Brief Report

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

Nisar Sonam Poonam, Md Shahid Alam, Dipankar Das, Jyotirmay Biswas

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 swelling 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, strong positivity for vimentin and focal S-100 positive areas (Fig. 2B & 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 ± 9.20 years.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 of...
reported by Shibata Nako et al. Stagner AM et al. noticed lower lid involvement in 4 of the 5 patients in their case series. No cases of solitary eyelid neurofibroma were reported in 649 cases of benign eye lid tumors studied by Sean Paul et al., or 1541 cases studied by Gundogan FC et al. A thorough review of literature and Medline search revealed only seven cases of solitary neurofibroma of the eyelid reported (Table 1). 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. 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. 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 malignacies. 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. Schwann cells stain positive 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.

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.

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.
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 their 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

Funding

No funding or grant support.

Conflicts of interest

The following authors have no financial disclosures.

Poonam NS, Alam MS, Das D, Biswas J.

Authorship

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

Acknowledgements

None.

References

1. Mohammadi A, Rosa M, Rhatigan R. Eyelid basal cell carcinoma associated with solitary neurofibroma. J Cutan Pathol. 2010;37:707–709.
2. Nako Shibata, Kitagawa K, Noda M, Sasaki H. Solitary neurofibroma without neurofibromatosis in superior tarsal plate simulating a chalazion. Graefes Arch Clin Exp Ophthalmol. 2012;250:309–310.
3. Stagner AM, Jakobiec FA. Peripheral nerve sheath tumors of the eyelid dermis: a clinicopathological and immunohistochemical analysis. Ophthal Mic Plast Reconstr Surg. 2015;32:40–45.
4. Paul S, Vo DT, Silikis RZ. Malignant and benign eyelid lesions in San Francisco: study of a diverse urban population. Am J Clin Med. 2011;8(1):40–46.
5. Gundogu FC, Yolu U, Tas A, et al. Eyelid tumors clinical data from an eye center in Ankara, Turkey. Asian Pac J Cancer Prev APJCP. 2015;16(10):4265–4269.
6. Yohay K. Neurofibromatosis type 1 and associated malignancies. Curr Neurol Neurosci Rep. 2009;9(3):247–253.
7. Rodriguez FJ, Folpe AL, Giannini C, Perry A. Pathology of peripheral nerve sheath tumors: diagnostic overview and update on selected diagnostic problems. Acta Neuropathol. 2012;123(3):295–319.
8. Jakobiec FA, Jones IS. Clinical ophthalmology. Neurogenic tumors. 1976;4(41):1–45.
9. Krol EM, El-Fanek H, Borruus J. Solitary neurofibroma with malignant transformation: case report and review of literature. Conn Med. 2015;79:217–219.