OPEN

PRS GLOBAL OPEN

VIEWPOINT
Reconstructive

Orbital/Periorbital Plexiform Neurofibromas: Classification and Surgical Strategies for a Better Outcome

Akiko Yoshinaga, MD; Itaru Tsuge, MD, PhD; Susumu Saito, MD, PhD; Naoki Morimoto, MD, PhD

Neurofibromatosis type 1 (NF1) is an autosomal dominant disease occurring in approximately one in 3500 births. Plexiform neurofibroma (PN) involving the eyelid, orbit, periorbital, and facial structures has been labeled as orbital-periorbital PN (OPPN).[1] OPPN affects less than 10% of NF1 patients, and most of them track along the distribution of the trigeminal nerve.[1] OPPN can invade and destroy surrounding tissues, causing proptosis, ptosis, amblyopia, and facial disfigurement leading to visual dysfunction and social distress. Surgical management of OPPN is challenging for not only functional and cosmetic reasons but also because of the risks of complications, including bleeding and brow ptosis, caused by damage to the temporal branch of the facial nerve.[1–3] All OPPN cases reportedly developed postoperative damage of the temporal branch of the facial nerve when conventional debulking was performed in the temporal lesion.[3] Unfortunately, there is no consensus yet concerning a surgical solution for OPPN.[1] The present study explored the surgical outcomes in terms of the preoperative tumor location.

Medical records of NF1 patients with a surgical history of OPPN who visited our clinic were retrospectively reviewed (April 1, 2005, to January 31, 2021). Patients were assessed based on axial-section magnetic resonance imaging or computed tomography findings and categorized into two groups by preoperative tumor location: anterior OPPN (OPPN-A) for tumors that had not penetrated the retrobulbar area and posterior OPPN (OPPN-AP) for tumors that extended beyond the retrobulbar area (Fig. 1). The visual function was evaluated preoperatively by an ophthalmologist. The pre- and postoperative margin reflex distance 1 (MRD1) were measured, and postoperative photographs of all the patients were evaluated based on the Whitaker classification[4] in a blinded fashion by two plastic reconstructive surgeons. This study was approved by the ethics committee of Kyoto University Graduate School and Faculty of Medicine, Kyoto, Japan (ethical approval number: R2902).

Five patients were enrolled (Table 1). The three patients with OPPN-A showed good results with regard to both the improvement of MRD1 and aesthetic outcome, while these results were poor in the two patients with OPPN-AP. Two of the three patients with OPPN-A had sustained damage to the temporal branch of the facial nerve during surgery at their previous hospital. We performed brow lift for the brow ptosis and obtained a good outcome. In OPPN-AP, conservative surgery did not markedly improve the MRD1 or aesthetic outcome because of severe infiltration into surrounding critical tissues, such as the levator, nerves, and orbital bone. Orbital exenteration and reconstruction need to be considered in blind OPPN-AP patients.[2]

We suggest that treatment strategies for OPPN-A and OPPN-AP should be managed separately, as the surgical outcomes are markedly different. Although there is a surgical classification based on orbital soft tissue, bony involvement, and blindness, there is no further consensus concerning surgical management for OPPN.[2] Similar to periocular skin tumor,[5] classifying OPPN based on its depth as OPPN-A or OPPN-AP is useful for deciding on surgical strategies.

Akiko Yoshinaga, MD
Department of Plastic and Reconstructive Surgery
Graduate School of Medicine, Kyoto University, Kyoto, Japan
Kawaharacho 54, Shogoin, Sakyo-ku,
Kyoto, 606-8507, Japan
E-mail: akikoyoshinaga0530@gmail.com

DISCLOSURE

The authors have no financial interest to declare in relation to the content of this article.

REFERENCES

1. Avery RA, Katowitz JA, Fisher MJ, et al;OPPN Working Group. Orbital/peri-orbital plexiform neurofibromas in children with neurofibromatosis type 1: multi-disciplinary recommendations for care. *Ophthalmology*. 2017;124:123–132.
2. Erb MH, Uzcategui N, See RF, et al. Orbitotemporal neurofibromatosis: classification and treatment. *Orbit*. 2007;26:223–228.
3. Hu X, Bogari M, Tan A, et al. Initial exploration on temporal branch of facial nerve function preservation in plexiform neurofibroma resection. *J Craniofac Surg*. 2016;27:1589–1592.
4. Denadai R, Buzzo CL, Takata JP, et al. Comprehensive and global approach of soft-tissue deformities in craniofacial neurofibromatosis type 1. *Ann Plast Surg*. 2016;77:190–194.
5. Martel A, Lassalle S, Picard-Gauci A, et al. New targeted therapies and immunotherapies for locally advanced periocular malignant tumours: towards a new "eye-sparing" paradigm? *Cancers*. 2021;13:2822.

From the Department of Plastic and Reconstructive Surgery, Graduate School of Medicine, Kyoto University, Kyoto, Japan.

Received for publication April 13, 2023; accepted May 16, 2023.

Presented at the 11th Annual Meeting of The Japanese Society of Recklinghausen Disease in Tokyo, Japan.

Plast Reconstr Surg Glob Open 2023; 11:e5108; doi: 10.1097/GOX.0000000000005108; Published online 6 July 2023.

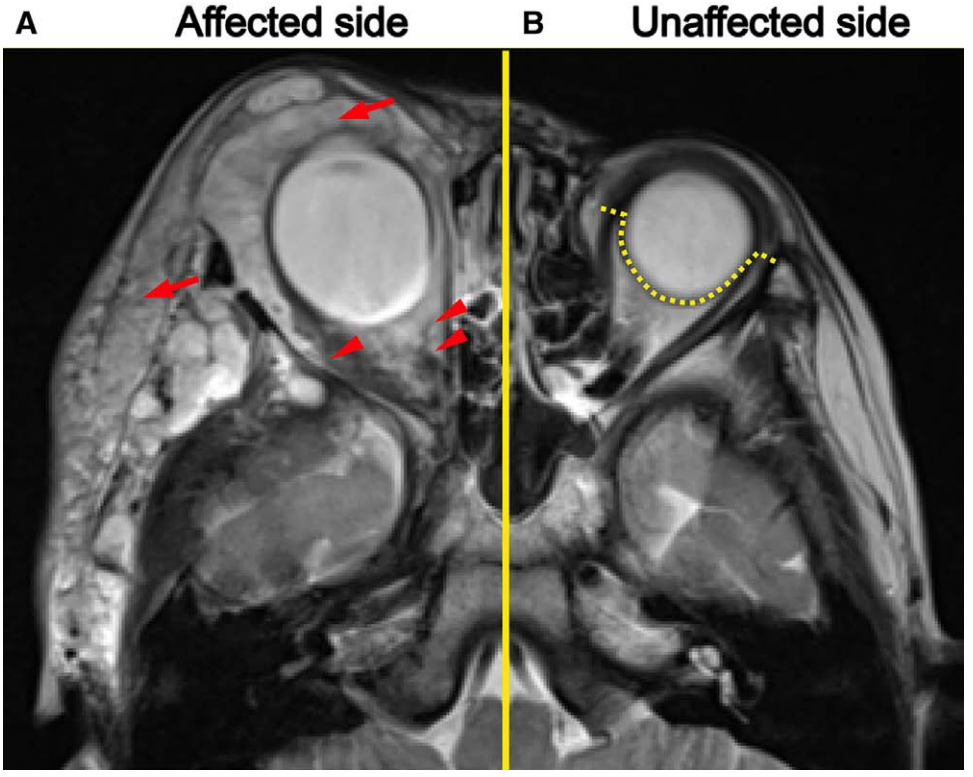

Fig. 1. Proposed classification for OPPN based on tumor localization using magnetic resonance imaging. T2-weighted MRI from the patient #4 with OPPN-AP. A, The tumor compressed extraocular muscles and invaded to surroundings of optic canal (arrowheads). B, The dotted line, defined by the posterior aspect of the eye ball, indicates the border line between OPPN-A and OPPN-AP. Arrows and arrowheads indicate OPPN-A and OPPN-AP, respectively.

Table 1. Summary of the Backgrounds and Surgical Outcomes

ID	AAFE (y)	Sex	Anterior/ Anterior + Posterior	Bone, Y/N	Visual Function	No. Surgeries	Preoperative/ Postoperative MRD1 (mm)	Aesthetic Outcome (Whitaker Score)	Postoperative Complications
1	56	M	Anterior	N	ND	11	−5/1	I	Brow ptosis
2	61	F	Anterior	N	ND	13	0.5/2	I	Brow ptosis
3	49	M	Anterior	N	ND	3	ND/1.5	I	None
4	22	F	Anterior + posterior	Y	Blind	1	−5/−5	IV	None
5	21	F	Anterior + posterior	Y	Amblyopia	3	−0.5/−5	IV	Ptosis

"Bone" indicates bony involvement.
The Whitaker classification comprises four categories: category I, no refinements or surgical revisions advisable or necessary; category II, soft-tissue or lesser bone contouring revision was advisable; category III, major osteotomies or bone graft repositioning advisable (not as extensive as the original procedure); category IV, major craniofacial procedure advisable (duplicating or exceeding the original procedure).[4] "Brow ptosis" refers to the descent of the eyebrow caused by damage to the temporal branch of the facial nerve, and "ptosis" refers to the drooping of the upper eyelid caused by damage to the levator.
AAFE, age at final evaluation; N, no; ND, evaluations were not done; Y, yes.