

Pseudoptosis after Aberrant facial nerve regeneration (AFR): an under-recognized entity

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Abstract

Aberrant facial nerve regeneration (AFR) is not well recognized as a cause of pseudoptosis. Signs of AFR includes pseudoptosis, narrow palpebral aperture and prominent nasolabial fold on side of paralysis.

We present a series of six cases who had a significant pseudoptosis after facial nerve paralysis (FNP). All patients had a prominent nasolabial fold on the side of paralysis and a narrow palpebral aperture which further narrowed on the jaw movements. Margin Reflex distance (MRD)1 and MRD2 were reduced in all cases on the side of FNP. Levator palpebrae superioris (LPS) action and Bell's phenomena were good in all the cases. All the cases were diagnosed as old lower motor neuron FNP with pseudoptosis due to AFR.

In conclusion, AFR as a cause of pseudoptosis is less well recognized. It is important to keep in mind this entity for proper management and to prevent the patient's dissatisfaction.

Keywords: Aberrant facial nerve regeneration, Eyelid, Martin-Amat Syndrome, Ptosis, Marcus-Gunn jaw winking, Synkinesis.

Introduction

Pseudoptosis is a condition resembling ptosis, due to abnormalities other than those found in the eyelid elevator muscles. Causes include blepharophimosis, dermatochalasis, ipsilateral hypotropia, phthisis bulbi, microphthalmos, anophthalmos, or a decrease in orbital volume as in enophthalmos.¹ Pseudoptosis may also be caused as a delayed complication of facial nerve palsy(FNP). It occurs as a result of increased orbicularis tone due to aberrant facial nerve regeneration (AFR). AFR is often under-recognized cause of pseudoptosis.² Signs may be very subtle. Diagnosing AFR is necessary as management differs from other form of ptosis.

Case History

We had six cases of aberrant regeneration due to FNP. The previous episode of lower motor neuron (LMN) FNP was confirmed by their history, inability to raise the eyebrow, absent forehead wrinkles and inability to blow air. None of these cases had a history of diurnal variation, fatigue, diplopia, trauma and lid surgery. Three patients presented with narrow palpebral aperture and other cases had complaints unrelated to pseudoptosis. The duration between FNP and AFR ranged from 6 months to 8 years (Table 1).

All patients had reduced margin reflex distance (MRD)1 and MRD2, no lagophthalmos, prominent nasolabial folds and deviated angle of mouth on the side of FNP (Fig. 1). Levator palpebrae superioris (LPS) action and Bell's phenomena were good in all cases. The eyelid crease in downgaze was normal. The extraocular movements,

corneal sensations and pupillary examination were normal. Ice pack test was negative. Ptosis of all the patients increased with jaw movements (Fig. 2). Details of ptosis examination are given in Table 2. Case A and C had completely forgotten the history of FNP which they recalled only when they were asked the directed question about it. Case D had both eye ptosis, right eye (RE) had moderate aponeurotic ptosis, and left eye (LE) had ptosis secondary to AFR. Case F had post-traumatic FNP with severe lagophthalmos of LE for which temporary tarsorrhaphy was done. Tarsorrhaphy was released after 3 months. After the release of his tarsorrhaphy, his LE further narrowed to the present state.



Fig. 1: Showing features of Aberrant Facial Nerve Regeneration

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Table 1: Demography and Presenting Symptoms

Case No.	Age	Sex	Duration between AFR and FNP	Presenting Symptom
1.	50	F	1 year	Small LE
2.	40	F	4 year	Small LE
3.	55	F	4 year	Irritation LE
4.	50	F	6 months	Decreased vision
5.	55	F	8 years	Consultation for DR, HR
6.	40	M	8 months	Small eye

RE: Right eye; LE: Left eye; AFR: Aberrant facial nerve regeneration; F: Female; M: Male; FNP: Facial nerve paralysis, DR: diabetic retinopathy, HR: hypertensive retinopathy

Table 2: Findings of Ptosis examination

Case No.	Visual Acuity		LPS Action in mm		MRD1/ MRD2 in mm		VPFH in mm		Amount of ptosis in mm	
	RE	LE	RE	LE	RE	LE	RE	LE	RE	LE
1.	20/20	20/100	12	12	4.5/ 5.5	-1.0/ 4.5	10.0	3.5	-	5.5
2.	20/20	20/20	15	14	4.5/ 5.5	0.0/ 4.5	10.0	4.5	-	4.5
3.	20/50	20/100	11	12	2.5/ 5.5	1.0/ 4.5	7.0	5.5	2.0	3.5
4.	20/20	20/20	14	13	4.5/ 6.0	3.5/ 5.0	11.5	8.5	-	1.0
5.	3.28/200	20/200	14	14	3.0/6.0	1.0/4.5	9.0	5.5	1.5	3.5
6.	20/20	20/20	12	12	4.5/4.5	1.0/4.0	9.0	5.0	-	3.5

RE: Right eye; LE: Left eye; LPS: Levator palpebrae superioris; MRD: Margin Reflex distance; VPFH: Vertical palpebral fissure height



Fig. 2: Martin Amat Syndrome

Discussion

We present here a series of six cases of pseudoptosis caused by AFR which is a delayed complication of LMN FNP. Timely diagnosis of cases prevented unnecessary investigations and LPS surgery.

Usually, the patients of FNP present with lid retraction, wide palpebral aperture (increased MRD1 and MRD2), lagophthalmos on the paralysed side, and prominent nasolabial folds and deviation of angle of mouth towards the opposite side. On the contrary, all of our patients had a narrow palpebral aperture (reduced MRD1 and MRD2), prominent nasolabial folds and a paradoxical deviation of angle of mouth towards the side of FNP without any lagophthalmos. Presence of these signs on the side of FNP

is an indicator of AFR. This occurs due to increased tone of orbicularis and other facial muscles. AFR is not a well-recognized cause of pseudoptosis. These signs may be subtle. They can be made prominent by instructing the patients to puff their cheeks which increases pseudoptosis and eyelid closure.^{2,3,4}

When the initial FNP is mild, recovery is usually complete and rapid. However, in severe initial damage, there is faulty regeneration within the facial nerve itself leading to AFR with proprioceptive impulses associated with muscle stretch acting as the trigger.⁵ This causes synkinesis leading to hypertonic facial muscles i.e. they contract when they should be at rest. Unlike other skeletal muscles, facial muscles lack spindles. Without these spindles, there is no awareness of the contraction, and the muscles remain in a state of constant tension.

Hemifacial spasm (HFS) is a close differential diagnosis. HFS is characterized by irregular, involuntary muscle contractions on one side of the face whereas muscle is in a constant state of contraction in AFR. HFS may occur due to *emphatic transmission* which is an electrical activity crossing from one demyelinated neuron to another resulting in a false synapse,⁶ *abnormal activity of axons at the facial nerve root* secondary to compressive damage/demyelination⁷ and "*Kindling*" which involves increased excitability of the facial nerve nucleus due to feedback from a damaged facial nerve.⁸

It is important to observe the effect of jaw movement on ptosis. Martin Amat syndrome (MAT) is a form of acquired facial synkinesis manifesting as involuntary eyelid closure on jaw movement. It was present in all of our six cases (Figure 2). MAT should not be confused with Inverse Marcus-Gunn phenomenon. Though both have eye closure on jaw movement, the mechanism of synkinesis is different

in both. The term inverse Marcus-Gunn syndrome should be reserved only for a congenital lesion, where the mechanism of lid closure is because of inhibition of the LPS rather than orbicularis oculi contraction as seen in MAT. MAT should also be differentiated from Marcus-Gunn Jaw winking Phenomena (MGJWP), a congenital form of synkinesis where there is upper eyelid elevation. The synkinetic movements in Marin-Amat syndrome are opposite to that seen in MGJWP.^{5,8,9,10}

Botulinum toxin injection in orbicularis muscle is helpful in management.¹¹

As pseudoptosis simulates aponeurotic ptosis, there is a potential risk of a patient undergoing LPS surgery if the condition is not recognized. LPS is normal functionally and structurally. Doing LPS surgery may give an unpredictable postoperative upper lid height as a result of increased orbicularis tone which can lead to patient's dissatisfaction.² In cases of lagophthalmos in FNP where the risk of corneal exposure is high, a temporary procedure like tarsorrhaphy should be done. A correction with an irreversible permanent procedure (LPS recession, Spacer graft) should not be done early in course because if these patients develop AFR it may further narrow down the palpebral aperture. These patients should be followed up at periodic intervals as the reported incidence of synkinesis in facial nerve palsy varies greatly ranging from 9 to 55%.¹² The exact timing when a permanent procedure should be done is still a dilemma as AFR may develop from 4months to years.^{2,3}

In conclusion, AFR is not a well-recognized cause of pseudoptosis. It is important to diagnose it for the proper management and to prevent the patient's dissatisfaction.

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Conflict of Interest

None.

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