Case report

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Clinical Case of Facial Muscles Function Recovery in a Patient with Idiopathic Lesions of the Facial Nerve with Unfavorable Myographic Predictors

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ABSTRACT

The facial nerve (FN) palsy is a disease of the peripheral nervous system that leads to aesthetic, organic and functional disorders. The causes of the disease are different, the most common is the idiopathic form Bell's palsy. With a conservative approach to therapy, up to 80% of patients note the complete recovery of the lost functions of facial muscles, the others have the likelihood of severe consequences. Electroneuromyography is used to assess the risk of a negative outcome of the disease. In cases of confirmation of severe nerve damage, surgical treatment is suggested. The article presents a clinical case of complete recovery of the function of facial muscles in a patient with severe damage to the a nerve and an unfavorable prognosis. The course of the disease, complications of Bell's palsy and methods of their correction are described. Timely correct choice of treatment tactics and prevention of complications minimizes negative consequences. When working with a patient, a differentiated approach is important depending on the clinical situation.

Keywords: facial nerve palsy, Bell's palsy, facial asymmetry, synkinesis, electroneuromyography

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BFB – biofeedback
BTA – botulinum toxin type A
CT – computed tomography
ENMG – electroneuromyography
FN – facial nerve
MRI – magnetic resonance imaging
NEMG – needle electromyography
PB – Bell's palsy
PMU – potential of motor units
TE – therapeutic exercises

INTRODUCTION

Neuropathy of the facial nerve (FN) is a common disorder of the peripheral nervous system. The disease is characterized by one-sided decrease in strength or complete disappearance of movements in the facial muscles of the face [1], leading to aesthetic, organic and functional consequences. The weakness of the facial muscles can be combined with secondary disorders of vision, hearing, taste, speech and eating disorders. These disorders significantly worsen the quality of life of patients, often lead to depression and social isolation [2]. The reasons for the development of the disease can be very diverse: tumors, trauma, infections, metabolic disorders, vascular disorders, and others [3]. However, the pathology can be idiopathic, in which case it is called Bell's palsy (PB). PB is a kind of diagnosis of exclusion.

With a conservative approach to therapy, up to 80% of patients have complete restoration of the lost functions of facial muscles in PB, in 13–15% symptoms remain mild, and the rest of the patients remain with severe consequences [4]. Surgical treatment of PB, which consists in decompression of the FN canal, is usually recommended in cases of severely unfavorable prognosis of facial muscle recovery. However, the effectiveness of this method is still the subject of scientific discussion. To assess the likelihood of a negative outcome of the course of PB, a clinical assessment of the functional state of facial muscles, as well as data from stimulation and needle electromyography (ENMG and NEMG) are used. A decrease in the amplitude of the M-response by 90% and lower in comparison with the healthy side and the absence of motor unit potentials (PMU) during voluntary muscle contraction are the most accurate instrumental predictors of an unfavorable outcome [5]. We report a case of successful restoration of the function of facial muscles in a patient with severe BP and myographic predictors of an unfavorable outcome.

Case report

Patient I., 27 years old, became acutely ill in December 2018, when, against the background of complete well-being, a significant asymmetry of the face appeared due to the absence of movements of the facial muscles of his right half. The patient had no other complaints. From the anamnesis it is known that the patient suffers from chronic ulcerative colitis of unspecified etiology, in connection with the disease, he regularly undergoes a long course of treatment with antibacterial and hormonal drugs. A clinical examination revealed that movements in the mimic muscles of the right half of the face were absent, the tone in them was reduced, according to the House-Brackmann scale, the VI degree of damage to the right FN was established. Otherwise, the neurological status was normal.

According to instrumental research methods, no pathology was revealed (magnetic resonance imaging (MRI) of the brain, computed tomography (CT) of the temporal bones). Laboratory indicators (clinical, biochemical, coagulological blood tests, general urine analysis) did not exceed the permissible values. Antibodies to the herpes simplex virus - anti-HSV (type 1) IgG 10.9 (the result is positive if the value is more than 1.1) were revealed. The Bell's palsy was revealed, prosoplegia on the right, treatment was prescribed - prednisolone 60 mg per day for 5 days with a dose reduction, valacyclovir 500 mg 2 times a day for 10 days, kinesio taping of the muscles of the right half of the face. It was recommended to refrain from carrying out active physiotherapy exercises (LFK) due to the lack of movement in the affected muscles and data on the degree of nerve damage (Fig. 1).

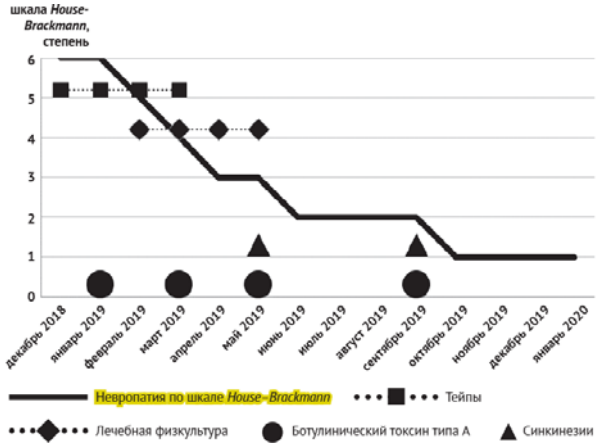

Fig. 1. Correlation of clinical manifestations dynamics of the right facial nerve neuropathy according to the House–Brackmann scale and methods of treatment in patient I. at different stages of the disease

The graph on the X-axis represents the severity of clinical manifestations of neuropathy on the House – Brackmann scale, on the Y-axis - the patient's recovery time. The graph reflects how the complex of therapeutic measures changed depending on the dynamics of the clinical picture (types of medical intervention were prescribed and canceled).

On the 7th day of the disease, the patient underwent ENMG and NEMG (Fig. 2). According to ENMG data, signs of pronounced axonopathy of the right FN were revealed, while the decrease exceeded 90% in comparison with the healthy side. With NEMG, the circular muscle of the eye and mouth was examined. In both cases, it was not possible to register PMU on the affected side during an attempt to voluntary muscle contraction.

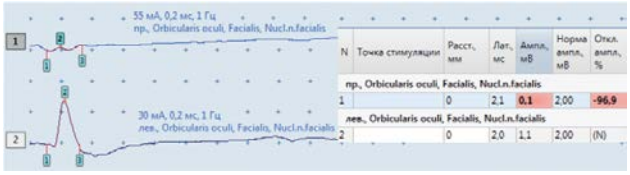

Fig. 2. Asymmetry of M-responses. Curve 1 — M-response of the right orbicular muscle of the eye. Curve 2 is the response of the left orbicular muscle of the eye. The difference in the amplitudes of the M-responses is 96.9%

According to the results of the myographic study, a neurosurgeon's consultation was indicated: the absence of positive dynamics from conservative therapy and the presence of damage to the axons of the right FN more than 90% according to ENMG data indicate a high risk of an unfavorable functional outcome, in connection with which, in order to improve the prognosis of restoration of the volume of movements of facial muscles, an operation is indicated - decompression of the intratemporal (labyrinth) segment of the right FN in the middle cranial fossa. The patient was recommended to continue kinesio taping of the right half of the face to improve blood circulation, stimulate the lymphatic and nervous systems, tone the muscles, and also refrain from excessive facial activity (avoid excessive facial movements with maximum amplitude). The patient underwent preparation for surgical treatment, but later refused.

A month after the onset of the disease, the clinical degree of damage to the mimic muscles of the right half of the face was assessed as plegia, according to the House – Brackmann scale - grade VI (Fig. 1; Fig. 3, row 1). The mimic muscles of the left half of the face (conditionally healthy) became hyperactive, their tone increased. Injections of botulinum toxin type A (BTA) were performed in a conditionally healthy side, and 30 U of incobotulinum toxin (drug) was injected into the target muscles at a time [6–9]. At a follow-up examination a week after botulinum therapy, the patient reported that he had sensations of movement in the corner of the mouth from the affected side. Positive dynamics was clinically noted, minimal movements appeared in the mimic muscles of the lower third of the face, according to the House – Brackmann scale, the V degree of damage to the right FN was established (Fig. 3, row 2). An EMG was performed: when examining the circular muscle of the eye and the circular muscle of the mouth on the right, moderately pronounced spontaneous activity in the form of fibrillation potentials was revealed, and with an arbitrary contraction - single PMU. Taking into account the appearance of movements in the facial muscles of the affected half of the face, the patient was trained in exercise therapy according to the Kabat method [8].

Fig. 3. Photo registration of the patient's examination before treatment (1), in February, one week after the first injection of botulinum toxin type A (2), one year after treatment (3). A — at rest; B — the eyebrows are raised as much as possible; C — smile at maximum amplitude

The effect of BTA injections persisted for 8 weeks. Further, the excessive activity of the muscles of the conditionally healthy half of the face returned, their tone increased (the duration of the disease was 3 months), so the patient again began to experience discomfort. Clinically, there was a positive dynamics in the form of an increase in the amplitude of the smile on the affected side, according to the House – Brackmann scale, grade IV of the right FN was found. The patient reported that after the BTA injection he underwent a course of treatment for colitis, with which he associated a decrease in the effect of the procedure. A repeated botulinum therapy procedure was performed - 30 U of incobotulinum toxin was injected into the target muscles of a conventionally healthy half of the face at a time. A follow-up examination a week after botulinum therapy showed clinically positive dynamics, movements appeared in the mimic muscles of the middle third of the face, the eye began to close with an effort, according to the House-Brackmann scale, grade III of the right FN was established.

Eight weeks after the second procedure of botulinum therapy, the patient came to an appointment with complaints of involuntary contractions in the muscles of the affected side of the face (disease duration - 5 months). Clinically, there was a positive dynamics - less effort was required to close the eye, movements appeared in the upper third of the face, according to the House-Brackmann scale, grade II-III damage to the right FN was established. However, oro-ocular synkinesia was revealed, as well as a feeling of constriction in the area of the platysma on the right. The third injection of BTA was performed in the affected side in order to relieve synkinesis, in the conditionally healthy side to maintain the symmetry of the face. 15 Units of incobotulinum toxin were injected at the same time into the target muscles of the right half, 26 Units - in the left. It is recommended to refrain from performing active exercise therapy. A follow-up examination one week after the third botulinum therapy showed clinically positive dynamics, improved movements in the mimic muscles of the upper and middle third of the face, the smile became symmetrical when it was visually monitored with a mirror, the House-Brackmann scale established II degree of damage to the right FN. During provocative tests, as well as at rest, synkinesis was not revealed.

At a scheduled appointment 16 weeks after the third procedure of botulinum therapy (the duration of the disease was 9 months), the patient reported that the symmetry of the face had practically recovered, but mild synkinesis was again disturbing. According to the House – Brackmann scale, grade II damage to the right FN

was established; oro-ocular synkinesia on the right was objectively revealed, which intensified after provocative tests. The fourth injection of BTA (incobotulotoxin) was performed into the mimic muscles of the affected side at a dose of 15 U in order to relieve synkinesis, in a conventionally healthy side at a dose of 26 U - to maintain facial symmetry. At the follow-up examination a week after the fourth procedure of botulinum therapy, the face was symmetrical at rest and in motion, synkinesis was not detected both at rest and during provocative tests.

On a routine examination 16 weeks after the fourth procedure of botulinum therapy (the duration of the disease is 13 months), the patient had no complaints. The volume of active movements of the facial muscles was full, the face was symmetrical at rest and during movement, objectively synkinesis was not revealed (Fig. 3, row 3). The patient was advised to apply for an appointment in case of facial asymmetry or involuntary movements in the face and neck area.

DISCUSSION

Specialists of the N.V. Sklifosovsky Research Institute for Emergency Medicine often consult patients with acutely developed facial asymmetry, with PB as one of the reasons. The prevalence of the disease is 20–32 observations per 100,000 population per year [3], which indicates the urgency of the problem. The difficulty in working with patients with PB is the lack of uniform standards for diagnosis, treatment and rehabilitation [8].

Clinical methods for assessing FN function include generally accepted neurological examination and special scales that allow objectifying the severity and dynamics of the process. According to the Facial Nerve Disorders Committee of the American Academy of Otolaryngology - Head and Neck Surgery, the House-Brackmann scale (1985) is the "gold standard" for diagnosing FN function, which takes into account the movement of the eyebrow, closing the eye, raising the angle of the mouth on the affected side in comparison with a conditionally healthy side (Table 1). This scale makes it possible to assess the degree of dysfunction of facial muscles in six degrees, where I degree is the absence of dysfunction, and VI is total damage [10]. In the clinical analysis, the assessment is made on the House – Brackmann scale, but it should be noted that it does not take into account all the clinical features of the course of the disease.

Table 1

House– Brackmann scale

Degree of impairement	Description	At rest	In motion
I	Norm	The function of the facial nerve is not changed	The function of the facial nerve is not changed
II	Mild dysfunction	Symmetrical facial expression	Slight asymmetry of the angle of the mouth when smiling
III	Moderate dysfunction	Symmetrical facial expression	Inability to fully raise the eyebrow upward, moderate asymmetry of the mouth when smiling
IV	Moderate dysfunction	Symmetrical facial expression	The eyebrow does not rise, the eye does not close completely, the asymmetry of the mouth at maximum effort
V	Severe dysfunction	Severe facial asymmetry	Movement on the affected side is barely noticeable. Inability to close the eyes, weak movement of the corner of the mouth
VI	Total dysfunction	Decreased muscle tone	Complete lack of movement

Patient I. underwent a full range of necessary paraclinical studies. To exclude masses and alternative diagnoses, the patient underwent MRI of the brain and CT of the temporal bones [11]. Herpes simplex virus is considered the main probable cause of PB. As a result of a viral infection, FN edema develops, it is compressed in the temporal bone canal [12, 13]. That is why patient I. was tested for antibodies to the herpes simplex virus, but no data on the presence of an acute process were obtained.

The patient was prescribed treatment in accordance with modern clinical guidelines. All patients with PB within 3 days from the moment the first symptoms appear, corticosteroids are prescribed, as well as antiviral drugs in case of confirmed infection and / or with severe paresis or plegia of facial muscles [11; 14]. There are no clinical recommendations regarding the use of physical therapy in the acute period of PB. In the studies conducted, the groups of subjects were heterogeneous, therefore, it is not possible to evaluate and compare the results [8]. Doctors are currently applying positional treatment using adhesive plaster

immobilization and a more modern method - kinesio taping. Kinesio taping in complex treatment limits the development of asymmetry of nasolabial folds, the development of synkinesis, promotes early recovery, but has no evidence base, since the method was previously used in all studies in conjunction with other methods of treatment [8]. Patient I. underwent kinesio taping from the first days, which, in our opinion, made it possible to avoid overstretching of the paretic muscles and to maintain facial symmetry in the future. When moderate movements appear in the affected side, kinesio taping is canceled. Therapeutic exercises were recognized as one of the most effective methods of physical rehabilitation [8], however, patient I. was advised to refrain from performing active exercises due to the absence of movements in the facial muscles and a high percentage of FN damage according to myography data. Therapeutic exercises in this clinical case could lead to an aggravation of the situation, an increase in asymmetry due to the absence of a positive effect on the affected side and excessive activation of the muscles of the conventionally healthy side.

With the lesion of FN in the acute and in the chronic period, it is necessary to consider not only conservative treatment, but also the possibility of an operative one [8]. One of the most common types of surgical intervention at an early stage in the development of plegia of facial muscles is the method of decompression of the labyrinth segment of the facial nerve with a pronounced degree of damage according to clinical and myographic criteria. However, there is no clear understanding of the time frame in which one can reliably judge the prognosis of the disease, the benefits and risks of the operation. We faced this problem when assessing the clinical situation of patient I.

Esslen E. (1973) with the help of ENMG established that in case of FN lesion, axonal degeneration of more than 90% is critical, is detected already on the fourth day after the onset of symptoms and corresponds to a poor recovery prognosis. The author was the first to suggest using myography as a method of early diagnosis and predicting outcome in neuropathy [5, 15, 16]. According to L.A. Isakova and G.O. Penina (2017), ENMG should also be performed no earlier than 3-5 days after the development of neuropathy, since there is a delay in changes in ENMG when compared with the clinical picture [17]. L. Thomander, E. Stalberg (1981) specified that the maximum difference in the amplitude of the M-response of the affected and conditionally healthy side is observed on average on the 10th day from the onset of symptoms [18]. According to N.P. Gribova and O.S. Galitskaya (2009), myographic study has a higher informative value on days 10-14 from the onset of the disease, especially with a combination of ENMG and NEMG [19]. Neuromuscular restructuring is likely to occur during the first 10 days of illness [20], while degeneration of FN nerve fibers (Wallerian degeneration) may occur within the first 72 hours [21, 22]. Spontaneous activity (SA) can be recorded at different stages of the disease in the form of significant fibrillation potentials and positive acute waves, which is also a poor prognostic sign [23]. ENMG and NEMG were performed on the 7th day of the disease (Fig. 2). According to ENMG data, signs of significant axonopathy of the right FN were revealed, the decrease in the amplitude of the M-response of which exceeded 90% compared to the healthy side. With NEMG, the orbicular muscles of the eye and mouth were examined. In both cases, it was not possible to register PMU on the affected side during an attempt to voluntary muscle contraction. In the case presented by us from practice, the patient had relative indications for surgical treatment, since according to the data of myography and clinical examination, the prognosis of restoration of facial functions was unfavorable. In the study by U. Fisch (1981), 14 patients with Bell's palsy and a decrease in the amplitude of the M-response on the affected side by more than 90% underwent decompression of the FN labyrinth segment. The author noted the best outcomes of the disease after surgical treatment in comparison with the results in 13 patients who refused surgery [24]. B.J. Gantz et al. (1999) offered an algorithm for choosing treatment tactics depending on the EMG results: surgical treatment is indicated for total plegia of facial muscles for a period of up to 14 days from the onset of the disease, asymmetry of M-response amplitudes of more than 90% according to ENMG and the absence of PMU when trying to voluntary muscle contraction (Fig. 4). In other cases, the authors recommended the use of prednisone. According to the results of the study, the best outcomes were observed after surgical treatment in comparison with conservative treatment (91% versus 42%) [25, 26]. Similar observations were published by L. Thomander, E. Stalberg and B. Mamoli (1981) [18, 27].

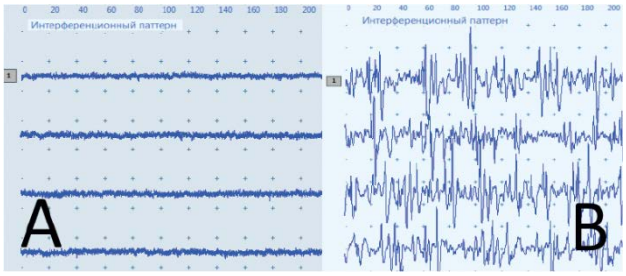

Fig. 4. Myographic image during voluntary contraction of the orbicular muscle of the eye: A — right orbicular muscle of the eye; B — left orbicular muscle of the eye

The risk of complications of surgical treatment is 3–10% for damage to the auditory nerve and 4% for liquorrhea; therefore, it is not recommended to be used as a routine method of treating PB [11]. The decision on the choice of treatment tactics for PB in case of poor prognosis should be made individually. We presented a case from practice, in which, despite a poor prognosis, there was a complete restoration of the function of the facial muscles. However, it cannot be ruled out that the choice of surgical tactics would lead to faster recovery.

With the development of acute PB, facial asymmetry occurs due to the weakness of the affected (ipsilateral) side. The conditionally healthy (contralateral) side is compensatory for the work, over time it becomes hyperactive, the facial muscles of the face are hypertrophied, the asymmetry of the face is aggravated. In patient I., a month after the onset of the disease, there was no dynamics on the affected side, however, the conditionally healthy side had already become more active. Starting from the acute period of FN neuropathy of any genesis, BTA injections can be performed. The procedure is carried out exclusively in the mimic muscles of the conventionally healthy side in order to correct asymmetry, prevent the development of hyperkinesis and hypertrophies, and also more quickly restore the innervation of the affected side. Experimental studies have shown that temporary chemical denervation with BTA on the contralateral side improves morphological and functional regeneration of the FN after its damage. With the strengthening of the evidence base, botulinum therapy can be a standardized and recommended technique [7, 8]. In the described clinical case, the first injection of BTA was carried out a month after the onset of the disease and led to the activation of the affected side.

The introduction of BTA in the chronic period into the conditionally healthy side is also safe and effective at any time of the disease, and the asymmetry of the face decreases not only due to the relaxation of hyperactive hyperactive muscles of the contralateral side, but also to some increase in muscle strength on the ipsilateral side [7, 8]. The presented graph shows that after each injection of BTA in patient I., an increase in the strength of the facial muscles of the affected side of the face was observed.

When the first movements appeared in the facial muscles, the patient was trained in the methods of exercise therapy, since exercise therapy is one of the most effective methods of physical rehabilitation, especially in combination with biofeedback (BFB) [7-9]. Our experts recommend using a mirror as a biofeedback.

After damage to the FN, even with its optimal reconstruction, innervation is disturbed due to incorrect orientation of axonal sprouting and polyinnervation of the end plates of the facial muscles. Over time, pathological movements develop on the affected side - synkinesis, which affect the symmetry of the face, are difficult to control medically and non-medically. Patient I. had synkinesis on the 5th month of the disease. A lot of research is devoted to prevention and treatment of synkinesis using BTA. When the drug is injected only into the muscles of the affected side, the patient's face will be asymmetric; therefore, the introduction of BTA into the mimic muscles of both the affected and conditionally healthy half of the face is justified [7, 8]. T. Kasahara et al. (2017) offered the use of kinesio taping in the development of orocular synkinesia; in a small study, the method showed its effectiveness. However, kinesio tape was applied to the face in an unconventional way (around the mouth) and the sample of patients was extremely small [28]. Currently, our specialists do not use this technique, since more convincing data on the effectiveness of the method are required. The treatment of synkinesis in Patient I was successfully carried out by the administration of BTA.

CONCLUSION

The given clinical example demonstrates the successful restoration of the function of facial muscles in a patient with Bell's palsy in the course of conservative therapy with instrumental predictors of an unfavorable outcome. In this regard, we believe that the data of electroneuromyography / needle electromyography should be taken into account as an additional point. The algorithms for the timing of the

study require clarification, but it is recommended to start it no earlier than 7 days from the first signs of the disease. The development of synkinesis remains a complex problem and requires timely detection, therefore, patients with PB should be warned of the risks of developing delayed complications and be under the supervision of a neurologist, and have a schedule of scheduled examinations.

Timely correct choice of tactics for treatment and prevention of complications minimizes the negative consequences of this disease.

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