Short communication

Use of botulinum toxin in a patient with pendular congenital nystagmus

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Abstract

Case report: We report the case of a 5 month-old male diagnosed with congenital nystagmus and oculocutaneous albinism. The initial examination showed pendular horizontal nystagmus with high amplitude and without blocking position or foveal fixation periods. A 2.5 IU injection of botulinum toxin was administered in the horizontal rectus muscles of both eyes in two sessions separated by 6 weeks. This led to a decrease in amplitude of nystagmus and early development of binocular visual acuity of 4.8 cycles/cm.

Conclusion: Faced with diagnosis of horizontal nystagmus in the early stages of development, and in order to avoid periods of foveal fixation, the use of botulinum toxin leads to a temporary reduction in its amplitude and an improvement in visual acuity with low complication rates. Given the possibility of spontaneous improvement described in these patients, studies are needed with longer follow-up to establish the advantage of long-term treatment.

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Empleo de toxina botulínica en paciente con nistagmus congénito de tipo pendular

Resumen

Caso clínico: Se presenta el caso de un varón de 5 meses de edad, diagnosticado de nistagmus congénito y albinismo oculocutáneo. En la exploración inicial se evidenció un nistagmus horizontal pendular de gran amplitud sin posición de bloqueo ni períodos de fijación foveal. Se realizó una inyección de 2,5 UI de toxina botulínica en los músculos rectos horizontales de ambos ojos en dos sesiones separadas por 6 semanas permitiendo una disminución de la amplitud del nistagmus y un desarrollo inicial de la agudeza visual binocular de 4,8 ciclos/cm.

Conclusión: Ante el diagnóstico de nistagmus horizontal en etapas precoces del desarrollo, de amplitud tal que impida períodos de fijación foveal, el empleo de toxina botulínica permite

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Palabras clave:
Síndrome de nistagmus infantil
Nistagmus pendular
Albinismo oculocutáneo
Toxina botulínica

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una disminución temporal en la amplitud del mismo y una mejora de la agudeza visual con baja tasa de complicaciones. Ante la posibilidad descrita de mejoría espontánea de estos pacientes, especialmente durante el primer año de vida, son necesarios estudios con mayor tiempo de seguimiento para establecer la ventaja de este tratamiento a largo plazo.

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Introduction

Infantile nystagmus syndrome (INS) is a motor ocular disorder that presents at birth or in early childhood and is clinically characterized by involuntary eye oscillations. Incidence is estimated in one case for every 6550 or 0.015%. 1,2 Said movements usually have a fast and a slow stage, although they can also be purely pendular. They are generally horizontal with a small torsional component. Additional clinical characteristics include increased intensity of nystagmus with fixation and reduction of intensity with sleep or lack of attention, intensity variations in different eye positions, abnormal positions of the head or association with strabismus. INS can be associated with acquired or congenital visual sensory system defects. Pendular, sudden or spring-like forms of nystagmus without foveal fixation periods are associated to poor visual development, whereas the forms with long foveal fixation periods indicate good vision. Accordingly, the visual acuity of patients with INS depends on the existence or absence of foveal fixation periods.

Nystagmus treatment includes botulinum toxin injections 3-5 or broad resections of horizontal rectus muscles. 6,7 Botulinum toxin has been used for treating various types of nystagmus for 30 years and it must be considered as a reasonable and preferable option to surgery in many cases. Repeated botulinum toxin injections can be made regularly in the rectus horizontal muscles 6,9 or in the retrobulbar space in selected cases. 10 The main complications in the treatment of nystagmus with botulinum toxin are ptosis, lack of ocular alignment control and exceptionally retrobulbar hemorrhage or ocular perforation. Treating congenital nystagmus with botulinum toxin causes a reduction of the nystagmus width, a partial increase of visual acuity and improvements in the abnormal position of the head. 11

The objective of the clinical case presented below is to illustrate the possibility of a significant magnitude of visual development at an early stage of development in a patient with wide nystagmus. The use of botulinum toxin provided excellent results which are probably in direct relationship with the absence of noticeable anatomic involvement.

Discussion

The visual acuity of patients with INS depends on the one hand on the anatomic alterations and on the other on the variable functional disorders associated to the width of the movement. This patient exhibited broad pendular nystagmus without locking position and without any foveal fixation period. Accordingly, adequate visual development was lacking. In the absence of foveal fixation periods, such as with our patient, the use of botulinum toxin applied in the rectus horizontal muscles during the first days of life enabled a significant reduction in the width of the nystagmus and in this case a remarkable improvement of visual acuity with low complication rate.

It is difficult to establish the efficacy of this technique with a view to its generalization as the literature describes a spontaneous improvement in the first year of life of these patients. In addition, it is not clear to what extent this spontaneous improvement of patients is the result of the degree of anatomic involvement, which is a factor that cannot be modified with any therapeutic intervention, or the amplitude of the movement. Accordingly, it is essential to carry out studies with longer follow-up periods to determine the efficiency of this treatment in the long-term and to assess whether the patients that improve with botulinum toxin would also improve spontaneously due to having less anatomic alterations.

Clinical case

Male patient, 5 months old, diagnosed with congenital nystagmus. Relevant personal history includes the existence of oculocutaneous albinism. At diagnostic time it was not possible to record preferential vision due to wide pendular horizontal nystagmus preventing foveal fixation periods. The patient did not exhibit nystagmus obstruction in any gaze position or in convergence. Biomicroscopic exploration did not reveal relevant findings except for slight iris hypopigmentation with a slight degree of translucence. The ocular fundus exhibited moderate hypopigmentation with a physiological appearance of the optic nerve head. The patient did not exhibit torticollis at diagnostic time and the found refraction defect was not significant.

After finalizing the diagnostic and the various supplementary assessments, an injection of 2.5 UI of botulinum toxin was injected in the horizontal rectus muscles of both eyes, enabling a significant reduction in the width of the nystagmus assessed with a video recording and visual acuity measured with the preferential vision test of 2.3 cycles/cm. Six weeks after the first injection, the width of the nystagmus began to increase. Therefore a second botulinum toxin was injected having the same characteristics as the first one. This enabled a visual acuity in the right eye of 2.3 cycles/cm and of 4.8 cycles/cm in the left eye, with the binocular visual acuity being of 4.8 cycles/cm. At this point it was decided to perform an hourly occlusion of the left eye and a close follow-up of the patient with successive video graphic recordings, detection of abnormal head positions or appearance of refraction defects which could require treatment. No prominent complications were observed concerning the development of horizontal or vertical strabismus.
Conflict of interests

No conflict of interest has been declared by the authors.

REFERENCES