Case Report

Bilateral orbital isolated (solitary) neurofibroma in the absence of neurofibromatosis — A case report

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Abstract

Isolated neurofibroma is a slowly progressive tumor rarely found in the orbit accounting for less than 1% of the space occupying lesions of the orbit. It usually presents in the 2nd to 5th decades of life with proptosis, swelling, visual changes, ptosis, diplopia or pain. Almost all cases reported are unilateral. We report a 23-year-old female with no systemic features or family history of neurofibromatosis who presented with right upper eyelid swelling. Radiological studies revealed bilateral identical masses in the superior orbits. The patient underwent surgical excision of the lesion on the right side and it was proved to be neurofibroma. She did not have a recurrence with a follow up period of 2 years but developed sensory deficit. This is the fifth reported case of bilateral isolated neurofibroma.

Keywords: Neurofibroma, Neurofibromatosis, Orbit, Nerve

Introduction

Neurofibroma is a peripheral nerve tumor rarely found in the orbit accounting for less than 1% of the space occupying lesions of the orbit. It can occur in association with familial neurofibromatosis, however it can be found as a solitary lesion. Isolated (solitary) neurofibroma is an uncommon subtype. Garrity and Henderson had only 9 cases in their large series of peripheral nerve sheath tumors. All reported cases except 4 were unilateral. Herein we report a case of bilateral isolated orbital neurofibroma in the absence of neurofibromatosis.

Case report

A 23-year-old healthy female presented with non-progressive right upper eyelid painless swelling for 8 months. It was not associated with other ocular or visual complaint. The patient’s medical history and family history were unremarkable particularly for signs and symptoms of neurofibromatosis.

On Examination, a soft, non-tender, compressible, mobile mass was appreciated in the right upper eyelid. The Hertel exophthalmometer (at a base of 105) measured 22 mm on the right eye and 19 mm on the left eye with no changes...
during Valsalva’s maneuvers. The extraocular muscles movement was full. The superior fullness over the right upper eyelid was evident to the patient and disturbing for her cosmetically (Fig. 1A). Systemically, the patient did not also have any signs suggestive of neurofibromatosis.

Ultrasound B-scan was done on the left side and showed a cystic lesion suggestive of a unilateral dermoid cyst.

Orbital computed tomography (CT) showed a cylindrical shaped hypodense mass measuring approximately $4.4 \times 1 \times 1.3$ cm in the superior aspect of the right orbit causing an inferior displacement of the superior rectus muscle as well as the right globe. Incidentally, a similar, smaller, hypodense lesion was noted in an identical location within the superior aspect of the left orbit with similar inferior displacement of the left superior rectus muscle. The remaining extraocular muscles were preserved (Fig. 1B and C).

The lesion was excised through upper eyelid skin crease incision and superior orbitotomy. It appeared intraoperatively to be cystic in nature (Fig. 2A) arising in relation to the supra-orbital nerve and extending deeply along the course of the frontal sensory nerve into the superior orbital fissure (Fig. 2B).

Grossly, the tissue consisted of a cystic fusiform mass attached to a slender-like structure representing a nerve. The whole specimen measured $4 \times 0.3 \times 0.2$ cm, was serially sectioned and submitted “en toto.”

Histopathologically, the unremarkable part of the nerve tissue was evident, however the dilated part observed grossly showed many myelinated axons, and collagen fibers loosely distributed within a myxoid stroma. The tissue was surrounded by a thin layer of perineurium and continues as a thickened unremarkable nerve (Fig. 2C and D).

Post operatively, the patient complained of decreased sensation and numbness over the distribution of the frontal nerve and was improving gradually over few months. She had no recurrence 2-years post-surgical treatment and possible excision of the lesion on the left side is being considered.

Discussion

Peripheral nerve tumors are rarely found in the orbit as they account for only 2% of the space occupying lesions. Out of these, 0.7% of the lesions are neurofibromas. Isolated neurofibroma, a solitary subtype, is relatively uncommon to be encountered in the orbit in the absence of systemic features of neurofibromatosis. This lesion has been described with variable configuration in the English-written literature. Rose and Wright reported a 46% incidence of neurofibroma in orbital peripheral nerve sheath tumors. Patients presented with slowly progressive clinical features such as swelling, proptosis, visual changes, ptosis, diplopia or pain. Family history and/or systemic neurofibromatosis was present in 28% of patients. Their report also showed that the tumors had a smooth contour on CT scan with 41% extension to the superior orbital fissure, as seen in our case. In their series, the nerve involved in almost all cases was the frontal nerve. However, disturbance of the nerve function was uncommon.

Braich recently reviewed 18 published articles that included 45 patients with isolated neurofibroma reported in the literature. He noted 4 cases only with bilateral lesions, as seen in our case. In his review, he concluded, almost equal distribution between males and females, a wide age-range from 1.5 to 82 years and median age of 32–38 years. It is a slowly progressive lesion presenting mostly with proptosis in 69% of the cases and with a variable duration of symptoms ranging from 0.2 to 32 years.

These tumors are usually described to be well-circumscribed during surgery. Krohel mentioned detailed pathologic description grossly as gray to white masses with little vascularization. Microscopically, the tissue is characterized by interwoven strands of spindle-shaped cells with elongated processes in a bundle arrangement with a mixture of normal nerve fibers. Out of 9 specimens, only one was found to show encapsulation of the neoplasm.

Where possible, complete resection of the tumor tends to be curative. Although, sensory deficit postoperatively is expected and was evident in 72% of patients in one study, and has been also noted in our patient. No evidence of recurrence of the neoplasm has been mentioned even to 23 years of follow up.

In conclusion, Isolated neurofibroma is a rare tumor to be found in the orbit and bilateral occurrence is even less likely.
to be encountered in patients without systemic neurofibromatosis. It is a slowly progressive lesion that occurs mainly in adults without sex predilection. The commonest presentation is proptosis; however, diplopia and pain can happen. Surgical excision of the neoplasm is curative with no tendency for recurrence, but patients usually suffer from sensory deficit after the procedure.

Declaration statement

This case report was prepared in accordance with the ethical standards of the human ethics in accordance with the Helsinki Declaration. A general informed consent was taken from the patient, which includes permission for anonymous reporting.

Conflict of interest

The authors declared that there is no conflict of interest.

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