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Brief Report

Solitary eyelid neurofibroma presenting as tarsal cyst: Report of a case and review of literature

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ABSTRACT

Purpose: To report a rare case of solitary eyelid neurofiboma presenting as tarsal cyst.
Observation: A 64 year old male, presented with a painless, non progressive swelling in the right upper eye lid. Examination revealed a non tender, firm, mass adherent to the underlying tarsus. Excision via a conjunctival approach was performed and the histopathology was suggestive of neurofibroma. Immunohistochemistry was positive for Vimentin and focally positive for S-100. The patient did not have any features of neurofibromatosis.
Conclusion: and Importance: Solitary neurofibroma of the eyelid does not have any systemic association with neurofibromatosis, and only seven such cases have been reported in English literature. We report a case of solitary neurofibroma of the eyelid presenting as tarsal cyst. A thorough review of literature of previously reported cases is included.

1. Introduction

Neurofibromas are benign peripheral nerve sheath tumors arising from the non myelinated Schwann cells, perineural fibroblasts or both. Neurofibromas can present as a solitary lesion or in association with systemic neurofibromatosis, which is often referred to as Von Recklinghausen disease. They are commonly seen over the trunk, head and neck but can affect any organ, bone or soft tissue. Plexiform and diffuse types of neurofibromas are seen more commonly in the orbit and eyelid, compared to the solitary or localized. Solitary neurofibroma of the eyelid is extremely rare. A thorough Medline search shows only 7 published cases of solitary neurofibroma of the eyelid,.[1–3] We describe an unusual case of a solitary neurofibroma of the upper eyelid presenting as tarsal cyst in an adult male.

1.1. Case report

A 64 year old male, was referred to our oculoplasty clinic with a chief complaint of a painless, non progressive mass in the right upper lid present over a 3 years period. There was no history of redness or tearing. The patient denied any history of trauma or previous surgery. There was no history of any systemic illness or similar masses anywhere else on his body. On examination, his best corrected visual acuity was 6/36 in the right eye and 6/9 in the left eye. Slit lamp examination showed nuclear sclerosis in both eyes, which was more advanced in right eye. There was a firm non tender, non-mobile mass measuring 6 × 5 mm adherent to the posterior border of the underlying tarsus (Fig. 1A and B). The remainder of the ophthalmic and systemic examination was unremarkable. Based on the clinical findings, a differential diagnosis including chalazion and tarsal cyst were considered.

The mass was totally excised via conjunctival approach. Histopathological examination showed non capsulated, firm mass composed of multiple spindle shaped tumor cells with wavy nuclei and intervening collagen suggestive of neurofibroma (Fig. 2 A). Immunohistochemistry was done to confirm the diagnosis which showed strong positivity for vimentin and focal S-100 positive areas (Fig. 2 B & C).

There was no recurrence at 6 months of follow up.

2. Discussion

The cause of solitary neurofibromas is unknown. In most cases they have no sex predilection and are commonly seen in young adults.[4] However, amongst the published cases of solitary neurofibromas affecting the eyelid (Table 1), a strong female preponderance is seen (6, 85.71%) and the mean age at presentation was 69.85 ± 9.20years.[1–3] Our patient was an elderly male with involvement of the upper eyelid and the lesion had the appearance of a chalazion, similar to that

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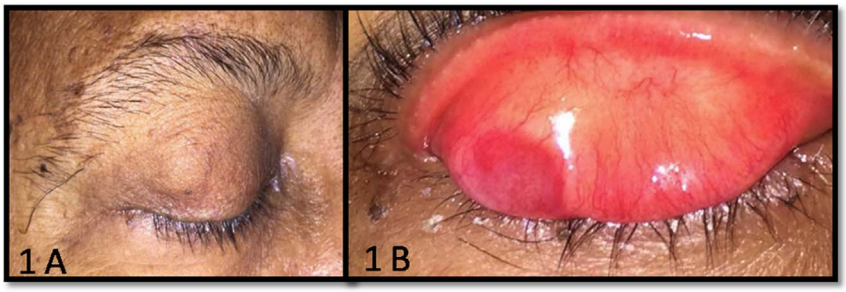

Fig. 1(A): External photograph of the patient showing well localized right upper lid mass.. (B): External photograph of the right everted eyelid showing the mass involving the tarsal plate.

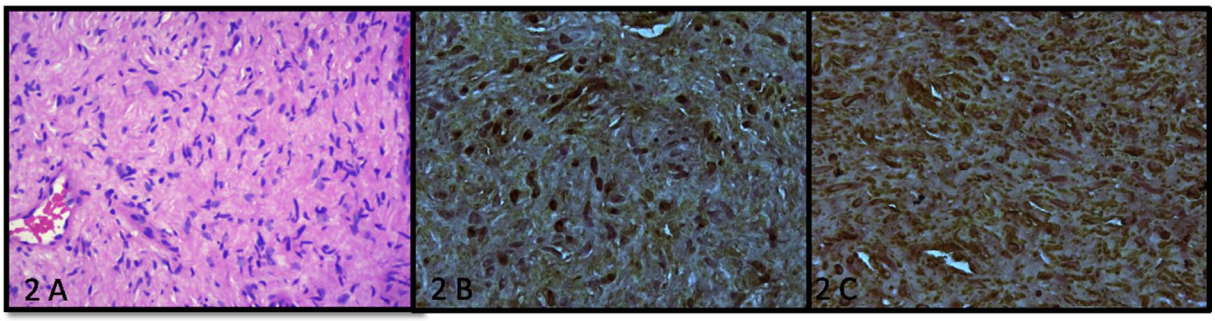

Fig. 2(A): Microphotograph (Hematoxylin & Eosin 20 X) showing benign looking spindle shaped tumor cells intermixed with collagen fibers. Tumor cells have wavy nuclei without atypia. Features suggestive of neurofibroma. (B): Immunohistochemistry image (40X) showing focal S-100 positive cells.. (C): Immunohistochemistry image (40X) showing diffuse positivity for Vimentin.

Table 1
Previous reported cases of solitary eyelid neurofibroma.

Author, year	Age (years)/sex	Site	Associated condition	Management	IHC
Amir Mohammadi et al., 2010	81/M	Upper lid	Basal cell carcinoma, Adenocarcinoma lung	Biopsy	S-100 positive
Nako Shibata et al., 2011	72/F	Upper lid	Past history of malignant lymphoma	Biopsy	*S*— 100 positive, CD34 positive
Anna Stagner et al., 2016	59/F	Lower lid	–	Excision biopsy	S-100 positive,
	57/F	Upper lid	–	Excision biopsy	CD 34 positive,
	76/F	Lower lid	–	Excision biopsy	Glut-1 and EMA negative
	77/F	Lower lid near medial canthus	–	Excision biopsy	
	67/F	Lower lid	–	Excision biopsy	
Nisar S et al., 2016 (Present study)	64/M	Upper lid	–	Excision biopsy	Vimentin positive, S-100 focally positive

reported by Shibata Nako et al.[2] Stagner AM et al. noticed lower lid involvement in 4 of the 5 patients in their case series.[3]

No cases of solitary eyelid neurofibroma were reported in 649 cases of benign eye lid tumors studied by Sean Paul[4] et al., or 1541 cases studied by Gundogan FC et al.[5] A thorough review of literature and Medline search revealed only seven cases of solitary neurofibroma of the eyelid reported (Table 1).[1–3] None of the patients had any of the features of systemic neurofibromatosis. Complete excision and immunohistochemistry confirmed the diagnosis in all cases.

Mohammadi A et al. have reported a case of solitary eyelid neurofibroma with co existing basal cell carcinoma.[1] This patient was also found to have an adenocarcinoma of the lung which eventually proved fatal. Shibata Nako et al. reported that their patient had a past history of treatment for diffuse large B cell lymphoma.[2] Mutations in NF-1 gene in Neurofibromatosis 1 results in loss of neurofibromin which keeps the Ras proto oncogene in its inactive form. This loss of neurofibromin predisposes the patient to many neural and other systemic malignancies.[6] Whether the same holds true for solitary neurofibroma is not known, but it is advisable to evaluate all these patients systemically.

Histopathology of a solitary eyelid neurofibroma is similar to that of solitary neurofibromas seen elsewhere. These tumors are non-encapsulated, well circumscribed and composed of spindle shaped cells with fusiform or wavy nuclei, and a mixture of Schwann cells, endoneurial fibroblasts and perineural like cells within a collagen and connective tissue matrix.[5,7]Schwann cells stain positive for S-100 protein whereas endoneurial fibroblasts and perineural cells do not. Intervening fibroblasts show partial CD34 positivity. Neurofibromas are negative for epithelial membrane antigen and keratin.[8]

One can differentiate schwannomas from neurofibromas based on histopathological examination and immunohistochemistry. The Antoni A and Antoni B pattern is classically seen on histopathological examination in schwannomas. Schwannomas show strong positivity for S-100 protein marker, CD 56 and calretinin, they may or may not stain positive for CD 34. Neurofibromas are weakly positively for S-100 and CD 34, faintly positive for neurofilament and may or may not stain positive for calretinin.[3,7,8]

Solitary neurofibromas are benign and can be observed however, when indicated, a complete excision of the neurofibroma is curative. Incomplete excision may result in recurrence. Though malignant transformation is extremely rare, Krol EM et al. have reported malignant transformation in a case of recurrent solitary neurofibroma.[9]

3. Conclusion

Solitary neurofibroma is a rare entity and very few cases are reported in literature. Since it may be a precursor to systemic neurofibromatosis a thorough systemic evaluation is mandatory. Recurrence, malignant transformation and associated systemic malignancy have been reported in literature. Thus, a regular follow up and routine systemic evaluation is indicated. Ophthalmologists should be aware of its occurrence and include neurofibroma in theirs differential for eyelid tarsal cyst.

Patient consent

Patient gave written consent to publish his photograph and case history for scientific purpose, even if the photograph is identifiable.

Disclosures

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Conflicts of interest

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Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

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