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Neurofibromatosis and brainstem implants: What to do?

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Abstract. – **The histopathological changes in the temporal bones of one donor individual deceased for complications of neurofibromatosis type 2 are studied. The different modalities of presentation of neurofibromatosis and the criteria for the differential diagnosis are presented. The possibilities of the auditory brainstem implants in this patology are discussed.**

Key Words:

Neurofibromatosis, Brainstem implants.

Introduction

Neurofibromatosis and bilateral acoustic neuromas should be carefully separated. Neurofibromatosis is a condition characterized by multiple cutaneous neurofibromas and cafè-au-lait spots. This is inherited as an autosomal dominant in a little more than 50% of cases. Meningiomas, astrocytomas and other neoplasms may be associated with it, but acoustic neuromas are rare. Bilateral acoustic neuroma is not associated with cutaneous neurofibromas and cafè-au-lait spots, but this location of the neural tumour and its bilaterality are inherited as an autosomal dominant. Such a family, which was first described in 1930[1], was more recently found to have almost 100 members with definite or possible bilateral acoustic neuroma[2].

Pathological appearances

Neurofibromatosis is a disease transmitted genetically, altering nerve cell growth.

Two types of neurofibromatosis can be distinguished. One has prevalent tropism for the peripheral system (NF-1), the other for the central system (NF-2).

The different modalities of presentation and the criteria for the differential diagnosis are expressed in Table I

Subjects with NF-2 have a greater risk of developing other tumors, such as schwannomas, meningiomas, gliomas, ependymomas, and plexiform neurofibromas.

Patients with NF-2 show an alteration of the acoustic nerve and thus no connection between the ganglion of Corti and the cochlear nuclei. A stimulus carried with a cochlear implant to the scala tympani would not in that case be transmitted to the Central Nervous System, due to the alteration in the acoustic nerve.

Materials and Methods

We have studied the histopatological changes of one case of neurofibromatosis. This patient was a donor and agreed during his life to donate post mortem his temporal bones to the House Ear Institute as a contribution to a better knowledge of temporal bone diseases.

We have removed the temporal bones in our usual way[3-5].

Results and Discussion

At post-mortem neural neoplasms are frequent in both eighth nerves and other central nerves. There are often many small schwannomas and neurofibromas growing on cranial nerves. Meningiomas, usually multiple, are also present in this condition. As well as the major acoustic tumours,

Table I. Different modalities of presentation and criteria for the differential diagnosis between NF-1 and NF-2 .

Definition
NF-1
Also called von Recklinghausen's disease or peripheral neurofibromatosis
NF-2
Also called bilateral hereditary syndrome of bilateral schwannomas.
Genetic
NF-1
Dominant autosomic disease. Transmitted to 50% of offspring. Gene located at band 17q on the long arm of chromosome 17, near the centromere
NF-2
Dominant autosomic disease. Transmitted to 50% of offspring. Gene is located in the middle of chromosome 22's long arm.
Incidence
NF-1
1/4000
NF-2
1/40000
Age
NF-1
The skin spots appear mostly on the trunk within the first 4 years of life.
NF-2
Signs may manifest from 10 to 60 years, but in general later than those of NF-1.
Diagnosis
NF-1
At least 2 of the following signs:
• More than 5 coffee colored spots larger than 5 mm in children and 15 mm in adolescents and adults.
• Two or more neurofibromas or one plexiform neurofibroma.
• Freckles on the armpit or groin.
• Glioma of the optic nerve.
• Two or more raised melanocytic hamartomas, yellow or darker, on the surface of the iris.
• Specific osseous lesion, such as a displasia of the sphenoid or a thinning of the cortex of the long bones, with or without pseudoarthritis.
• Close relative with NF-1 defined with the aforementioned criteria
NF-2
Subject has bilateral schwannomas of the vestibular nerve or a close relative with NF-2, or at least 2 of the following symptoms:
• Cutaneous or subcutaneous neurofibromas
• Plexiform neurofibromas.
• Schwannomas.
• Gliomas.
• Posterior juvenile subcapsular cataract.

meningiomas and neurofibromas of microscopic size may be present, the former on the meninges in the vicinity of the acoustic neuromas and sometimes even intermixed with them microscopically. In neurofibromatosis, neuromas are histologically identical to those of the single acoustic tumours, but are more invasive in their behaviour, tending to involve the cochlea and vestibule[6]. The case described by us is of this type (Figure 1). The aggressive and infiltrating behaviour of neurofibromatosis makes very difficult the removal of acoustic neuroma tumor with hearing preservation. In these cases we face the problem of giving the sensation of sound to an otherwise deaf patient. This possibility is given by the auditory brainstem implant which permit bypassing both the cochlea and the cochlear nerve to directly stimulate the cochlear nuclei.

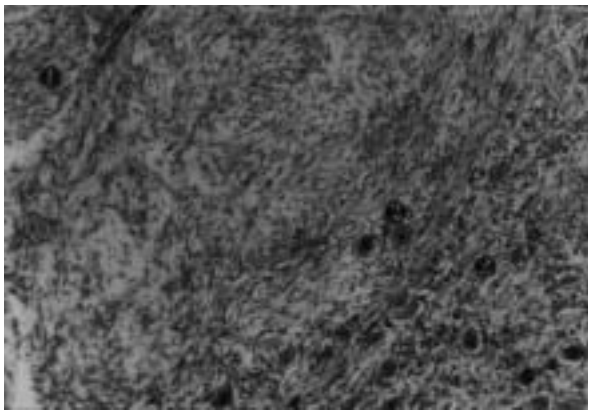

Figure 1. Neurofibromatosis II. The tumor is pleomorphic containing, remnants of schwannoma (1), ganglion cells (2), and meningioma (3). *×120*

Currently, patients who can receive an auditory brainstem implant are patients with bilateral acoustic neuromas and type 2 neurofibromatosis, according to a protocol monitored by the US Food and Drug Administration (Tables 2, 3).

Anatomy

The structures in anatomic relationship with the lateral process of the fourth ventricle are the following: the foramen of Luschka, the choroid plexus, and the floccule.

- The extremity of the lateral recess forms the foramen of Luschka, which is found at the edge of the bulbopontine sulcus. Just superior to the foramen is the root of nerves VII and VIII and inferior is the root of nerves IX and X.
- The choroid plexus is attached to the inner surface of the choroid tela and protrudes from the foramen of Luschka below the zone of entrance of nerves VII and VIII and slides to overlap the posterior part of nerves IX and X.

Table II. FDA Criteria for implantation .

- Presence of bilateral tumors of nerves VII/VIII involving the internal auditory canal or the cerebellopontine angle
- Familiarity with the English language.
- Age at least 15
- Good psychological state
- Willingness to take part in research studies.
- Realistic expectations

Table III. FDA criteria for the auditory brainstem implant in NF-2 patients.

- Secondary tumor in the only hearing ear
- Any tumor larger than 1.5 cm in the hearing ear
- Limited life expectancy due to other tumors, medical problems, or advanced age
- Usable hearing with a tumor that does not demonstrate significant growth in MRI checks and stable hearing demonstrated in audiometric tests
- Situations in which a tumor is observed in an ear with residual hearing of an NF-2 patient with bilateral neuromas, if the tumor does not endanger life nor significantly alter vital functions

- The floccule is a cerebellar lobe which protrudes from the margin of the lateral recess and adheres to the rostral margin of the lateral recess and to the foramen of Luschka. The tenia passes across the roof of the lateral recess and outlines the area of the ventral cochlear nucleus.

The stimulation should be carried to the ventral cochlear nerve, which is the most representative nucleus for the transmission of stimuli to the central auditory paths.

The further the electrode is inserted into the recess, the better is the result and the fewer are the side effects.

Implantation technique

The translabyrinthine approach is the most common approach used at the House Ear Clinic for VIIIth nerve neuroma removal, but it is also the approach that gives the best surgical visibility for the positioning of a brainstem implant[7-11]. The electrode is inserted completely into the foramen of Luschka, to give the best yield. After insertion, the implant's function is evaluated in relation to its position and all of the electrodes are stimulated. The evoked potentials are sought, as well as any stimulation of nerves V, VII, and IX, and alterations in the vital signs[12-14].

The postoperative treatment is thus similar to that of translabyrinthine surgery.

The second day after surgery, patient's conditions permitting, a light pulsating signal may be administered, with progressively increasing intensity. The best results and the fewest side effects have been obtained with a 300Hz sinusoid[15-17].

Conclusion

Among 46 NF-2 patients implanted between 1979 and 1999, 28 have obtained auditory benefit, with no significant side effects, in our review of the House Ear Clinic.

Patients who have received an auditory brainstem implant report hearing a sound similar to that of a badly tuned radio station. Sounds are heard but not understood, but all users are aware of environmental sounds and show improved lip-reading scores.

It is important to follow very carefully NF2 patients with MRI, avoiding the tumor removal when otoneurological symptoms are absent. Should these symptoms present, the greatest lesion must be operated first. ABI rehabilitation program must be started *before* the tumor removal on the contralateral side and the implant operation.

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