

We report a case of conjunctival embryonal rhabdomyosarcoma in a 9-year-old boy to highlight its presentation, diagnosis, management, and follow-up.

CASE REPORT

A 9-year-old boy presented to a tertiary care ophthalmology centre with a conjunctival lesion in his right eye that had persisted for several months (Figure 1, left image). His past medical, birth, and ocular history was unremarkable, with no comorbidities. Uncorrected visual acuity was 6/6 bilaterally. Systemic examination revealed multiple hyperpigmented macules over the lower back, arms, and fingers (Figures 2). Although these cutaneous findings raised the possibility of an associated systemic or syndromic condition, no definite syndromic diagnosis was established at presentation.

His past medical, birth, and ocular history were unremarkable, and no comorbidities were reported. Visual acuity was 6/6 in both eyes. On systemic examination, multiple pigmented spots were noted on his lower back, fingers, and arms (Figure 2).

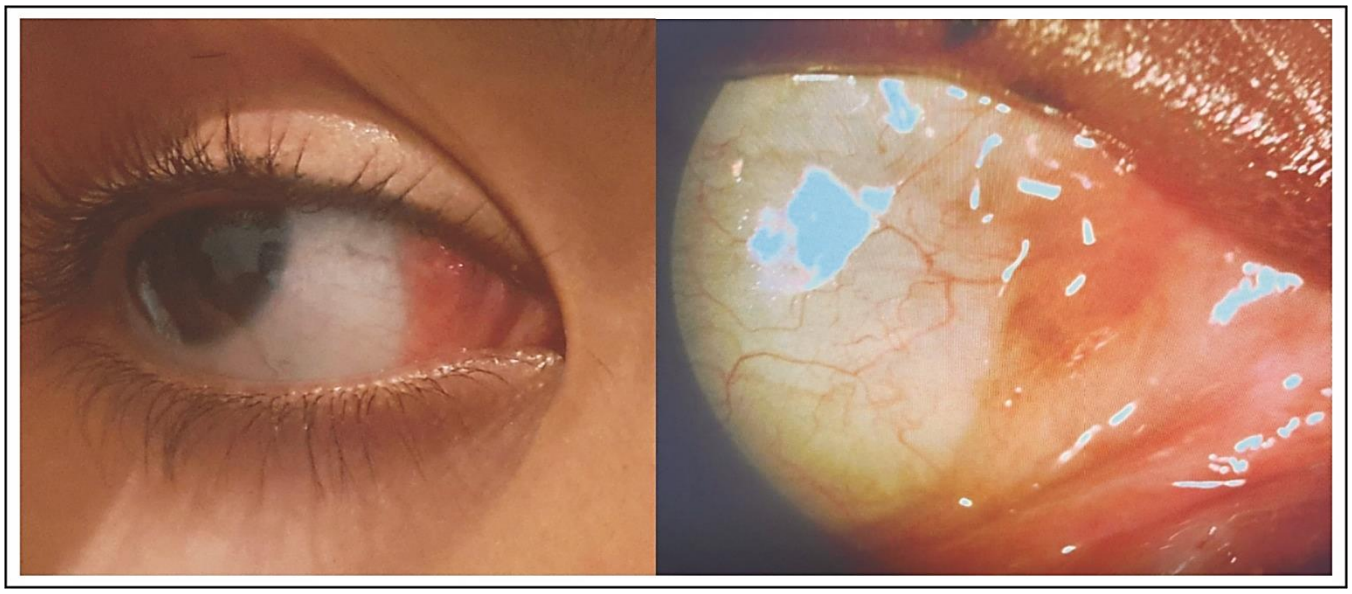

Figure 1: Conjunctival Lesion in right eye before treatment (Left).Postoperative photograph showing a keloid-like scar at the excision site on follow-up(Right).

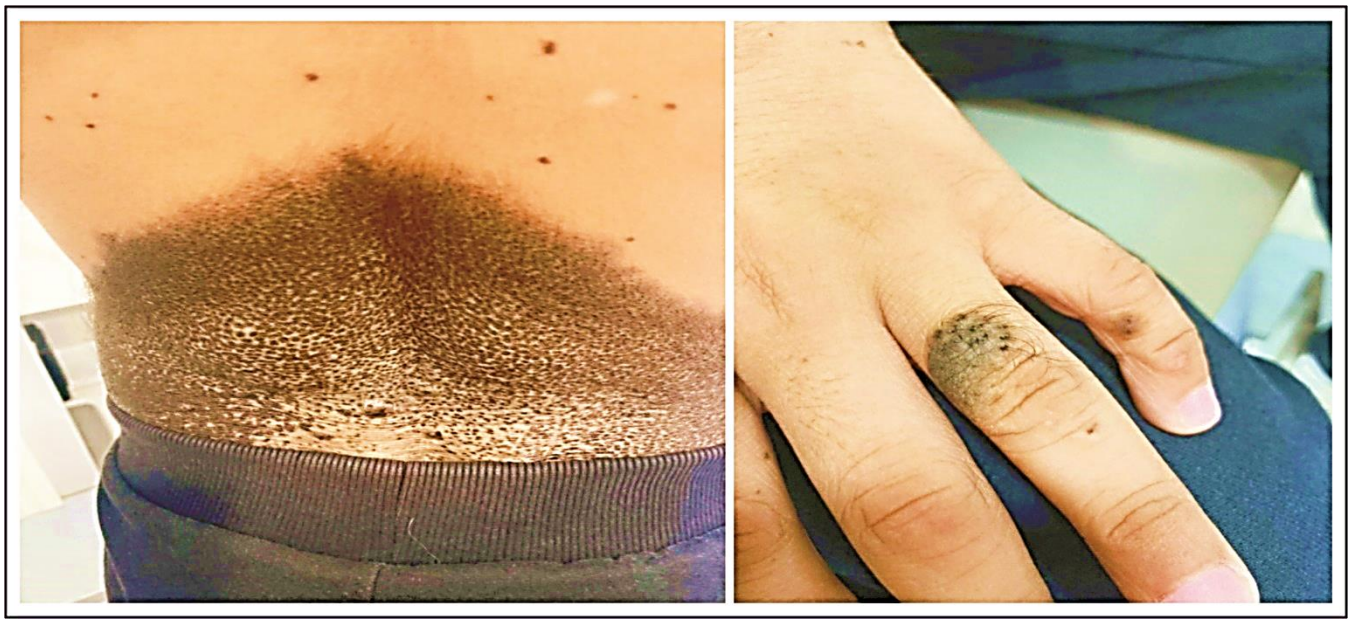

Figure 2: Pigmentation on lower back (Left). Hyperpigmented macules over the hand/fingers.

Slit-lamp examination showed an elevated nodular conjunctival lesion measuring approximately 10×2mm near the medial canthus of the right eye. In view of its suspicious appearance, an excisional biopsy was performed under general anesthesia after obtaining informed parental consent. Intraoperatively, the lesion was confined to the conjunctiva and Tenon's capsule, with no invasion of deeper tissues or adjacent structures.

Gross examination revealed a polypoidal soft-tissue fragment. Microscopically, the specimen was lined by benign squamous epithelium with an underlying band-like infiltrate of atypical spindle cells. These tumor cells showed moderate pleomorphism, hyperchromatic nuclei with inconspicuous nucleoli, and moderate to abundant eosinophilic to clear cytoplasm. Numerous mitotic figures were identified, indicating brisk proliferative activity.

Immunohistochemical analysis was performed for further characterization. The tumor cells showed diffuse positivity for desmin and focal positivity for myogenin, supporting skeletal muscle differentiation. They were negative for S-100 and HMB-45, helping exclude melanocytic and neural neoplasms. The Ki-67 proliferative index was 40%–50%, consistent with a high-grade malignant neoplasm. Based on the combined histomorphology and immunophenotypic findings, a diagnosis of embryonal rhabdomyosarcoma of the conjunctiva was established.

The patient was referred to pediatric oncology services and commenced systemic chemotherapy, with subsequent continuation of treatment at another oncology centre. At 24-month follow-up, he demonstrated satisfactory clinical recovery, with a keloid-like scar at the surgical site and no evidence of local progression (Figure 1, right image). He remains on topical anti-inflammatory, and lubricating eye drops for symptomatic relief. Serial photographic documentation has been shared with the treating oncology team for ongoing evaluation.

DISCUSSION

Rhabdomyosarcoma (RMS) is the most common soft-tissue sarcoma in the pediatric population and adolescents, with the orbit representing a recognized ophthalmic site of disease. However, primary conjunctival RMS is exceptionally rare, and most available literature consists of isolated case reports and small ophthalmic series. In the large ophthalmic series by Shields et al, conjunctival involvement accounted for only 4 of 33 cases (12%), underscoring the rarity of this presentation.[2] More recent case reports by Bakhtiary and Barkley, Morales et al, and Pennington et al, similarly describe conjunctival RMS as an unusual localized ocular surface lesion that may clinically mimic a benign conjunctival mass.[1,4,5]

The present case is comparable to previously reported conjunctival RMS cases in several respects. Similar to the reports by Bakhtiary and Barkley and Morales et al, our patient was a child with a localized conjunctival lesion, preserved visual acuity, and histopathologic confirmation of embryonal rhabdomyosarcoma after excision biopsy.[1,5] Pennington et al, described a rapidly enlarging conjunctival mass in a young boy that was initially suspected to be benign but was ultimately diagnosed as embryonal/botryoid RMS, emphasizing the importance of early biopsy in atypical conjunctival lesions.[5] Polito et al, also reported primary botryoid conjunctival RMS confined to the conjunctiva, supporting that conjunctival disease may present without deep orbital extension.[6] Taken together, these reports and the current case highlight that conjunctival RMS can present deceptively as a superficial lesion despite its malignant nature.

Histopathologically, RMS comprises several subtypes, including embryonal, alveolar, pleomorphic, and spindle cell/sclerosing variants.[7] Among pediatric ophthalmic cases, embryonal RMS is the predominant subtype and is generally associated with a more favorable prognosis than alveolar RMS.[2,5,8] In our patient, the diagnosis was supported by the morphology of atypical spindle cells together with immunohistochemical positivity for desmin and myogenin, findings consistent with skeletal muscle differentiation and in keeping with previously reported conjunctival embryonal RMS cases.[1-7]

An additional notable feature in our patient was the presence of multiple hyperpigmented macules on the back, arms, and fingers. This finding is not routinely emphasized in published conjunctival RMS reports. Hyperpigmented or café-au-lait–type macules have a broad differential diagnosis and may occur as isolated cutaneous findings or in association with neurocutaneous syndromes. In the absence of a confirmed syndromic diagnosis in our patient, a definite association cannot be established. Nevertheless, their presence is clinically noteworthy and may justify careful systemic examination and, where appropriate, dermatologic, or genetic evaluation.

From a treatment standpoint, management of RMS depends on site, stage, histology, and extent of residual disease. In ophthalmic RMS, multimodal management with surgery, systemic chemotherapy, and selected radiotherapy has substantially improved outcomes.[2,8] Based on the clinical confinement of the lesion to the conjunctiva and Tenon's capsule, the absence of deeper invasion intraoperatively, and the use of excision followed by systemic chemotherapy, the present case is most consistent with localized disease, although precise IRSG grouping (Intergroup Rhabdomyosarcoma Study Group))would depend on margin status and formal metastatic staging. If the lesion was completely excised with no residual disease, it would be compatible with Group I; if

microscopic residual disease remained, Group II would be more appropriate.[9]

This case is important because rhabdomyosarcoma in Pakistan has been reported to uncommon and often associated with late presentation and poor prognosis. Karachi registry data identified only 10 ocular/orbital cases between 1998 and 2002, while a North-West Pakistan case series reported just 12 orbital cases over 5 years, underscoring the rarity of ophthalmic RMS and the importance of early recognition of unusual conjunctival presentations.[10] Clinicians should therefore include rhabdomyosarcoma in the differential diagnosis of any atypical or rapidly evolving conjunctival lesion in a child, in order to ensure timely multidisciplinary treatment and close follow-up.

CONCLUSION

Conjunctival rhabdomyosarcoma is an exceptionally rare presentation of a common pediatric soft tissue sarcoma. This case underscores the importance of considering RMS in the differential diagnosis of conjunctival lesions in children to enable timely diagnosis and effective treatment.

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Conflict of Interest: Authors declared no conflict of interest.

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