Isolated neurofibroma of the eyelid mimicking recurrent chalazion

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Neurofibroma is a peripheral nerve sheath tumor which is seen in neurofibromatosis type 1 and is characterized by various ophthalmic manifestations. Solitary neurofibroma of the eyelid is rare. We report a case of a 53-year-old female patient who presented with a painless swelling in the left upper lid of 4 years' duration. She had undergone surgery for the same lesion twice. The lesion was excised and histopathological examination revealed a solitary neurofibroma. She did not have any other features of generalized neurofibromatosis.

Key words: Lid tumor, neurofibroma, recurrent chalazion

Neurofibromatosis type 1 (NF-1) is caused by a mutation in a gene located on chromosome 17. Although one-half of the patients inherit gene mutation in an autosomal dominant pattern from their parents, the other half present sporadically. Solitary neurofibromas, without any features of generalized NF, are rare. Eyelid tumors are usually benign and frequently masquerade as chalazion. In cases of recurrent chalazion, in addition to meibomian gland adenocarcinoma, neurofibroma should be considered as one of the differential diagnoses.

Case Report

A 53-year-old female patient presented with complaints of painless, gradually progressive mass in the left upper lid associated with drooping of the lid for the past 5 years [Fig. 1].

She had undergone surgery for the left upper lid mass twice, 3 years ago elsewhere. She had a history of previous surgery, which mentioned the procedure as chalazion incision and curettage (I & C). The swelling did not disappear. On examination, the best-corrected visual acuity was 6/6 N6 OU. There was a 15 mm round, well-circumscribed mass in the left upper lid not involving the lid margin. Eyelashes were intact.

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Rest of the anterior segment examination was normal. Anterior segment examination of the right eye was normal. Intraocular pressure by Goldmann's applanation tonometry was 12 mmHg OU. Fundus examination was normal OU. A diagnosis of recurrent chalazion was made.

The patient was planned for I & C of the lesion under local anesthesia. During the procedure, tarsus was found thickened. No fluid or granulomatous material was found. A small piece of tarsus was excised and sent for histopathology examination.

Histological examination revealed a tumor composed of fascicles and bundles of spindle-shaped cells with wavy-to-elongated nuclei, with tapered ends and eosinophilic cytoplasm surrounded by fibrocollagenous stroma with entrapped nerve bundle along with lymphocytes and few mast cells [Figs. 2 and 3]. Immunohistochemistry with S-100 showed diffuse positivity in tumor cells confirming the diagnosis of neurofibroma [Fig. 4].

When the patient came for review, she was examined in detail for other signs of NF. However, we found none. There was no family history of NF. Hence, a diagnosis of isolated solitary neurofibroma of the eyelid was made.

She underwent excision of the lesion with lid reconstruction by using Tenzel's flap method under general anesthesia [Fig. 5]. During the surgery, the lesion was found to be not very vascular.

Discussion

NF-1 (von Recklinghausen type 1) is inherited as autosomal dominant disease.[1] According to the diagnostic criteria for NF-1 as described by the National Institutes of Health in 1987, an individual has NF-1 if two or more of the following specific features are present – Café au lait spots on the skin, iris Lisch nodules, optic pathway glioma, freckles in the axilla or groin area, neurofibroma, plexiform neurofibroma, bony dysplasia, and a first-degree relative with NF-1.[2] Solitary

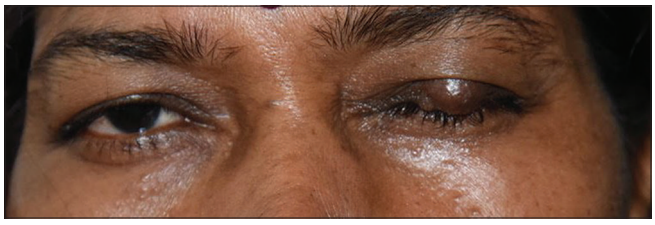

Figure 1: Clinical photograph showing a well-circumscribed round swelling in the left eye upper lid

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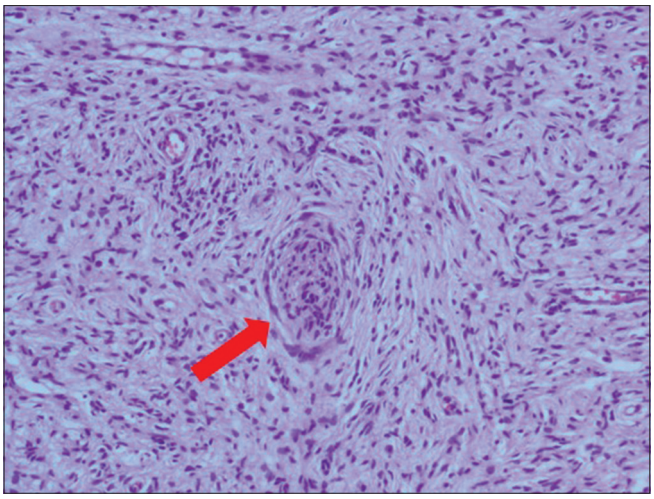

Figure 2: Microscopic section showing bundles of spindle-shaped cells with wavy-to-elongated nuclei, surrounded by fibrocollagenous stromal with entrapped nerve bundle (arrow) along with lymphocytes and plasma cells in (H and E, ×40)

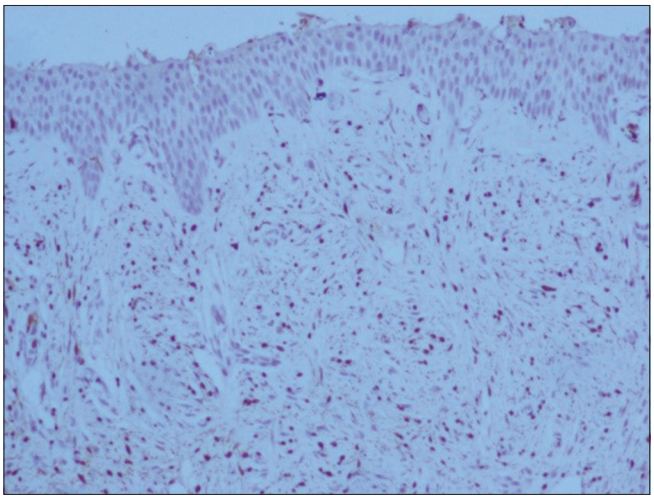

Figure 4: Immunohistochemistry with S-100 marker showing diffuse positivity of tumor cells

neurofibromas have been reported in unusual sites such as tongue, peritoneum, mandible, and palatine tonsil.[3]

In a retrospective study conducted by Ozdal *et al*. for assessing the accuracy of clinical diagnosis of chalazion, it was found that 6.4% of the cases were misdiagnosed as chalazion.[4] In a study conducted by Huang *et al*., majority of the eyelid tumors are benign.[5] Eyelid tumors of different origins such as sebaceous gland carcinoma and neurilemmoma can masquerade as a chalazion.[6,7]

In the present case, the solitary neurofibroma was seen involving the tarsal plate which differs from the article reported by Shibata *et al*., where the neurofibroma was seen beneath the tarsal plate, and two other cases reported by Chen *et al*. and Yeon Soo Kang, where the neurofibroma involved the eyelid margin.[3,8,9] Histopathology of the excised specimen in doubtful cases helps in the proper diagnosis as in our patient.

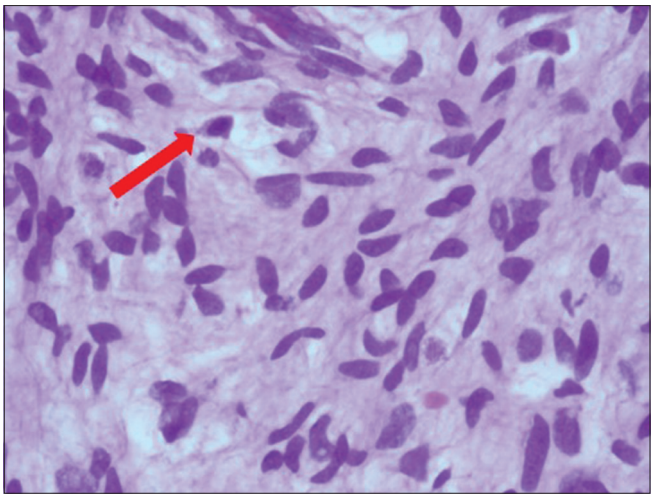

Figure 3: Microscopic section showing mast cell (arrow) interspersed with the tumor cells in (H and E, ×100)

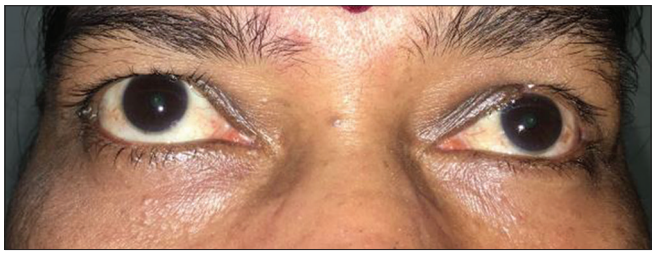

Figure 5: Postoperative photograph

Conclusion

We conclude that isolated neurofibroma can present like chalazion. A histopathological evaluation is important, especially for recurrent or atypical chalazion.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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