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Case Report

Bulbar conjunctival plexiform schwannoma in a 5-year-old patient; expect the unexpected!

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Abstract

Rare, atypical ophthalmological conditions in adults include bulbar conjunctival plexiform schwannomas, which are usually asymptomatic. Few case reports in the literature indicate the presence of orbital/conjunctival schwannomas in adult patients and, rarely, among children under the age of 12. We report a case of a 5-year-old girl who presented in an outpatient clinic with inferior temporal conjunctival nonpigmented cystic lesion of a 10 × 10 mm size. Upon examination, we could not identify a feeding vessel. The mass was mobile and not fixed to the sclera. The history indicated a 1-year duration but the mass in the left eye had progressively increased in size during the last 2 months prior to presentation. There was no traumatic injury or past history of ophthalmic surgery. Surgical excision of the cyst was sucessfully performed, and histopathological examination confirmed bulbar conjunctival plexiform schwannoma diagnosis. Upon regular follow-up evaluation, there was no evidence of recurrence or malignant transformation. Although it is extremely rare to encounter conjunctival schwannomas in children, it should be considered in ovoidal well-circumscribed orbital swellings, particularly those that appear with no history of trauma or surgery to the eye. Surgical excision is effective and safe therapeutic intervention.

Plain language summary

A 5-year-old girl consulted our ophthalmology clinic for an unusually large swelling in her left eye. It had been increasing in size over the last 2 months before the consultation. She reported no injury or surgery at the swelling site. Apart from the mass, no other abnormality was noted. The family agreed to the surgical removal of the mass, which was performed. Pathological examination revealed a rare tumor called a bulbar conjunctival plexiform schwannoma. This case report is of interest as this type of tumor is rare in children. This case report confirms that surgical removal of these tumors is safe, feasible, and effective as a treatment for this rare condition.

Keywords: bulbar conjunctival plexiform schwannomas, conjunctival cyst, none pigmented cyst

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Introduction

Bulbar conjunctival plexiform schwannomas are considered a group of unusual and ocular malformations. Although they are rare types of clinically diverse peripheral nerve sheath tumors, they are even rarer among pediatric populations. It is well known that orbital schwannomas involve the head and neck in the majority of cases; however, they rarely occur within the orbital cavity.[1] Patients with neurofibromatosis commonly develop other forms of peripheral nerve sheath tumors that include neurofibromas and malignant peripheral nerve sheath tumors. Orbital involvement is seen in 11–28% of patients with neurofibromatosis-1

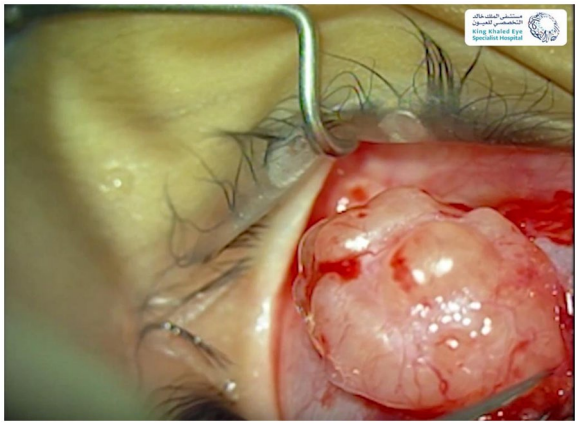

Figure 1. Macroscopic examination revealed a lobular whitish conjunctival mass in the left eye.

or a family history of neurofibromatosis; however, the risk of specifically developing orbital schwannoma within those populations is 1.5%.[2] Benign schwannomas usually present between the second and sixth decade of life, and there has been no racial association identified.[3] These tumors rarely undergo malignant transformation.[4] Surgical approach for the treatment of orbital schwannomas depends largely on the tumor location; therefore, variation in size and tumor location often creates unique therapeutic challenges.

Although several case reports described the occurrence of orbital schwannomas among adults[5,6] and children over 12,[7,8] it is rare to encounter Schwann cell hyperplasia among the under-fives. We present the first case of pediatric conjunctival schwannoma that responded favorably to surgical excision. We have followed the CARE guidelines during the writing up of the current case report.

Case report

We report a case of a five-year-old girl who presented in an outpatient clinic with left eye inferior temporal conjunctival cystic lesion of a 10 × 10 mm size, and it was not pigmented. Upon examination, we could not identify a feeding vessel. The conjunctival mass was mobile and not fixed to the sclera. The history indicated a one-year duration but the mass in the left eye had progressively increased in size during last 2 months prior to presentation. There was no traumatic injury or past history of ophthalmic surgery. A systemic evaluation revealed no abnormalities.

Clinical findings

Examination on presentation showed left eye inferionasal conjuctival mass in the fornix without redness, normal visual acuity of 20/20. In addition, there were full extra ocular movements.

Furthermore, no evidence was found for strabismus, nystagmus, or abnormal head posture. Cyclorefraction was +3.00 – 1.00 × 180 in both eyes. The slit lamp examination of the both eyes were within normal range, and the fundus examination of both eyes showed a healthy disc and flat retina in both eyes.

Ultrasonographic microscope imaging of the left eye showed a large conjunctiva lesion detected inferior nasal with no clear invasion to the sclera (see Figure 1).

Diagnostic assessment

Upon *in situ* macroscopic assessment, there was a sizable conjunctival mass in the inferionasal fornix in the left eye. It consisted of lobular whitish tan firm tissue, and upon sectioning, the cut surface was white smooth with a gross lobular pattern.

The main differential diagnoses were pyogenic granuloma, post-traumatic conjunctival cyst, conjunctival lymphoma or amelanotic melanoma. There were no systematic inflammatory symptoms to suggest a diagnosis of the former or history of trauma to suggest the latter. Furthermore, both conditions were duly excluded by histopathological examination of the tissue biopsy.

Therapeutic intervention

Surgical excision of the cyst was performed, and microscopic histopathological examination was conducted using Hematoxylin and Eosin stain. It confirmed bulbar conjunctival plexiform schwannoma diagnosis, left conjunctiva.

Follow-up and outcomes

The patient's eye appearance was noted to be normal at subsequent follow-up visits following the removal of the conjunctival mass. There were no signs of recurrence or malignant transformation.

Patient perspective

The family was willing and agreeable to the joint decision of surgical removal for the mass for diagnosis and cosmesis. They were content with the outcome.

Discussion

We present this pediatric case for a conjunctival mass that was open to a wide range of differential diagnoses. Clearly, the absence of a history for trauma and previous surgery ruled out the possibility of an inclusion cyst. Confirmation of the diagnosis was by microscopic histological investigation, the bulbar conjunctival plexiform schwannoma.

Schwannoma is caused by Schwann cells' hyperplasia. Schwann cells are responsible for the production of myelin sheath in the peripheral nervous system. It is usually a slowly growing ovoidal-shaped and well-demarcated swelling and constitutes 1% of all tumors that occur in the orbital area.[9]

The molecular etiology of Schwann cells' hyperplasia is poorly understood; however, it is thought to bear a close resemblance to the pathological processes that lead to the development of neurofibromatosis 1 and 2 NF-1 and NF-2. In NF-1, Schwann cell hyperplasia is promoted by unbridled Ras gene signal transduction related to biallelic inactivation neurofibromin on 17q11.2, believed to play a vital role in tumor suppression.[10] Similarly, in NF-2, Schwann cell hyperplasia is enhanced with deficiency of the Merlin gene (22q11.2).[11]

Our current case of a pediatric congenital bulbar conjunctival schwannoma is an extremely rare finding. In the adult population, we found several case reports of schwannomas within the globe, with infiltration of the ciliary body,[12,13] choroid,[14] iris,[15] sclera,[16] and posterior ciliary nerve.[17] It is quite rare for the involvement of optic nerve by Schwann cells hyperplasia,[18,19] which has been attributed to autonomic perivascular nerves surrounding the nerve sheath.[20]

Our patient responded favorably to surgical excision. This is in line with the existing literature that recommends excision for conjunctival schwannomas, given their solid consistency and easy separation from adjoining tissue.[21] There are no reports of recurrence or malignant transformation. Cryotherapy would be indicated only should the sclera be involved or malignancy is suspected.[21]

This current case report is a once-off observation, which should be considered before generalizing our results to patients of different demographic and clinical characteristics. We recommend follow-on research of longitudinal nature for how patients progress after surgical excision. Also, genetic and molecular underpinnings of conjunctival schwannoma require focused laboratory and clinical investigation.

Conclusion

Although it is extremely rare to encounter conjunctival schwannomas in children, it should be considered in ovoidal well-circumscribed orbital swellings, particularly those that appear with no history of trauma or surgery to the eye. Surgical excision is an effective and safe therapeutic intervention, even in very small children.

Declarations

Ethics approval and consent to participate

Ethical approval was obtained from the Research and Ethics Committee affiliated at King Khalid Eye Specialist Hospital (RD/26001/IRB/0158-22). Informed consent to participate was obtained from the patients' parents at the time of treatment.

Consent for publication

The parents of our patient provided informed consent for the publication of her case findings and the images presented.

Author contributions

Mona Kenani: Methodology, Writing – original draft.

Rafaa Babgi: Supervision, Writing – review & editing.

Sultan Bakri: Conceptualization, Writing – review & editing.

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Competing interests
The authors declare that there is no conflict of interest.

Availability of data and materials
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Supplemental material
Supplemental material for this article is available online.

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