Supranuclear Pathways for Facial Movements


Abstract

The authors report on the case of a young man with a mild head injury and an isolated palsy of voluntary facial movements, attributed to a midbrain traumatic hematoma. This exception to the generally accepted conjunction between brainstem contusion and poor prognosis pertains to a special entity of midbrain laceration due to hyperextension of the head, with minimal influence on the level of consciousness. The clinical presentation of this lesion with facial palsy sparing emotion-related movement has rarely been described and offers a clue for exploring the neuroanatomy of facial movement.

According to the authors, this is a case of supranuclear facial palsy at the level of the cerebral peduncle. This explanation is plausible, because “in supranuclear lesions there may be a dissociation of emotional and voluntary facial movements; often, some degree of paralysis of the arm and leg or aphasia (in dominant hemisphere lesions) is conjoined,” as has happened in the described patient.

First of all, there is the problem that the contusion seems to be outside the cerebral peduncle (Figs. 1A and 3A of the article), while, as correctly described by the authors, “the corticobulbar fibers for face movement lie medially to the ones for the extremities in the cerebral pedicle.” The corticospinal tract of this patient was probably injured at the level of the internal capsule and not at the level of the midbrain peduncle. However, the main problem is that this is obviously a case of upper facial paresis, which should not occur in a supranuclear facial injury, given that the corticopontine innervation of the upper facial muscles is bilateral.

This bilateral innervation occurs at the level of the pons, near the facial nucleus. An alternative hypothesis is that this case could be a kind of facial apraxia.

Facial (including orofacial and buccofacial) apraxia occurs in areas connected to the premotor cortex. We describe such a case from our department. A 25-year-old woman suffered a head injury after a car accident. She was initially lethargic, but her level of consciousness quickly improved to normal. She had right facial paresis, as is typical in cases of facial nerve palsy. Facial movements were easier during mimicry. She could sometimes perform spontaneous movements of her face, which she could not do on demand. She had difficulty in her verbal expression; verbal pronunciation was better during repetition. She also has difficulty swallowing, with diminution of her gag reflex, which was not accompanied by hypesthesias of the oropharynx. All these clinical signs fluctuated at consecutive clinical examinations (Fig. 1).

A neuroradiological investigation revealed contusions in the areas of Broca and left lower parts of the precentral gyrus, in central motor areas for the face and oropharynx, as well as adjacent perisylvian areas. There were no brainstem abnormalities (Figs. 2–4).

This was a case of orofacial dyspraxia, which is characterized by difficulty in facial movements, easier movements after mimicry, and spontaneously and fluctuation in their clinical presentation. It is also characterized by dyspraxia in talking, especially in difficult syllables, easier mimicry, and fluctuation in expression of language. Finally, it is characterized by diminished gag reflex without hypesthesia of the oropharynx.

Clinical and imaging studies show a strong correlation between orofacial dyspraxia and lesions in the frontal operculum. This condition may also occur with subcortical lesions involving periventricular and/or peristriatal white matter as well as the basal ganglia (circuit of cortex/basal ganglia/thalamus/cortex).

Fig. 1. Left: Photograph illustrating facial dyspraxia. Right: Photograph illustrating oral dyspraxia (published with permission of the patient).
Finally, it is well known that oropharyngeal dyspraxia can accompany the mutism associated with removal of vermian tumors (circuit of cortex/cerebellum/thalamus/cortex).¹

Orofacial dyspraxia can happen in patients with lesions of areas connected to the premotor area. We must remark that subcortical and basal ganglia lesions tend to produce mainly emotional disturbance of facial movements.

The case described by the authors is quite different from the one that we have described.

In our opinion, the patient the authors have described has suffered a mixed lesion. If we compare Fig. 3A of the original article with a standard anatomical atlas,² we can see that the upper part of the lesion is partly within the internal capsule, but mainly within the ventral anterior nucleus of the thalamus, which is well known as part of the prefrontal loop.³ It is probably an interesting case combining the clinical picture of supranuclear facial nerve palsy with that of orofacial dyspraxia. Such a combination could explain the entire spectrum of the clinical picture of the patient.

Nikolaos Sakellaridis, M.D.
Christolos Kelesis, B.Sc.
KAT National Hospital
Kifissia, Attica, Greece

References

RESPONSE: In regard to the point of view expressed by Drs. Sakellaridis and Kelesis, there are a number of facts we wish to point out.

At first, our patient’s photographs have probably been misinterpreted, as it is clear that he suffers from central facial palsy. In addition, the presence of a definite supranuclear lesion combined with the absence of petrous bone fractures renders the hypothesis of peripheral facial lesion unacceptable.

Drs. Sakellaridis and Kelesis propose an alternative concept to explain the findings in our patient, the one of facial
The phenomenon of isolated voluntary central facial palsy per se is not uncommon and occurs typically in cases of lesions of the internal capsule, along with hemiplegia. This voluntary/emotional dissociation is attributed to a presumed different pathway considering the emotional components of facial movements. Drs. Sakellaridis and Kelesis propose the term “apraxia”; we disagree with this, given that in our case there is a clear anatomical lesion interrupting directly the central connections of the facial nerve. Moreover, our patient did not present with fluctuations in his condition, but, on the contrary, had a constant facial palsy which persisted only to voluntary movements.

The case presented by Drs. Sakellaridis and Kelesis is indeed a good example of facial apraxia as the lesion does not affect the facial pathway, but its presumed connections to other, “modulating” centers. Interestingly, in other cases the emotional portion of the facial movements is affected to other, “modulating” centers. Interestingly, in other cases the emotional portion of the facial movements is affected with such lesions.

Finally, the imaging studies in our patient reveal a hematoma extending from midbrain to the thalamus. The absence of hemiplegia implies the integrity of the internal capsule. Furthermore, the fact that mainly the midbrain was injured in this patient is explained by the mechanism of trauma (hyperextension), which is the essence of our article and is supported by the relevant literature.

References

Evaluation Methods in the Assessment of Peripheral Nerve Regeneration

To the Editor: We read with interest the article by Vleggeert-Lankamp on the reliability of various methods for peripheral nerve regeneration (Vleggeert-Lankamp CL: The role of evaluation methods in the assessment of peripheral nerve regeneration through synthetic conduits: a systematic review. Laboratory investigation. J Neurosurg 107:1168–1189, December, 2007).

Abstract

Object. A number of evaluation methods that are currently used to compare peripheral nerve regeneration with alternative repair methods and to judge the outcome of a new paradigm were hypothesized to lack resolving power. This would too often lead to the conclusion that the outcome of a new paradigm could not be discerned from the outcome of the current gold standard, the autograft. As a consequence, the new paradigm would incorrectly be judged as successful.

Methods. An overview of the methods that were used to evaluate peripheral nerve regeneration after grafting of the rat sciatic nerve was prepared. All articles that were published between January 1975 and December 2004 and concerned grafting of the rat sciatic nerve (minimum graft length 5 mm) and in which the experimental method was compared with an untreated or another grafted nerve were included. The author scored the presence of statistically significant differences between paradigms.

Results. Evaluation of nerve fiber count, nerve fiber density, N-ratio, nerve histological success ratio, compound muscle action potential, muscle weight, and muscle tetanic force are methods that were demonstrated to have resolving power.

Conclusions. A number of evaluation methods are not suitable to demonstrate a significant difference between experimental paradigms in peripheral nerve regeneration. It is preferable to apply a combination of evaluation methods with resolving power to evaluate nerve regeneration properly.

Reliability of experimental methods in demonstrating significant differences between experimental paradigms in nerve regeneration is a key issue in light of translating basic science results to patients, and we wish to congratulate the author for having critically and extensively treated this issue. However, we think that the author did not address 2 methodological points that are very important for the interpretation of experimental nerve repair data—namely the methods for sampling nerve fibers and the availability of alternative methods for the behavioral assessment of motor function recovery—and we wish to draw the author’s attention to these 2 points.

Regarding the first point, the essence of sampling is represented by the selection of a part of a finite population of objects that allows the researcher to infer conclusions about the entire population being guided by the body of principles and procedures for statistical inference. If faults occur with the sampling strategy, the investigator will infer an erroneous conclusion. Despite the widely acknowledged importance of this issue in experimental medicine and biology, most papers reporting quantitative data on peripheral nerve regeneration, including those cited by Vleggeert-Lankamp, contain either methodological sampling errors or, more commonly, insufficient methodological information on the sampling strategy to allow a clear evaluation of the reliability and significance of the results. In the case of peripheral nerve fibers, bias can occur at 2 main levels: 1) the selection of the histological sampling fields throughout the nerve profile; and 2) the inclusion/exclusion rules for the nerve fibers crossing the sampling fields (edge effect). In the selection of the histological sampling fields, it must always be kept in mind that nerve fibers are usually heterogeneously located throughout a nerve cross-sectional profile. Therefore, it is fundamental the adoption of a sampling strategy that guarantees that all areas of the nerve have the equal opportunity of being sampled (“equal opportunity rule”). Unfortunately, most papers provide no, or little, information on the procedure used to select the histological sampling fields. In some reports, authors have indicated that the sampling fields were selected as “representative” without providing any information about the criteria for the identification of a “representative” sampling field (an approach that is subjected to the investigator-related bias given that the researcher might involuntarily influence...
Neurosurgical forum

the final quantitative estimations by subjectively choosing the representative sampling fields. Even more undervalued is the importance of the rules for determining whether an axon profile crossing the sampling field borders shall be included or excluded from the sample. In fact, variability in the morphology of nerve fiber profiles (especially in their size) causes significant differences in the probability of profiles being intersected by the sampling frame edges: larger fibers will have a higher probability of intersecting the frame edges and thus partially falling in more that one sampling field than smaller fibers ("edge effect"). Therefore, if the investigator includes all edging fiber profiles, quantitative estimations will be biased toward a systematic overestimation of both the total number and mean size of fibers in that nerve trunk and vice versa if any edging profile is excluded from sampling. Unfortunately, most studies on nerve repair and regeneration do not provide any information on "what happens" when a fiber profile intersected the histological field edges.

To cope with the aforementioned sources of bias, the "equal opportunity" rule should be respected by adopting a set of sampling rules that assures that any histological field inside the nerve profile and any fiber profile inside the sampled fields have the same chance of being sampled irrespectively of morphological heterogeneity. Procedures based on systematic random sampling and dissector probe have been described and successfully used for comparing different peripheral nerve regeneration paradigms.

Regarding the methods for the behavioral assessment of motor function recovery, we agree with Vleggeert-Lankamp on the conclusion that the sciatic functional index is no longer the gold-standard technique to measure hindlimb function after tubulization for peripheral nerve repair. A combination of tests, each examining particular components of recovered sensorimotor function, has been recommended for an overall assessment of rat sciatic nerve regeneration. However, the variety of experimental methods used to assess behavioral recovery, especially motor abilities, often makes it difficult to compare results between studies.

An alternative test to examine the motor recovery was proposed by Thalhammer et al. as part of neurological evaluation in the rat sciatic nerve model and later applied to the nerve guidance channel research. Motor performance is evaluated by measuring the extensor postural thrust, that is, the force in grams applied to a digital balance.

In the last few years, computerized rat gait analysis has made a profound effect on the assessment of sciatic nerve recovery. Measurement of the gait-stance duration with the aid of video technique was first reported by Walker et al. This kinematic parameter was used to study functional nerve recovery after bridging a 15-mm gap in the sciatic nerve in rats with a biodegradable nerve guide. Recently, a new functional index, the toe out angle, was applied to study the recovery of a 15-mm gap repaired with a synthetic conduit. This study focused on the exorotation of the foot to score the walking pattern of the rats in the transverse plane.

We also agree with Vleggeert-Lankamp about the importance of considering the sensory perception in recovery from sciatic nerve injury. In our lab, instead of electrical or mechanical stimulation we have been using the heat stimulation to evoke the withdrawal reflex of the hindlimb.

This differs substantially from the traditional hotplate recordings, by measuring the withdrawal reflex latency of a single limb.

Finally, we fully agree with Vleggeert-Lankamp that a combination of evaluation methods is absolutely recommendable and we hope that the methodological issues that we have addressed in this letter might complete those reported by Vleggeert-Lankamp and help both experimental researchers in selecting the most appropriate experimental protocol for nerve repair investigation and clinicians in correctly interpreting the results of basic science studies.

Stefano Geuna, M.D.
University of Turin School of Medicine
Turin, Italy

Artur S. P. Varejão, D.V.M., Ph.D.
University of Trás-os-Montes e Alto Douro, CITAB
Vila Real, Portugal

References

14. Varejão AS, Cabrita AM, Patrício JA, Bulas-Cruz J, Gabriel RC,
Intracraniatal Hypotension

To THE EDITOR: We read with great interest the article by Zada et al. (Zada G, Pezeshkian P, Giannotta S: Spontaneous intracraniatal hypotension and immediate improvement following epidural blood patch placement demonstrated by intracraniatal pressure monitoring. Case report. J Neurosurg 106:1089–1090, June, 2007).

Abstract

The presentation of spontaneous intracraniatal hypotension (SIH) can be associated with various clinical and neuroimaging features that may impede a rapid diagnosis of this entity. The authors report the case of a patient who presented with bilateral third cranial nerve palsies and bilateral subdural hematomas. Intracraniatal pressure monitoring proved to be useful in the diagnosis and management of SIH in this patient.

The authors reported on a 44-year-old patient in whom SIH was diagnosed and dural cerebrosplinal fluid (CSF) leakages were found with the aid of CT myelography at the C1–3 epidual space (Fig. 3 in their article) and the T6–10 level. They successfully treated the patient by using an epidual blood patch and demonstrated, for the first time, the usefulness of intracraniatal pressure monitoring during the disease diagnosis and treatment of such patients. The case is well documented; however, there are some points regarding the CSF leakage at the C-2 level on which we are obliged to comment.

In an MR imaging study by Yousry et al.,1 retrospinal circumscribed fluid collections at the C1–2 levels were detected in 6 (67%) of 9 patients with spontaneous SIH headache and in 4 (36%) of 11 patients with postlumbar puncture headache. At this point, the first possibility that comes in mind regarding the pathophysiological mechanism is a possible dural leakage point at the C-2 level. However, detection of the retrospinal fluid even in patients with postlumbar puncture headache in whom the leakage point is in the lumbar region makes this theory less probable. In accordance with this, Yousry et al. suggested that the fluid collection might be due to a cascade of events beginning with the dilation of the venous plexus at the C1–2 level for compensating the decrease in CSF volume. They proposed that if the dilation of the venous plexus is restricted, or not sufficient, subsequent collection of transudate occurs. Thus agreeing with Yousry et al., we believe detection of contrast agent on CT myelography or from our personal experience on MR myelography at the C1–2 levels is most probably transudate rather than CSF leakage as in the case presented by Zada et al. In conclusion, we believe there is a possibility that retrospinal fluid collections may be detected at the C1–2 levels in patients with SIH that should not be regarded as CSF leakage unless proven otherwise. It is important to keep this possibility in mind while managing such cases.

SAIT ALBAYRAM, M.D.
MUSTAFA ONUR ULÜ, M.D.
CHAN ISLER, M.D.
Istanbul University
Istanbul, Turkey

Reference


Pituitary Adenomas


Abstract

Object. The authors devised an extended transphenoidal ap-
proach involving a submucosal posterior ethmoidectomy that allows for adequate exposure of the cavernous sinus. To evaluate the adequacy of this approach for removal of adenomas invading the cavernous sinus, the authors retrospectively analyzed the surgical outcomes obtained in treated patients.

Methods. During a 9-year period, 36 patients with pituitary adenomas extending into the cavernous sinus underwent tumor removal at Kinki University Hospital. In the authors’ technique of extended transsphenoidal surgery, the inferior wall of the affected cavernous sinus was entirely exposed, not only to permit safe removal of the tumor but also to secure the petrous portion of the internal carotid artery (ICA). For prevention of intraoperative injury to the cranial nerves, a low-profile pressure sensor was attached on the eyelid to detect eye movements in response to electrical stimulation of the cranial nerves.

Results. Total or subtotal tumor removal was achieved in 72% of 36 patients. In eight (67%) of 12 patients with growth hormone–secreting adenomas, hormonal remission was achieved postoperatively. Postoperative transient double vision was observed in 27% of the patients, but no serious complications, such as permanent cranial nerve palsy or ICA injury, occurred.

Conclusions. These reasonable surgical results obtained in the present series of patients suggest that the extended transsphenoidal approach is safe and effective for removal of adenomas within the cavernous sinus. These preliminary results may lead to a reevaluation of the role of surgery as the therapeutic strategy for invasive pituitary adenomas.

We would like to express some observations and ask the authors for some clarifications regarding several points.
1) Kitano et al. reported that to reduce intraoperative blood loss, the head and the upper part of the body were elevated at an angle of 30°; they reported 1 case of pulmonary embolism. Pulmonary embolism is a well-known complication of a sitting or semisitting operative position and especially in posterior cranial fossa surgery, but this complication has also been reported, although more rarely, in transsphenoidal surgery when the head is elevated at only 20° and when only the intercavernous sinus is opened. An opened cavernous sinus bleeds more than an intercavernous sinus, thus the risk of venous air embolism is not negligible in the described approach with the head elevated at an angle of 30°. We would like to know if Doppler ultrasonographic cardiac monitoring or, better, transoesophageal echocardiography was used during surgery and if a preoperative study for screening for eventual patent foramen ovale was conducted in the patients.
2) The authors performed the operations with the patient’s head secured in 3-point pin fixation. Rigid fixation of the head disables any usual manipulation of the patient’s head during the operation, which is performed to improve the operative view. Therefore, we would like to know whether the authors encountered some related difficulty during the approach.
3) Kitano et al. reported on 36 patients, including 4 with prolactin (PRL)-secreting tumors, who underwent an extended transsphenoidal approach to the cavernous sinus. However, in their Results the authors reported that for PRL-secreting tumors the surgical strategy was to only decompress the optic chiasm, and then the tumor remnant was treated with medical therapy. We would like to know if the described technique for the cavernous sinus was performed also in patients with PRL-secreting tumors. In our experience we have observed that in voluminous invasive prolactinomas, medical therapy was more effective after a surgical reduction of the tumoral mass, but it is always advisable not to remove the intracavernous component of the adenoma, especially if it is a prolactinoma.
4) Finally, we would like to know why the 2 patients affected by growth hormone–secreting adenomas in whom normalization of insulin-like growth factor-I was not achieved after surgery and postoperative medical treatment did not undergo radiotherapy. Nowadays, with modern radiotherapeutic techniques, like stereotactic radiotherapy or intensity-modulated radiotherapy, an optimal preservation of pituitary function is possible. In our personal experience, we did not observe any important side effect of postoperative radiotherapy, even after 6–7 years of follow-up.

Mario Francesco Fraioli, M.D.
Laura Moschettoni, M.D.
University of Rome “Tor Vergata”
Rome, Italy
Chiara Fraioli, M.D.
CIRAD Villa Benedetta
Rome, Italy

References

Response: We thank Dr. Fraioli and colleagues for their comments regarding our article. They raise several important questions, and we will summarize our comments on these issues.
1) In the Results section of our paper, we stated that “postoperative pulmonary embolism [was observed] in 1 patient” and did not make further descriptions about this complication. Dr. Fraioli et al. have interpreted this complication to mean that the air embolism was associated with intraoperative head elevation. We also regret that we did not address the relationship between the intraoperative head elevation and the risk of air embolism in our text. However, the patient who experienced pulmonary embolism had not suffered air embolism but deep venous thrombosis, which is a well-known complication of neurosurgical procedures. She became symptomatic on the 3rd postoper-
ative day, and she was successfully treated with thrombolytic therapy using urokinase.

We completely agree with Dr. Fraioli et al. that the use of previously reported monitoring techniques for a sitting operative position will reduce the risk of air embolism, although the rate of air embolism remains to be elucidated. Fortunately, we did not encounter a pulmonary air embolism as an operative complication. In our experience, tumor removal is not always associated with massive venous bleeding if the tumor completely invades into the cavernous sinus. Therefore, our approach is started at the usual position of the standard transsphenoidal approach. If massive bleeding from the cavernous sinus is encountered, the upper half of the patient’s body would be gradually elevated to diminish the bleeding during surgery. We think this intraoperative postural change is important to avoid air embolism. To make it easier and safer, the patient’s head should be secured with 3-point pin fixation.

2) For the midline transsphenoidal approach, the patient’s head is tilted left, facing toward the surgeon who is standing on the right side of the patient’s chest. The surgical field obtained by the transsphenoidal approach will be expanded laterally to expose the cavernous sinus in our extended approach. Thus, the laterally extended portion of the tumor can be removed through an oblique transsphenoidal approach, which permits direct observation of the cavernous sinus and the ICA. However, this oblique approach is uncomfortable for the surgeon, especially when the tumor extends to the right cavernous sinus and access may be hindered if the patient has poor neck flexibility. So we prefer to use rigid pin fixation of the patient’s head and to tilt the operating table downward to the lesion side during the oblique approach. This intraoperative positional change will provide more comfortable access to the right cavernous sinus because of avoidance of a looking-down posture of the surgeon with an overhanging position of the operative microscope.

3) The current strategy for managing a PRL-secreting adenoma extended into the cavernous sinus is medical therapy with or without operation. Volume reduction of the tumor might play an important role in a rapid improvement of neurological deficits. Therefore, surgical removal of the tumor is indicated in patients with progressive visual loss. However, there is some difficulty in controlling the extent of tumor resection to obtain optimal tumor resection. During the removal of invasive adenomas, the surgeon sometimes unintentionally strays off into the cavernous sinus. In these situations, there is a possibility to perform a blind dissection around the ICA, resulting in severe complications. On the other hand, the extended transsphenoidal approach makes it possible to visualize directly the cavernous sinus and the ICA under the operating microscope, which is difficult by standard transsphenoidal approach. Therefore, we believe the medial approach to the cavernous sinus, which we describe in our article, is useful for a reliable and safe removal of these invasive adenomas.

4) Microsurgery is the gold standard for treatment of growth hormone (GH)-secreting adenomas. It offers the advantages of rapid normalization of hormone hypersecretion and the chances of cure of the disease. Our results indicate that > 50% of patients with invasive GH-secreting adenomas can be treated successfully with microsurgery alone. With the development of medical therapies (somatostatin analogs and the GH receptor antagonist), medical therapies are the well-established second-line treatments for patients with acromegaly. However, even if these treatment strategies are accepted, financial considerations of medical therapy indicate that some patients with acromegaly will require radiotherapy for hormonal or tumor mass control. Therefore, we believe that radiotherapy (both conventional external and stereotactic radiation therapy) continues to have an important role in controlling acromegaly in selected patients. Although the stereotactic technique has clear advantage over external radiation in terms of precise mapping to a defined tumor volume, it is necessary that the entire residual tumor can be accurately estimated on postoperative MR images. However, the residual tumors were difficult to define on postoperative MR images in our 2 patients, who had excess GH that persisted after radical surgery of the cavernous sinus. For these patients, irradiation should be delivered to include the entire residual tumor, which was approximately estimated by the preoperative tumor images.

The primary aim of therapy for acromegaly is to reverse the symptoms and signs of the disease and achieve a long-term survival. The decision should be individualized according to the patient’s age and physiological condition, as well as the biological behavior of the tumor. Radiation therapy may induce irreversible damage of pituitary function and arterial occlusion. In addition, radiotherapy is not always effective in every case. Therefore, we believe it is better to avoid radiation therapy for the treatment of benign tumors such as pituitary adenomas. Although postoperative nadir GH levels < 2.5 and IGF-I levels < 500 in our patients did not reach the strict criteria of biochemical remission, lowering GH concentrations rapidly achieved symptomatic relief and improved physical condition. Therefore, we did not accept rushed postoperative radiotherapy in the treatment of these patients; instead we selected follow-up endocrinological assessments scheduled at 3-month intervals. (DOI: 10.3171/JNS/2008/109/8/0362a)

MASAHIKO KITANO, M.D., PH.D.
MAMORU TANEDA, M.D., PH.D.
TARO SHIMONO, M.D., PH.D.
YUZO NAKAO, M.D., PH.D.
Kinki University School of Medicine
Osaka, Japan