Schwannoma: An Unusual Lacrimal Gland Tumor in a Child

To the Editors:

Schwannoma is a benign, slow-growing solitary peripheral nerve sheath tumor that preferentially involves spinal nerve roots and sympathetic, cervical, and vagus nerves. Schwannomas are uncommon in the orbit and usually arise from the supraorbital or supratrochlear nerve, generally occurring in adults.1 We describe an unusual case of calcifying lacrimal gland schwannoma in a child, which is extremely rare.

A 12-year-old girl presented with recurrent, painless, left upper eyelid mass for 7 years. The mass had twice been operated on elsewhere; the exact details were not known but the last surgery was done 1 year prior to presentation. Systemic examination was normal.

On examination, best-corrected visual acuity was 6/6 in both eyes. There was S-shaped deformity of the left upper eyelid with an underlying non-tender, firm, lobulated, and non-compressible mass (Figs. 1A and 1B). The rest of the ocular examination was normal. Computed tomography (Figs. 1C and 1D) showed a well-margined heterogeneously enhancing mass in the superotemporal quadrant of the left orbit. There were dense, nodular foci of calcification within the tumor. Transseptal lateral orbitotomy was done and a discrete mass in association with lacrimal gland was removed completely.

On gross examination, the mass was well encapsulated, measuring 3.9 × 2.8 cm. Cut section of the specimen revealed multilobulated, fish-flesh colored mass with foci of yellow discoloration (Fig. 1E). On microscopic examination, the parent lacrimal gland tissue could be well identified from the tumor. Verocay bodies and multiple foci of calcification were seen pointing toward schwannoma (Fig. 2). On immunohistochemistry, the tumor cells showed positive reactivity for S-100 antigen, confirming the diagnosis of schwannoma.

The postoperative course was uneventful (Fig. 1F). There has been no recurrence at 5 years of follow-up.

In association with the eye, schwannomas mostly arise within the orbit and rarely involve structures such as uveal tissue, conjunctiva, or eyelids.1,2 Schwannoma arising within the lacrimal gland is rare.3,4 The tumor in our case was seen in association with the lacrimal gland, but we could not directly identify the lacrimal nerve. The presenting age of the child was 12 years,
with the first appearance of the mass at the age of 5 years. This is uncommon; the usual presenting age is 20 to 50 years. Cystic and fatty degenerations are commonly known to occur within schwannomas, but calcification as seen in our case is extremely rare; only two cases, one orbital and one in the lacrimal gland region, have been reported before.\textsuperscript{3,5} Usually, calcification in the lacrimal gland region is associated with adenoid cystic carcinomas, pleomorphic adenomas, and dermoids.\textsuperscript{1}

To the best of our knowledge, calcifying lacrimal gland schwannoma in a child has not been reported previously. Because schwannomas are encapsulated non-invasive tumors, it is important to differentiate these tumors from other masses with a similar presentation to ensure appropriate management and because approximately 10% of cases of schwannomas are associated with multisystem disorders such as neurofibromatosis, schwannomatosis, multiple meningiomas, and Carney complex. However, these associations were not present in our case.\textsuperscript{6}

REFERENCES

Mandeep S. Bajaj, MD\textsuperscript{1}
Mridula Mehta, MS, DNB\textsuperscript{1}
Seema Sen, MD\textsuperscript{2}
Neelam Pushker, MD\textsuperscript{1}
Sumita Sethi, MS\textsuperscript{1}
Supriyo Ghose, MD, MNAMS\textsuperscript{1}
\textsuperscript{1}Oculoplastics & Paediatric Ophthalmology Services
\textsuperscript{2}Ocular Pathology Services
Dr. Rajendra Prasad Centre for Ophthalmic Sciences
All India Institute of Medical Sciences
Ansari Nagar, New Delhi, India

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Letters to the Editors

Transient Binocular Diplopia As a Rare Complication of Local Anesthesia

To the Editors:

We describe two cases with transient binocular diplopia following local anesthesia. The first case was a 13-year-old girl who was given a standard left superior alveolar nerve block with lidocaine hydrochloride 40 mg and adrenalin 0.025 mg for the treatment of superior left premolar 2. Thirty minutes after the local anesthesia, the child complained of double vision that was related to abducens nerve paresis of the left eye, which completely resolved within 45 minutes.

The second case was a 35-year-old man who was given right superior alveolar nerve and right infraorbital nerve block with lidocaine hydrochloride 40 mg and adrenalin 0.025 mg. Forty minutes after the local anesthesia, he complained of double vision that was related to partial oculomotor nerve palsy (Fig. A), which completely resolved within 4 hours (Fig. B). In both cases, the anesthetic agent was given after negative blood aspiration.

Several anatomic mechanisms have been suggested to explain how the anesthetic agent could access structures necessary to produce ophthalmoplegia. The first mechanism is that cavernous sinuses receive blood from the superior-inferior ophthalmic veins, superficial middle cerebral vein, and sphenoparietal sinus. These sinuses communicate anteriorly with the facial vein through the superior and inferior ophthalmic veins. Cavernous sinuses also communicate inferiorly with the pterygoid venous plexus and posteriorly with the basilar plexus, which in turn communicates with the internal vertebral venous plexus. The anesthetic agent from the pterygozygomatic-infratemporal fossa enters the pterygoid venous plexus, where it only causes paralysis of the lateral rectus muscle because of the separated abducens nerve by a thick wall of duramater.\textsuperscript{1}

The second mechanism is that the blood supply of the orbit is mainly from the ophthalmic artery and the infraorbital artery. The superior alveolar artery branches off the maxillary artery in the pterygopalatine fossa. It descends to the maxilla, where it gives off branches that supply the molar and premolar teeth, the maxillary sinus, and the gingiva. Passing of anesthetic agent into a branch of the facial artery carries it to the maxillary artery related to the orbital floor. Anomalous anatomical course of the superior alveolar artery, anastomotic link of the middle meningeal artery with the
The lacrimal branch of the ophthalmic artery and varieties of the maxillary artery should also be considered.

The third mechanism is that the infraorbital foramen is located 8 mm below from the infraorbital margin. Arteria, vena, and nervus infraorbitalis pass from the foramen infraorbitale. Facial angular artery, and facial angular veins are also running near the infraorbital foramen.

The fourth mechanism is reaching through the soft tissue by direct diffusion. Penetrating the orbital fossa via an anatomical defect of the maxillary sinus wall should also be considered.

Maxillofacial regional local anesthesia procedures are generally used in dental practice and maxillofacial surgery. Most of the extraocular nerve palsies in the literature are related to the mandibular block anesthesia. We present extraocular nerve palsies after the same regional local anesthesia for two different types of treatment procedures without any relation to mandibular block.

As soon as this complication appears, a detailed ophthalmological examination should be performed and the patient reassured by explaining the transient nature of the double vision.

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Osman Melih Ceylan, MD
Maresal Cakmak Military Hospital
Erzurum, Turkey
Fatih Mehmet Mutlu, MD
Halil Ibrahim Altinsoy, MD
Gülhane Military Medicine Academy
and Medical School
Ankara, Turkey

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Delayed Presentation of Children With Waardenburg Syndrome

To the Editors:

Waardenburg syndrome (WS), named after the Dutch ophthalmologist Petrus Johannes Waardenburg, is a rare hereditary disorder with ocular and systemic involvements. Based on genotypic and phenotypic variations, four different types of WS have been described, of which types 1 and 2 are the most common. According to the diagnostic criteria proposed by the WS Consortium, the major criteria are congenital sensorineural hearing loss (SNHL), pigmentary disturbances of the iris and hair, affected first-degree relative, dystopia canthorum, and a W index that exceeds 1.95 [The W index (significant if more than 1.95) is calculated using the following formula: X = (2a-0.2119c-3.909)/c Y = (2a-0.2479b-3.909)/b W = X + Y + a/b, where (a) inner canthal distance, (b) the interpupillary distance, and (c) the outer canthal distance, in mm.] The minor criteria are congenital vitiligo, synophrys, broad high nasal root, hypoplasia of alae nasi, and premature graying of hair. Individuals are considered to have WS type 1 if they fulfill two major (including dystopia canthorum) or one major and two minor criteria from the list. Individuals having other features but not dystopia canthorum are considered to have WS type 2. WS type 3 (or Klein–Waardenburg syndrome) and WS type 4 (or Waardenburg–Shah syndrome) are similar to WS type 1 but include skeletal muscle contractures and Hirschsprung’s disease, respectively.

We report a case series of six children (2 to 6 years old)
who presented with characteristic manifestations of WS, with an emphasis on late presentation and hence improper development of speech and language skills. The most common presenting complaint was inability to speak and hear, followed by difference in eye color.

Systemic examination revealed presence of hypopigmented patches on skin (cases 1, 2, and 4) and hypopigmented hair (cases 1 and 2). Ocular examination revealed heterochromia iridum in four cases and heterochromia iridis in two cases (Figs. 1A, 1B, and 1D). Pure tone audiometry revealed varying severity of SNHL in all six children. Magnetic resonance imaging three-dimensional reconstruction of inner ear (in one child with severe SNHL) revealed bilateral hypoplasia of cochlear cells (Fig. 1C). Considering the parental concerns, a trial of hearing aid was given and the parents were counseled for cochlear implant.

In WS, the auditory defects and pigmentary abnormalities are explained by improper differentiation of the melanocytes, secondary to mutations of various genes (PAX3, MITF, EDNRB, and SOX10).² Tissues derived from the neural crest and involved in the pathogenesis of WS are the cochlear striae vascularis, frontal bone, limb muscles, and enteric ganglia.

In newborns and infants, WS may be detected by pigmentary abnormalities and by hearing screening. There are two specific modalities for newborn hearing screening (ie, the transient otoacoustic emission or brain stem electric response audiometry).⁴ Treatment consists of auditory rehabilitation in the form of hearing aids and cochlear implant, where possible.⁵

In the current case series, the difference in eye color had been noticed by the parents since birth, but they did not consider it serious enough and presented later with SNHL. It is important to identify such children early in life to ensure complete evaluation and comprehensive management in the form of amplification, appropriate educational intervention, and cochlear implant where possible to assist adequate development of speech and language skills in the affected children.

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Mridula Mehta, MS, DNB
Sumita Sethi, MS
Neelam Pushker, MD
Mandeep S. Bajaj, MD
Supriyo Ghose, MD, MNAMS
Pediatric Ophthalmology Services
Dr. Rajendra Prasad Centre for Ophthalmic Sciences
All India Institute of Medical Sciences
New Delhi, India
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