Elephantiasis neuromatosa is a severe form of neurofibromatosis type 1, in which there is diffuse proliferation of Schwann cells and axons together with a bony hypertrophy, resulting in massive disfigurement. We herein report a case of this kind associated with a severely ptotic ear.

**CASE PRESENTATION**

A 40-year-old woman of a low socioeconomic status presented to our clinic from a remote province. She had a history of unilateral facial tumor and disfigurement, which started in her early childhood. There were no other health problems. Her family history was negative for such conditions.

When the patient took off her veil, an extremely disfigured face was noted. The left side exhibited lower facial and neck masses with an extremely hypertrophied and sagging skin (Fig. 1). Two main masses were noted: 1 over the mandibular border, causing an excessive gravitational pull on the cheek tissues along with eyelid ectropion, and a second, bigger bulge involving the neck region on the same side. The whole external ear was found to be hanging down, with the surrounding hypertrophied, saggy skin reaching to the level of the clavicle. Palpation of the masses revealed the classic “bag of worms” consistency. The patient had several café au lait spots and axillary freckles.

A diagnosis of neurofibromatosis type 1 was made, with a large plexiform neurofibroma evident clinically and on MRI. The tumor displayed significantly enlarged and rich vascularity via the external carotid system.

The patient’s main complaint, aside from the tumor, was the migration of her left ear to the top of her chest. She indicated that her hearing was very weak on the affected side, and she tended to manually manipulate her ear whenever she needed to hear better. A baseline audiogram confirmed a severe ipsilateral conductive hearing loss. The patient was well aware of the likely surgical sequelae: facial nerve dysfunction, hemorrhage, ear necrosis, and tumor regrowth.

The primary aim of the first and major surgical intervention was to lift the extremely descended ear. The chosen solution was a single large Z-plasty, aiming to transpose a flap containing the external ear, with its underlying extremely stretched auditory canal, to a proper location. In the second stage, further tumor debulking was performed. In the final stage, the floppy ear was set back significantly by obliteration of the auriculocephalic sulcus and was suspended by suturing to the mastoid fascia. The patient showed a remarkable improvement. A significant part of her neurofibroma was debulked, and the ear was salvaged, reduced in size, and transferred to a near-normal location. This was quite a unique and challenging case in terms of extreme external ear ptosis and malposition; however, success was achieved through adherence to basic principles and techniques, staging, collaborative support, and full commitment. (Plast Reconstr Surg Glob Open 2020;8:e3164; doi: 10.1097/GOX.0000000000003164; Published online 27 October 2020.)
angle of approximately 60-degree angle was used. To avoid compromising perfusion to the flaps, we executed this step of the operation first rather than focusing on debulking the tumor in the adjacent region. The ear fully survived but remained edematous for a few weeks. The pathology report confirmed the diagnosis of neurofibroma.

The second stage, performed about 2 months later, involved further tumor debulking, as well as ear reduction via a geometrical subunit excision, which essentially consisted of the classic combination of wedge and crescent excisions described by Tanzer in 1977. In addition, an attempt was made to stabilize the ear using concha-mastoid sutures. Overall, the patient’s ear had a much better location after the operation, but it continued to be quite floppy with a tendency to fall anteriorly, and edema recurred (Fig. 3).

The last stage was performed 6 months after the first, by which time the edema had settled significantly. It included a further reduction of earlobe tissue by a direct excision, in addition to a significant setback of the ear. This was achieved by an elliptical excision of tissues from the postauricular area (flap debulking), with the aim of obliterate the auriculocephalic sulcus. The ear cartilages were secured to the mastoid periosteum using 3-0 nylon sutures (Ethicon, Somerville, N.J.). Wounds healed well, and the ear was restored to a near-normal location and reasonable shape.

As anticipated, bleeding was significant, and the patient lost from 500 to 700 mL of blood during each of the first 2 operations. On both occasions, she received a few units of blood and was managed in the stepdown unit for the first couple of days after surgery.

**DISCUSSION**

Milder cases of external ear deformities associated with neurofibromatosis type 1 have been reported; these cases were caused by a direct mass effect on the ear and treated by excision of the neurofibroma and suspension. In our case, it was extreme gravitational forces and tissue expansion that were responsible for the descent of the ear to the level of the clavicle.

As a result of Z-plasty, alopecia was anticipated at the site of the cervical flap in the temporal area, which was a negligible issue for this patient. A smaller challenge after the 2 major stages was the tendency of the transferred ear tissue to fall anteriorly, making the ear protrude severely. There was no means of addressing this, other than making the ear adhere better by removing more tissues postauricularly. Recurrent lymphedema of the ear (pitting) responded gradually to massaging.

Surgically, neurofibromas behave very much like vascular malformations in terms of bleeding tendency. Hypotensive anesthesia, blunt cannula infiltration with dilute epinephrine, heavy surgical ties, and thrombin sealants are some of the essential measures to be used. Preoperative angiographic embolization should
be considered in thorough consultation with interventional radiologists. Tumor regrowth and recurrence are expected sequelae, requiring repeated surgical debulking and adjustments. Patients’ priorities need to be respected, as surgery in these patients is to some extent palliative.

Our patient was extremely grateful for the aesthetic outcome, and her hearing on the affected side returned close to normal. Our experience shows that by adhering to the basic historic principles of reconstruction, conducting the operation in several stages, and executing the intervention with full commitment, it is possible to succeed in achieving both the surgeon’s goals and the patient’s desires, even in such challenging cases.

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