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Brown's syndrome with Jaw-winking phenomenon: A rare-atypical association

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Abstract

Background: Brown syndrome is a rare mechanical problem, characterized by restriction of the superior oblique trochlea-tendon complex. The Marcus Gunn Syndrome is one of the more common congenital oculofacial synkineses that occur with a brief rising of the upper eyelid each time the jaw moves, calling Jaw-winking phenomenon, often associate with strabismus and amblyopia.

Case presentation: In our case report, we describe an atypical association with this uncommon syndrome: brown syndrome in one eye and Jaw-winking phenomenon in the fellow eye.

Conclusion: It is common the association of Marcus Gunn Syndrome with eye-motility alteration, but often in the same eye. This is a rare case with Brown Syndrome and Jaw-winking phenomenon not in the same eye.

Keywords

brown syndrome; Marcus Gunn; jaw-winking phenomenon

Abbreviations

BS: Brown Syndrome; D: Dioptre; DS: Dioptre Sphere; LE: Left Eye; MGS: Marcus Gunn Syndrome; RE: Right Eye

Introduction

A 50-year-old woman arrived at our clinic with a history of breech birth, strabismus and alteration of posture and head from birth. When she was 7yearsold, she was diagnosed with Brown's Syndrome (BS) in her right eye (RE) with active and passive restriction of elevation in adduction, divergence in upgaze, down shoot in adduction, hypotropia in primary position with compensatory chin elevation [1].

When she was 20 years old she underwent surgery on the tendon (recession of superior oblique tendon RE), twice. She did not know if other family members had similar syndromes. After the last surgery for many years, she did not complain about her sight.

About 20 years ago, shecomplained of occasional diplopia. She had visual acuity of 20/30 in her right eye (RE) and 20/25 in her left eye (LE) (ETDRS visual acuity chart) with a correction of -2.50 dioptre

sphere (DS) bilateral.

Our examination showed that her head was tilted to the left and she had mild blepharoptosis of the left upper eyelid. The Hirsch berg test with LE fixation showed -20 Dioptre (D) RE exotrophy with 6 D LE/RE in near vision and -18 D RE exotrophy with 12 D LE/RE in distance. The Monolateral Cover test demonstrated the capacity of the patient to maintain fixation with the RE, therefore she was able to alternate fixation with both eyes. The Prism Bar Cover test in near and distance confirmed the exo-hypotrophy of the RE with an angle of -30 D with 10 D LE/RE in near and an angle of -18 D with 12 D LE/RE in distance.

The Worth Four Dot Test revealed an alternating suppression although fixation was mostly observed with LE.

During the Red filter test, the patient complained about"transitory diplopia" only at the moment of the switch of the fixing eye, otherwise she did not complain about this problem.

Ocular movements showed a mild down-shoot in attempted elevation in adduction, with bilateral medial rectus hypo function.

The absence of homolateral Superior Oblique hyperfunction supported the BS diagnosis. During the orthoptic examination, we observed that, while the patient was speaking, the left eyelid tended to be more open, so we asked the patient to open her mouth: the upper eyelid retracted suddenly revealing a big portion of sclera (Figure 1). When the mouth was closed (Figure 2) the eyelid returned to its normal position (ptosis). She referred that she had been experiencing this peculiar phenomenon for many years, but she had never complained about it as she was not bothered. The Jaw-winking phenomenon or Marcus Gunn Syndrome (MGS), is characterized by eyelid ptosis, usually unilateral, associated with a retraction or higher elevation than the fellow eye of the affected eyelid at the jaw movement [2-4]. No surgical intervention is usually required [4]. MGS etiopathogenesis is not well understood even though the first hypothesis proposeda co-innervation between the elevator palpebrae superior muscle from the oculomotor nucleos and external pterygoid portion of the trigeminal nucleos [2,5].

Discussion

Brown' Syndrome and Marcus Gunn Phenomenon are congenital, rare, ocular disease. The first one syndrome occurs with evident restriction of active and passive elevation in adduction [1], instead the second one occurs with involuntary elevation of a ptotic eyelid secondary to mandible movements, often unilateral [5]. In literature was described a high incidence of strabismus and amblyopia associated with Jaw–Winking Phenomenon [5]. Pratt et al found a high percentage of association with amblyopia (59%) and other diseases with lower incidence like double elevator palsy, anisometropia and superior rectus muscle palsy [8]. Brodsky et al report a case reported of a child with synergistic divergence and MGS [10]. Isenberg et al described a case report of MGS associated with Duane's retraction syndrome in the same eye [11].

Our case demonstrates an association of Brown's Syndrome with co-existing Marcus Gunn Jaw-Winking Syndrome in the fellow eye, reported in only one other cases in literature: Artifoni et al in 1965 [7].

Conclusion

MGS is often associated with other eye disorders, such as strabismus and amblyopia [5-6,8-10], frequently in the same eye, but in our case it was associated with Brown's Syndrome in the fellow eye.

Figures

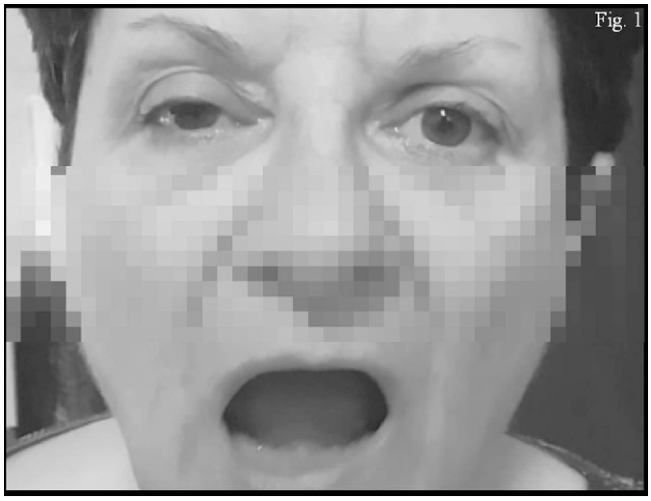

Figure 1: The pictures of the patient shows the the Jaw-winking phenomenon when the mouth is open.

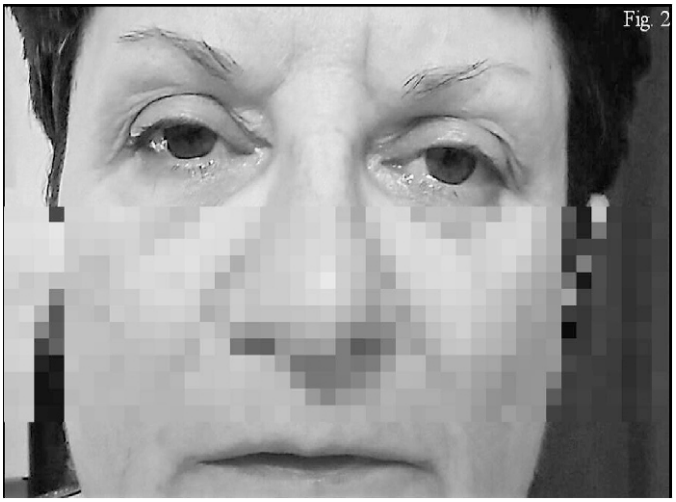

Figure 2: The picture of the patient shows the ptosis of the eyelid when the mouth is closed

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