Case Report

Schwannoma of the Caruncle: A Rare Tumour

Khushdeep Abhaypal,[1] Manpreet Singh,[1] Manu Saini,[1] Kirti Gupta,[2] Rajneesh Dhiman,[1] Pankaj Gupta[1]
[1]Department of Ophthalmology, Post Graduate Institute of Medical Education and Research, Chandigarh, India;
[2]Department of Histopathology, Post Graduate Institute of Medical Education and Research, Chandigarh, India

ABSTRACT

Introduction: Schwannomas, typically benign, encapsulated tumours arising from peripheral nerve sheaths, are commonly found in the orbit but are exceptionally rare in the caruncle. Here, a unique case of caruncular schwannoma is reported in an adolescent male who presented with gradual, painless swelling over a two-year period. This rarity emphasises the need to consider schwannomas in the differential diagnosis of caruncular masses.

Case: An 18-year-old male presented with a two-year history of progressively enlarging swelling adjacent to the right lacrimal caruncle, without associated symptoms or trauma. The mass was completely excised under local anaesthesia.

Observation: Histopathological analysis revealed a well-defined tumour consisting of spindle cells. Immunohistochemical staining for S-100 was strongly positive, confirming the diagnosis of schwannoma. This case is the first documented instance of caruncular schwannoma in a young individual, contributing to the limited understanding of this condition.

Conclusion: This case highlights a rare presentation of caruncular schwannoma in an adolescent male, marked by a painless, progressively enlarging mass adjacent to the right lacrimal caruncle over a two-year period. Complete excision under local anaesthesia allowed for histopathological confirmation of schwannoma, with immunohistochemical positivity for S-100. Post-operative healing was uneventful, providing an excellent cosmetic result with no recurrence observed at the two-year follow-up.

Key words: Caruncle; caruncular mass; lacrimal caruncle; neurilemmal tumour; neurilemmoma; schwannoma.

Financial Interest : Nil
Conflict of Interest : Nil

Received : 10.12.2023
Accepted : 25.01.2025

Corresponding Author
Dr. Pankaj Gupta,
Professor, Department of Ophthalmology, Advanced Eye Centre,
Post Graduate Institute of Medical Education and Research,
Chandigarh, India.
E-mail: drpankajkgupta@gmail.com

Access this article online
Website: www.nepjol.info/index.php/NEPJOPH
DOI: https://doi.org/10.3126/nepjoph.v16i2.59457

ISSN: 2072-6805, **E-ISSN:** 2091-0320

INTRODUCTION

Schwannoma, also known as a neurilemoma or neurilemmoma, is a benign tumour of the peripheral nerve sheath originating from Schwann cells. Schwannomas can manifest sporadically or in conjunction with conditions such as neurofibromatosis or schwannomatosis (Mrugala et al., 2005). In a series by Das Gupta et al., (1969) consisting of 303 patients with benign schwannoma, approximately 45% of tumours involved the head and neck. Schwannomas account for approximately 1% of orbital tumours, with a predilection for the superior orbit, reflective of their origin from cranial nerve branches traversing the superior orbital fissure (Sheilds et al., 2004). Other unusual locations include the eyelids, choroid, sclera, and conjunctiva (Le Marc'hadour et al., 1996). Notably, schwannoma of the caruncle is exceedingly rare, with only a single prior case documented in a 75-year-old female (Rennie et al., 1991). Here, a unique case of caruncular schwannoma is reported occurring in a young male patient, broadening the spectrum of potential diagnoses for caruncular lesions.

CASE REPORT

An 18-year-old male, previously healthy, presented with a painless mass in the medial canthal area of his right eye that had been progressively enlarging over a two-year period. The patient had no history of antecedent trauma or similar previous episodes. The mass was devoid of associated symptoms such as redness, watering, itching, or bleeding, and the patient had an unremarkable systemic medical history. On examination, the visual acuity was 6/6 in both eyes. The eyelids and periocular skin were normal. A small, fleshy, oval-shaped lesion measuring 5 x 4 x 4 mm was noted to arise from the right lacrimal caruncle (Figure 1). The rest of the ocular and systemic examinations were within normal limits. Nevi, being the most common caruncular lesion, was kept as the first possibility in this case.

The patient underwent complete excision of the mass under local anaesthesia. Intraoperatively, the mass was seen arising from the lacrimal caruncle without any underlying adhesions and was removed in toto. Histopathological examination revealed a well-circumscribed, encapsulated

Figure 1: Patient image with the arrow pointing towards the oval, fleshy lesion arising from the right lacrimal caruncle.

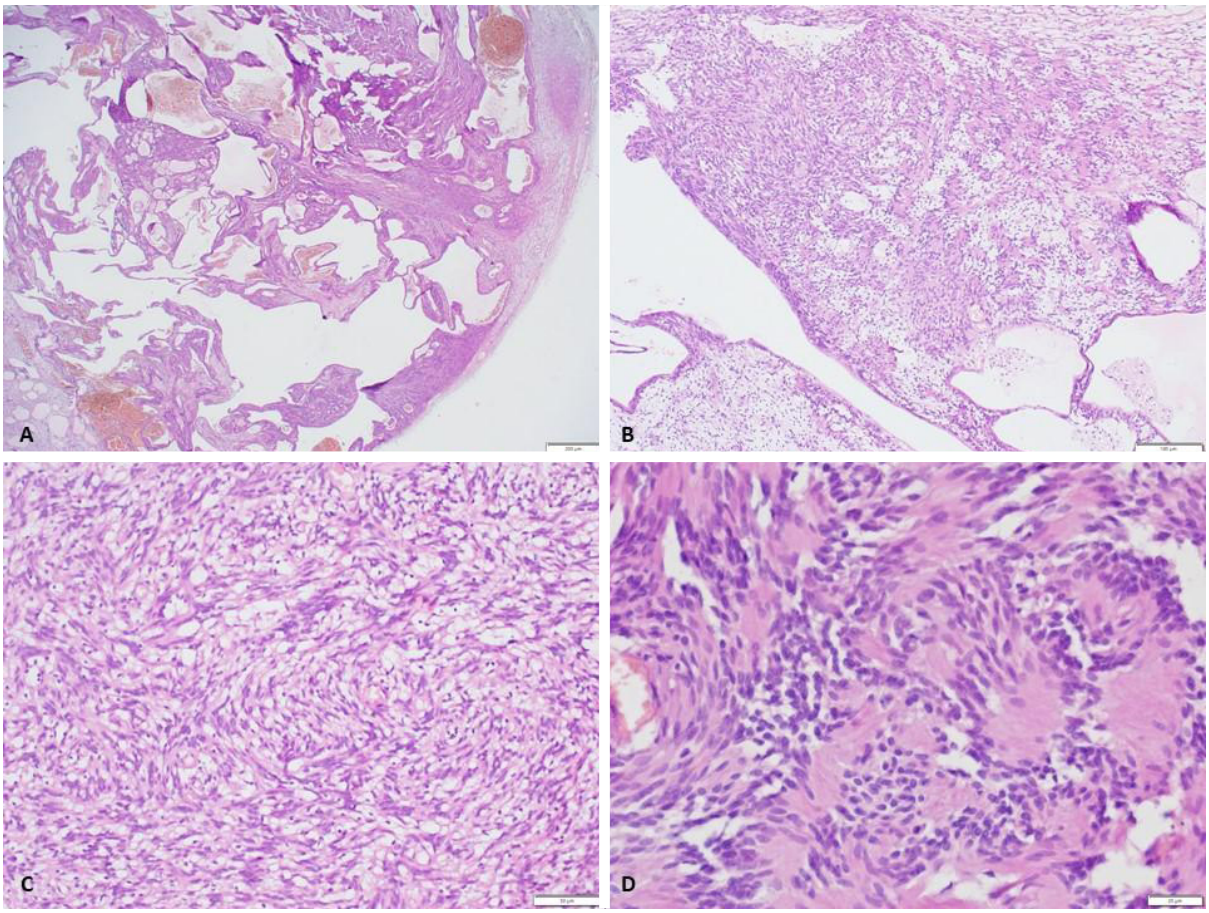

Figure 2: Histopathological examination of the caruncular tissue with hematoxylin-eosin stain; A, Scanner image depicting circumscribed lesion (scale bar 200µm); B, Low magnification depicting hypo and hypercellular areas interspersed with cystic areas (scale bar 100µm); C, High magnification depicting plump fusiform flattened endothelial cells (scale bar 50µm); D, Palisaded nuclei forming Verocay bodies (scale bar 20µm).

tumour (Figure 2A) predominantly composed of spindle cells arranged in alternating cellular and hypocellular patterns (Figure 2B). The spindle cells were monomorphic and arranged in interlacing fascicles, showing regimentation. These fusiform cells, characterised by flattened endothelial morphology (Figure 2C), featured bland nuclei with nuclear kinking, giving rise to the formation of Verocay bodies (Figure 2D). Additionally, the tumour displayed a moderate amount of eosinophilic cytoplasm interspersed with capillary channels. Immunohistochemical staining unequivocally confirmed the diagnosis of schwannoma by demonstrating strong positivity for the S-100 protein marker.

The post-operative healing was uneventful, and the patient had an excellent cosmetic outcome with no recurrence at two years follow up.

DISCUSSION

The caruncle, a distinctive, small, pinkish-red triangular mass situated in the medial canthus of the eye, is composed of conjunctival, cutaneous, and lacrimal elements. It encompasses sebaceous and sweat glands, as well as hair follicles, all enveloped by stratified squamous epithelium. It is innervated by the infratrochlear nerve and has a dense blood supply from the superior medial palpebral artery (Duke-Elder and Wybar, 1961). The caruncle is prominently located within the palpebral aperture, making it readily visible, and any alterations in its appearance are readily detectable. Lesions originating in the caruncle represent 1% of excised conjunctival lesions (Levy et al., 2009). The caruncle can be affected by an array of benign and malignant conditions, including cysts, papillomas, nevi, lymphomas, and adenocarcinomas. The rare and histopathologically diverse nature of caruncular lesions can pose a diagnostic challenge.

Melanocytic lesions, particularly nevi, constitute the predominant category of caruncular lesions (Kaeser et al., 2006; Santos and Gómez-Leal, 1994; Luthra, 1978). Previous comprehensive reviews of caruncular masses have consistently reported no cases of caruncular schwannoma. A review of 42 caruncular masses by Levy et al. (2009) revealed no cases of caruncular schwannoma. In a 16-year review of 31 caruncular lesions, melanocytic nevi were most common, followed by benign squamous papillomas; however, there was no case of a caruncular schwannoma (Yvon et al., 2022). A 20-year study that reviewed caruncular lesions found that epithelial inclusion cysts and sebaceous gland carcinoma were the most common benign and malignant lesions respectively among patients who underwent surgical excision (Alam et al., 2022). Schwannomas most commonly originate from the cranial and spinal nerve roots, with origins from peripheral nerves being relatively uncommon (Charles et al., 1997). Schwannomas of the lacrimal caruncle are extremely rare, with only one case being reported in the literature in a 75-year-old female (Rennie et al., 1991). In the present case, the most plausible origin of the schwannoma was deduced to be the branches of infratrochlear nerves supplying the caruncle.

Schwannomas are typically solitary tumours. The presence of multiple schwannomas in a patient often indicates an underlying syndromic association, which may include neurofibromatosis type 2 (NF2), schwannomatosis, or the Carney complex. Both NF-2 and schwannomatosis are autosomal dominant conditions caused by mutations in NF2 gene, SMARCB1 and LZTR1 genes respectively. NF2 is distinguished by the characteristic presence of vestibular schwannomas, which are not observed in schwannomatosis. The genetic predisposition to multiple schwannomas highlights the significance of genetic counselling and testing for affected individuals and their families. This facilitates early intervention and effective management of these potentially debilitating tumours.

The Carney complex is an autosomal dominant multiple neoplasia syndrome consisting of myxomas, spotty pigmentation, endocrine overactivity, and schwannomas. Notably, schwannomas associated with the Carney complex are recognised as psammomatous melanotic schwannomas, displaying characteristic psammoma bodies and melanin

on histopathology (Carney, 1995). Therefore, the diagnosis of Carney complex can be readily excluded by thorough pathological examination. Recently, conjunctival schwannoma has been described in association with multiple endocrine neoplasia type 2b, which is usually associated with mucosal neuromas that stain positive for neurofilament protein (Mirzayev et al., 2020).

The clinical presentation of caruncular schwannoma closely mirrors that of other caruncular lesions, featuring painless swelling, redness, and irritation. The definitive diagnosis of schwannoma is confirmed through histopathological examination of the excised mass. The histopathological features of schwannomas include a well-circumscribed, encapsulated mass consisting of spindle-shaped cells arranged in a characteristic pattern called Antoni A and Antoni B areas. Antoni A areas are composed of densely packed spindle-shaped cells that form a palisading arrangement around cystic spaces. These cells have elongated nuclei and eosinophilic cytoplasm. Spindle cells frequently exhibit nuclei regimentation, forming Verocay bodies. In contrast, Antoni B

Table 1: Depicting histopathological features of common caruncular lesions.

Origin	Type	Histopathological Features
1) Epithelial lesion	a) Epithelial inclusion cyst	Cyst wall lined by stratified squamous epithelium without keratinisation of inner layers
	b) Eccrine cyst/ hidrocystoma	Subepithelial location, lined by keratinised stratified squamous epithelium and filled with keratin material
	c) Sebaceous adenoma	Mature sebaceous lobules around a central duct
	d) Intraepithelial dysplasia	Intraepithelial dysplasia occupying lower third to half of epithelium with varying degree of hyperplasia but with intact basement membrane
	e) Basal cell carcinoma	Subepithelial nests of basaloid cells with peripheral palisading
	f) Squamous cell carcinoma	Irregular sheets and islands of dysplastic cells and scattered individual cells show keratinisation
	g) Oncocytoma	Benign epithelial cells with abundant eosinophilic granular cytoplasm
2) Melanocytic lesion	a) Nevus	Subepithelial nests of nevoid cells with increase in the basal layer melanocytes
3) Inflammatory lesion	a) Chronic non-specific	Inflammatory cells infiltration rich in lymphocytes with foci of granulation tissue
	b) With epithelial hyperplasia	Chronic non-specific inflammation with overlying epithelial hyperplasia
	c) Chronic granulomatous	Scattered epithelioid cells, poorly formed granulomas, with mixed inflammatory cells infiltration including multinucleated foreign body type giant cells
4) Lymphoid lesion		Monotonous population of small to medium sized lymphoid cells with scattered histiocytes

areas exhibit lower organisation, comprising less densely packed spindle-shaped cells with less well-defined cytoplasmic borders. Immunohistochemical staining of schwannomas typically shows strong positivity for S100 protein, which is a marker of Schwann cells (Mirzayev et al., 2020).

Malignant schwannomas have a less well-defined histopathological appearance, and may show features such as high cellularity, nuclear atypia, and increased mitotic activity. These tumours may also show loss of S100 protein expression and increased expression of other markers, such as p53 and Ki-67. The histopathological features of common caruncular lesions have been tabulated (Table 1).

Complete excision represents the treatment of choice for caruncular schwannoma. Despite the predominance of benign caruncular lesions, any alterations in appearance, including changes in color, size, or vascularisation, warrant prompt intervention and excision. When performing procedures in proximity to the caruncle, caution should be exercised to prevent inadvertent damage to the canalicular system. This is particularly significant, as the external surface of the caruncle is separated from the common canaliculus by an average distance of 0.85 mm, with the shortest recorded distance being 0.50 mm. Recurrence after complete excision is rare, and to the best of authors' knowledge, no case of malignant transformation has been reported in conjunctival or caruncular schwannoma.

CONCLUSION

Here a case of schwannoma originating in the caruncle has been presented, which broadens the range of possible diagnoses for lesions in that area. Histopathology with immunohistochemistry clinches the diagnosis, with complete excision offering a cure in most cases. Additionally, it is prudent to examine the patient, along with family members, to rule out genetic component in every case of solitary schwannoma.

REFERENCES

Agarwal R, Pal S, Chaudhary N, et al., (2019). Limbal schwannoma: Clinicopathological study of a case with review of literature. Oman Journal of Ophthalmology; 12(3): 197-199. DOI: 10.4103/ojo.OJO_81_2018 PMID: 31902998

Alam MS, Pal SS, Krishnakumar S (2022). Clinical and histopathological profile of primary caruncular lesions. Indian Journal of Ophthalmology; 70(6): 1975-1981. DOI: 10.4103/ijo.IJO_2724_21 PMID: 35647965

Carney JA (1995). Carney complex: The complex of myxomas, spotty pigmentation, endocrine overactivity, and schwannomas. Seminars in Dermatology; 14(2): 90-98. DOI: 10.1016/s1085-5629(05)80003-3 PMID: 7640202

Charles NC, Fox DM, Avendaño JA, et al., (1997). Conjunctival neurilemoma. Report of three cases. Archives of Ophthalmology; 115(4): 547-549. DOI: 10.1001/archopht.1997.01100150549020 PMID: 9109769

Das Gupta TK, Brasfield RD, Strong EW, et al., (1969). Benign solitary schwannomas (neurilemomas). Cancer; 24(2): 355-366. DOI: 10.1002/1097-0142(196908)24:2<355::aid-cncr2820240218>3.0.co;2-2 PMID: 5796779

Duke-Elder S, Wybar KC (1961). The anatomy of the visual system. In Duke-Elder S, ed. System of ophthalmology, vol 2. London, United Kingdom: Henry Kimpton, 1961:557-8.

Kaeser PF, Uffer S, Zografos L, et al., (2006). Tumours of the caruncle: A clinicopathologic correlation. American Journal of Ophthalmology; 142(3): 448-455. DOI: 10.1016/j.ajo.2006.04.035 PMID: 16935590

Le Marc'hadour F, Romanet JP, Fdili A, et al., (1996). Schwannoma of the bulbar conjunctiva. Archives of Ophthalmology; 114(10): 1258-1260. DOI: 10.1001/archopht.1996.01100140458019 PMID: 8859089

Levy J, Ilsar M, Deckel Y, et al., (2009). Lesions of the caruncle: A description of 42 cases and a review of the literature. Eye (London, England); 23(5): 1004-1018. DOI: 10.1038/eye.2008.316 PMID: 18927596

Luthra CL, Doxanas MT, Green WR, (1978). Lesions of the caruncle: a clinicohistopathologic study. Survey of Ophthalmology; 23(3): 183-195. DOI: 10.1016/0039-6257(78)90155-8 PMID: 216128

Mirzayev I, Gündüz AK, Cansiz Ersöz C, et al., (2020). Anterior segment optical coherence tomography, in vivo confocal microscopy, histopathologic, and immunohistochemical findings in a patient with multiple endocrine neoplasia type 2b. Ophthalmic Genetics; 41(5): 491-496. DOI: 10.1080/13816810.2020.1795891 PMID: 32718216

Mrugala MM, Batchelor TT, Plotkin SR (2005). Peripheral and cranial nerve sheath tumours. Current Opinion in Neurology; 18(5):604-10. DOI: 10.1097/01.wco.0000179507.51647.02 PMID: 16155448

Rennie IG, Parsons MA, Benson MT, (1991). Neurilemoma of the caruncle: A clinicopathological report. British Journal of Ophthalmology; 75(12): 749-751. DOI: 10.1136/bjo.75.12.749 PMID: 1768668

Santos A, Gómez-Leal A, (1994). Lesions of the lacrimal caruncle. Clinicopathologic features. Ophthalmology; 101(5): 943-949. DOI: 10.1016/s0161-6420(94)31233-4 PMID: 8190485

Shields JA, Shields CL, Scartozzi R, (2004). Survey of 1264 patients with orbital tumours and simulating lesions: The 2002 Montgomery Lecture, part 1. Ophthalmology; 111(5):997-1008. DOI: 10.1016/j.ophtha.2003.01.002 PMID: 15121380

Yvon C, Balamurugan T, Bianchi E, et al., (2022). Caruncular lesions: A 16-year single centre review in the United Kingdom. European Journal of Ophthalmology; 10:11206721221137938. DOI: 10.1177/11206721221137938 PMID: 36366790