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Case Report Open Access

Removal of a Solitary Neurofibroma from the Eyelid Margin: Adding to the Existing Case-Based Literature

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Abstract

Neurofibroma is a benign tumor of peripheral nerve sheath origin which is usually found as part of neurofibromatosis type-1 (NF1). This case report describes the removal of a solitary neurofibroma from the upper eyelid margin of an asymptomatic 59-year-old female with no other neurofibromatosis type-1 (NF1) features. The patient presented with a painless, cyst-like lesion on the left upper lid margin. A cyst of Zeiss was clinically suspected and surgical excision under local anaesthesia was agreed. Histopathological analysis of the excised material stated that the sample contained bland fusiform cells with wavy nuclei arranged in fascicles. Cells were strongly positive for S100. These findings are compatible with a benign neurofibroma. A survey of the existing (English) literature suggests that there have been only nine cases of solitary neurofibroma of the eyelid previously reported, all of them occurring in the last eight years. Because of the possibility of the recurrence, the transformation to malignant lesion and the association of systemic malignancy, neurofibroma should be one of the differential diagnoses of any tarsal cyst and ophthalmologists should be aware of that.

Keywords: Neurofibroma Type 1; Bland fusiform cells; Benign tumor; Eyelid

Case Report

A 59-year old Asian female presented routinely to our oculoplastic clinic complaining of a lump on her left upper eyelid which had been present for around 12 months. It was painless and had not significantly grown in size over this time and there was no history of any eye lid surgery or trauma but it was causing her cosmetic upset. On examination under a slit lamp a round yellowish lump of approximately 1 mm by 2 mm was noted centrally on the margin of the left upper eyelid. Clinically, a cyst of Zeiss was suspected and surgical removal was agreed. In addition to that, the patient had bilateral mild keratoconus which was worse in the right eye, showing Vogt' s striae in the posterior stroma at the corneal apex. The left cornea was normal on slit lamp examination. The best corrected visual acuity is 6/9 in the right and 6/6 in the left. Intraocular pressure was measured as 15 mmHg in both eyes. The rest of the anterior segment examination was otherwise normal and there was no other relevant ocular history.

Excision of the lump was performed under local anaesthesia *via* a conjunctival approach after informed consent. During excision, the material which was excised from the lump was noted to be solid and nodular and not consistent with a cystic material. The lump was entirely removed and the material was sent for Histopathology.

Histopathological analysis of the material described a solid nodular proliferation in dermis, with a spindle cell pattern. These were bland fusiform cells with wavy nuclei arranged in fascicles. There was no evidence of malignancy or dysplastic change. The cells were strongly positive for s100 and negative for SMA with no cystic changes. These findings are strongly suggestive of benign solitary neurofibroma.

The patient was recalled for further history and examination where it was confirmed that she had no history of any other lumps anywhere on her body, or any skin changes consistent with café-au-lait spots or auxiliary freckles and no history of malignancy. Her irides were darkly pigmented with no evidence of any Lisch nodules. She had no family history of NF1 or any known family disorder of soft tissue tumors. Hence, a diagnosis of isolated solitary neurofibroma of the eyelid was made. Examination of her posterior segment in both eyes after dilation revealed a healthy disc and macula with moderate amount of mid-peripheral hard drusen and no other abnormality detected. Unfortunately, we did not take a photograph of the lesion before the surgical excision as the lump was thought to be a cyst of Zeiss. The patient presented for a follow up after one year and there was no evidence of recurrence of the lesion.

Discussion

Neurofibroma is one type of peripheral nerve sheath tumor (PNST); a group of mostly benign tumors arising from nerve sheaths in the peripheral nervous system [1]. They can be classified according to which part of the nerve sheath they arise from: schwannomas, perineurinomas or neurofibromas. Despite their non-cancerous nature, they may cause pain or be disabling, depending on their location [2]. These tumors are often found in the context of neurofibromatosis type-1 (NF1) but solitary cases are not uncommon. Malignant transformation is possible in a minority of cases; malignant peripheral nerve sheath tumors (MPNSTs) comprise 2%-10% of all sarcomas depending on the source [3,4]. Around 50% of MPNSTs are found in patients with NF1 and 50% are sporadically occurring [4]. Lifetime risk of developing MPNST if you have NF1 is now thought to be as high as 8%-13% [3]; however, there is no published data on the incidence of malignant progression amongst solitary neurofibromas.

Previously reported cases

According to the Table 1 and previously reported cases, isolated eyelid neurofibromas has no location predilection, as it was noticed to affect upper lid, lower lid, medial canthus, upper tarsal conjunctiva and in our case it affected the lid margin.

Although solitary neurofibromas are recognized, we have only been able to find a very small number of prior published instances of solitary neurofibroma occurring on the eyelid Table 1. A large 2009 retrospective study of 5,504 eyelid tumors identified only four neurofibromas in total; it did not specify whether or not the patients had NF1[5]. One histopathological study from 2016 managed to isolate ten patients with solitary, benign PNSTs of the eyelid; five of these were found to be neurofibroma [6]. A 2010 case report describes an 81-year-old male patient found to have a solitary neurofibroma of the eyelid adjacent to a basal cell carcinoma [7]. We found only three other reported cases of solitary neurofibroma of the eyelid in the absence of NF1; one from 2012 and two from earlier in 2018 [8-10]. This makes a total of nine previously reported cases of confirmed solitary neurofibroma of the eyelid, all occurring in the last eight years. We felt it appropriate to add this case report to the available literature since a total of ten recorded cases makes this rare. However, the recent increased frequency of reported cases may suggest that solitary neurofibroma of the eyelid is less uncommon than previously thought.

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, CD 34 positive, Glut-1 and EMA negative
	57/F	Upper lid	-	Excision biopsy	
	76/F	Lower lid	-	Excision biopsy	
	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

Table 1: Previous reported cases of solitary eyelid neurofibroma.

Conclusion

Solitary Eye lid neurofibroma is a rare condition. Ophthalmologists should be aware to include it in their differential diagnosis of any eyelid lump. The reason for that is the Neurofibroma might be the first presenting sign for systemic Neurofibromatosis. Furthermore, solitary eyelid neurofibroma might transform to malignant lesion or it might be associated with systemic malignancy.

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