CO₂-Laser in the Removal of a Plexiform Neurofibroma from the Eyelid

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Abstract. We present a child with neurofibromatosis type I (NF-I) who underwent excision of a large upper lid neurofibroma using a CO₂-laser. Plexiform neurofibromas are notorious for their bleeding tendency and the inability to achieve complete surgical excision. The CO₂-laser is an adjunct to achieving better hemostasis and delineation of the tissue in the absence of natural tissue planes caused by the tumor's diffuse mode of growth. Using the CO₂-laser achieves better cosmesis, while reducing operation time and limiting complications. [Ophthalmic Surg Lasers 2000;31:432-434]

INTRODUCTION
Neurofibromatosis I, van Recklinghausen disease, is an autosomal dominant disease with incomplete penetrance and variable expression, found in about 1:3000 live births. It represents 95% of cases of neurofibromatosis. Diagnostic criteria include: 1) Six or more cafe-au-lait spots, >5 mm in prepubertal persons, and >15 mm in postpubertal persons; 2) Two or more neurofibromas of any type, or one plexiform neurofibroma; 3) Freckling of the axillary or inguinal regions; 4) Optic glioma; 5) Two or more Lisch nodules; 6) Osseous lesions such as sphenoid dysplasia and thinning of long bone (with or without pseudoarthrosis); 7) First-degree relative (parent, sibling, offspring) with neurofibromatosis I. A definitive diagnosis needs two or more criteria. Orbitotemporal involvement is frequent and may lead to blindness from involvement of the optic nerve, from glaucoma, or deprivation amblyopia from advanced ptosis. Isolated orbital neurofibromatosis has been described, but is rare.

CASE REPORT
A 10-year-old boy with neurofibromatosis I has been followed by our department since his birth. He has a healthy twin sister (Figure 1). The boy has severe neurofibromatosis with multiple cafe-au-lait spots, freckled skin in the axillae and inguinala, orbital involvement with NF-I, congenital glaucoma, mental retardation, gingival hypertrophy, spheroid dysplasia, and osseous deformation of the long bones of the legs, including a pseudoarthrosis of one of his tibia. The mother is the carrier, having only cafe-au-lait spots. The other 4 siblings are healthy. The deceased maternal grandfather is presumed to have been a carrier based on the reports of his daughter. No other family members are affected. Several of the mother's siblings died in childhood from causes unknown to us.

At the age of one week, the patient underwent filtration surgery in the right eye for congenital glauco-
ma. At age 10 months he had wandering eye movements in both eyes with a sluggish pupil in the left eye. At the age of 15 months the patient was observed to have OD occlusio pupillae. The plastic and reconstructive surgery department followed the patient for a cleft soft palate that had not been surgically corrected. At the age of 4 years, the right eye was found to be phthitic and the left eye was diagnosed with high myopia. Deformation of the upper eyelids of both eyes was present and advanced with each follow-up visit. Following, examining, and treating the child was extremely difficult because of his severe psychomotor retardation.

On computed tomography of the brain and orbits, a right optic nerve glioma, a right spheroid dysplasia with cranial content bulging into the right orbit, and an enlarged middle cranial fossae with enlarged cerebral ventricles, were diagnosed (Figure 2). The right upper lid was grossly deformed with elephantiasis neuromatosa of the lid. It was hanging a full 1 cm over the lower lid without closure over the phthitic eye that had copious mucous discharge unresponsive to conservative therapy (Figure 3). We considered performing neurofibroma excision with ptosis repair to enhance the patient's comesis and make him socially more acceptable. At the age of 10½ years, under general anesthesia, the patient underwent excision of the upper eye lid plexiform neurofibroma, using a CO$_2$- laser. Laser settings were continuous mode, at 7 watts (Laser Industries, model 30C). The incision was placed 4 mm above the superior eyelid margin to create the absent superior eyelid crease. The neurofibroma was excised, together with some of the overlying redundant skin. Dissection and debulking was done with the CO$_2$-laser to improve hemostasis. Blood vessels larger than 1 mm were coagulated with bipolar cantery. Still more profuse bleeding was observed than is usual with CO$_2$-laser-assisted blepharoptosis repairs. Good alignment of the lid margin was achieved by suturing the remaining eyelid tissue with a good cosmetic outcome. A week after surgery the patient was re-admitted because he had torn the lateral flap of the upper eyelid. The wound was re-sutured, and further follow-up was uneventful (Figure 4).

**DISCUSSION**

In this case, we excised a giant plexiform neurofibroma from the upper eyelid with the help of a CO$_2$-laser. The use of CO$_2$-laser in excision of neurofibromata is not new, though it is usually recommended for the removal of multiple small cutaneous lesions.$^{3,4}$ The advantages of using the CO$_2$-laser in this particular case is that the laser decreases the intraoperative hemorrhage, decreases operative time,$^5$ and enables the formation of new tissue dissection planes, which in this case had been utterly destroyed by the tumor. According to Jackson's classification of orbitotemporal neurofibromatosis, our patient suffers with type 3, the
most severe form including eyelid deformation, osseous defect in the sphenoid bone, intraorbital extension of the tumor, and an underlying blind eye (Table). He recommends that in such a case an extended exenteration be performed with reconstruction using bone grafts and a composite eye-skin prosthesis.6

Our patient, being severely mentally retarded, was clearly not a candidate for such extensive surgery and rehabilitation. Therefore, we opted for a limited excision with the aim of improving the cosmetic outcome. The parents were informed that the growth would most likely recur. In this case, surgery improved cosmetic appearance and alleviated the constant discharge from the phthitic eye. We believe that the CO₂-laser is an important tool in selected cases in which radical excision and major reconstruction are not an option.

REFERENCES