Acute Facial Diplegia

Marina Dididze, MD, PhD

No one involved in the planning of this CME activity have any relevant financial relationships to disclose. Authors/faculty have nothing to disclose.

CME is available 5/6/2008 - 5/6/2011

Copyright: 2008

American Association of Neuromuscular and Electrodiagnostic Medicine

2621 Superior Dr NW Rochester, MN 55901

The ideas and opinions in this Case Study are solely those of the author and do not necessarily represent those of the AANEM
CME Information  
Product: CS24 - Acute Facial Diplegia

Course Description  
Intended Audience  
This course is intended for Neurologists, Physiatrists, and others who practice neuromuscular, musculoskeletal, and electrodiagnostic medicine with the intent to improve the quality of medical care to patients with muscle and nerve disorders.

Learning Objectives  
Upon conclusion of this program, participants should be able to:  
1. evaluate patients with acute facial diplegia and formulate differential diagnosis.  
2. administer the specific tests to summarize the diagnosis.  
3. predict the role of electrodiagnostic examination to evaluate facial neuropathy.

Release Date: 5/6/2008  
Expiration Date: 5/6/2011. Your request to receive AMA PRA Category 1 Credits™ must be submitted on or before the credit expiration date.  
Duration/Completion Time: 1 hour

Accreditation and Designation Statements  
The American Association of Neuromuscular and Electrodiagnostic Medicine (AANEM) is accredited by the Accreditation Council for Continuing Medical Education to provide continuing medical education for physicians. The AANEM designates both sections of this enduring material for a combined maximum of 1 AMA PRA Category 1 credit™. Physicians should claim only the credit commensurate with the extent of their participation in the activity.

How to Obtain CME  
Once you have reviewed the materials, you can obtain CME credit by clicking on the link in the e-mail received when you purchased this product. Answer the questions and click submit. Once your answers have been submitted, you will be able to view a transcript of your CME by logging into www.aanem.org and clicking View Profile and then My CME.


**Acute Facial Diplegia: A Guillain-Barre Variant**

**May 2008**

CME Available from May 2008 through May 2011

**Accreditation Statement:** The AANEM is accredited by the Accreditation Council for Continuing Medical Education (ACCME) to sponsor continuing medical education (CME) for physicians. The AANEM designates this educational activity for a maximum of 1 *AMA PRA Category 1 Credits™*. Physicians should only claim credit commensurate with the extent of their participation in the activity. This activity has been developed in accordance with the ACCME Essentials.

**Presenting Symptom:** Acute bifacial weakness

**Case prepared by:** Marina Dididze, MD, PhD

**Affiliations:** Tbilisi State Medical University

**Disclosures:** No one involved in the planning of this CME activity have any relevant financial relationships to disclose. Authors/faculty have nothing to disclose.

**Please note:** The opinions expressed in these cases reflect the view of the authors and do not reflect official views or positions of AANEM. The AANEM is not liable for decisions made or actions taken by you or any third party in reliance on any of the information contained herein. Reference to any products, services, hypertext link to the third parties, or other information by trade name, trademark, supplier, or otherwise does not constitute or imply endorsement, sponsorship, or recommendation by the AANEM. Finally, we encourage you to read AANEM’s Privacy Policy.

**Software Requirement(s):** Each of the CME Application and Evaluation forms are online as PDF files, which can be viewed with Adobe Acrobat Reader software. Reader allows you to view, navigate, and print PDF files across all major computing platforms. To download a free copy of Reader, connect to the Adobe Acrobat site.

**Appropriate Audience:** Residents, neuromuscular fellows, and practicing physicians.

**Learning Objectives:** After completing this educational activity, participants will be able to:
1) Evaluate patients with acute facial diplegia and formulate differential diagnosis
2) Administer the specific tests to summarize the diagnosis
3) Predict the role of electrodiagnostic examination to evaluate facial neuropathy

**Level of Difficulty:** Intermediate/Advanced
Acute Facial Diplegia: A Guillain-Barre Variant

History

A 41-year-old otherwise healthy man has episode of laryngitis accompanied by intermittent diplopia and headache. A week later he experiences numbness in his left hand followed by weakness of the left facial muscles, which after three days spreads to the right facial muscles as well. The next day patient is admitted to the hospital at which time patient has moderate facial diplegia. He can not close both eyes tightly and purse lips, predominantly on the left side. Nasal labial folds are bilaterally flattened and mouth corners dropped. Tear flow is increased, but taste and hearing is normal.

Commentary I

Facial muscle weakness can be caused by central or peripheral lesions. The muscle responsible for voluntary smiling, grimacing and eyelid closure are innervated by the pathways originated in the facial motor area of precentral gyrus – Brodman’s areas 4 and 6. Axons from these cortical neurons descend as fascicles of the corticobulbar tract to the level of the lower pons, end in facial nerves nucleuses and the signals from here reach facial muscles via facial nerves. The portion of each facial nucleus responsible for the upper face muscle contraction is innervated by crossed and uncrossed corticobulbar tracts bilaterally from both the right and left precentral motor cortical areas. The lower face muscles are innervated only contra laterally. Thus the central supranuclear lesions – damage of corticobulbar tract within the cortical, extrapyramidal or brainstem areas – result in the contralateral paralysis of the lower face muscles with sparing of eyelid closure and forehead wrinkling, while peripheral lesions - damage the cranial VII nerve itself - cause paralysis of the whole face muscles. In addition to the motor supply (to the muscle of the face and also stapedius muscle), facial nerve involves (via nervus intermedius) parasympathetic fibers to the lacrimal gland, the submaxillary and sublingual salivary glands and sensory fibers from the taste receptors on the anterior two thirds of the tongue (chorda tympani fibers).

In our case the patient presents with bilateral weakness of upper and lower face muscles due to the both facial nerve palsies. Sparing of taste means that the lesion is above the point where the chorda tympani fibers join the facial nerve. Also preceding upper respiratory tract infection with headache and one episode of diplopia should be considered for the differential diagnosis. Bilateral facial palsy is most often a special finding of a systemic disease. The differential diagnosis in terms of etiology includes: idiopathic Bell’s palsy, inflammatory/autoimmune disorders, neoplasms, infections/viral, metabolic, trauma, congenital/inherited disorders; benign intracranial hypertension.

Bell’s palsy or acute idiopathic facial paralysis is the most common cause of facial neuropathy. Incidence rate of it is 23 per 100,000 annually according to Hauser at al. This disorder occurs at all ages and all times of the year and affects men and women more or less equally. It is more common in patients with diabetes and hypertension. Although the cause remains unknown, recent evidence suggests a possible association with Herpes simplex virus (HSV) infection.

Inflammation/autoimmune:

Facial diplegia can be the manifestation of GBS. Areflexia helps to distinguish GBS as the underlying etiology. Further investigations have to be done to establish the diagnosis in our case.

Approximately 10 % of patients with systemic lupus erithematosis (SLE) exhibit peripheral nerve involvement. Usually neuropathy appears in more advanced stages, but rarely could be the initial presentation. Sensory loss dominates and neuropathy has progressive and relapsing course. Which is not mentioned in our case – additional history is required.

Melkersson-Rosenthal syndrome (MRS) is a rare idiopathic non-caseating granulomatous condition. Facial palsy, lip-swelling and lingua plicata are its most common presenting features. The syndrome begins in childhood or adolescence and may be familiar. Our patient doesn’t have other features.
characterized for MRS and also the age of onset is not typical.

**Neoplasms:**
The facial nerve is the most common cranial nerve involved by meningeal (leukemia, lymphoma) tumors, but bilateral involvement is rare. Also other tumors should be considered: extra-axial – epidermoid cancer, ependimoma and cholesteatoma; acoustic neuromas, neurofibromas, tumors of parotid gland or granulomatosis at the base of the brain (histiocytosis). The onset is insidious and course progressive. Amyloidosis associated with crystal lattice deposits in the cornea, may involve the facial nerves. Pontine lesions, vascular or neoplastic may cause facial palsy, usually in conjunction with other neurological signs. The diagnosis is based on the clinical signs and MRI findings.

**Infections/viral:**
Syphilis was considered the sole cause of bilateral facial palsy almost a century ago; in 40s of last century seventh and eight nerves were found to be the most frequently involved cranial nerves in syphilitic meningitis.

Currently spirochetal facial weakness is more likely to be the result of Lyme disease. It was named after the town Lyme in Connecticut, where a cluster of cases was first recognizes in 1975. 7 years later Burgdorfer and colleagues identified the causative spirochetal agent, Borrelia burgdoferi, transmitted by ticks. Early skin manifestations of the disease had previously described in Europe as erythema chronicum migrans. Later manifestations with acute radicular pain followed by meningitis and frequently accompanied by peripheral and cranial nerve involvement, had long been known in Europe as the Bannwarth or Garin-Bujadoux syndrome. The identity of these diseases has been established and the entire group now is classed as borreliosis. Lyme disease is less acute than leptospirosis and less chronic than syphilis. It involves the skin, the nervous system, the heart and articular structures over a period of a year. In about half of the cases cranial nerve involvement becomes prominent within weeks of onset of the illness. In endemic areas perhaps one in four cases of ‘idiopathic facial palsy’ is caused by borreliosis (1). The diagnosis is likely when there has been a tick bite with erithema marginatum or arthritis.

Human immunodeficiency virus (HIV) is an infrequent cause of facial palsy. The facial palsy of Lyme and HIV are associated with a pleocytosis; serologic and SCF examination may be useful if there is s suspicious of either process.

Tuberculous infection of the mastoid and the middle ear or of the petrous bone is a cause of facial paralysis in parts of the world where tuberculosis is particularly common. Facial palsy may occur in infectious mononucleosis and also occasionally in poliomyelitis.

Facial nerve is frequently involved in leprosy (Hansen's disease), which is the classic example of an infectious neuritis, being due to the direct invasion of nerves by Mycobacterium leprae, locally causing the hypopigmented, lacking in sensation skin macule or papule. In addition to facial nerve involvement other peripheral nerves are also damaged with sensorimotor deficit and progressive cutaneous anesthesia. Tendon reflexes are usually preserved probably due to sparing of the muscular and larger sensory nerves.

Herpes virus commonly attacks the seventh nerve. Incidence of herpes zoster rises with age. If a cohort of 1000 people lived to 85 years of age, half of them would have had one attack of zoster and 10 would have had two attacks. Herpes zoster 'loves' the geniculate ganglion causing Ramsay Hunt syndrome, consisting of mostly painful facial palsy associated with a vesicular eruption in the external auditory canal, other parts of the cranial integument and mucous membrane of the oropharynx. Often eighth nerve is involved as well causing hearing loss and, vertigo. Varicella zoster viral infection can be detected in Ramsay Hunt syndrome before the emergence of the vesicles in a few hours by collecting exudates from the skin of the pinna and using PCR technique.

Facial diplegia occurs in approximately seven of every thousand patients with sarcoidosis (Uveo-parotid fever or Heerfordt Syndrome). The infectious etiology of sarcoidosis has never been established, but clinically and pathologically the disease is close to tuberculosis and other granulomatous infections. The
essential lesion consists of focal collections of epitheloid cells surrounded by a rim of lymphocytes. The sarcoid tubercules may be found in all organs and tissues, but most frequently in mediastinal and peripheral lymph nodes, lungs, liver, skin, eyes, phalangeal bones, parotid glands. While only 5% of sarcoidosis has neurological manifestation, half of these have facial nerve involvement and one-six have facial diplegia, where the paralysis on each side tends to be separated by weeks or longer.

**Metabolic:**
Mononeuritis multiplex rarely involves both facial nerves simultaneously and is frequently combined with diabetes.

**Congenital defects:**
Among congenital abnormalities Mobius Syndrome should be considered, due to in utero damage or maldevelopment of dorsal pontine tegmental structures. In this autosomal dominant disorder facial diplegia is accompanied by one of the following signs present at birth: convergent strabismus, complete external ophthalmoplegia, lingual palsy, clubfeet, mental defects, brachial disorders.

Bone overgrowth of the skull from any cause can invade cranial nerve foramina, but sclerosteosis seems particularly prone to affect the facial nerve bilaterally. This is a rare hereditary condition mostly affecting the families in South Africa, but some cases have been reported in Europe and USA.

Severe or complete facial palsy occurs in facioscapulohumeral dystrophy, sometimes preceding weakness of the shoulder girdle. Varying degrees of bifacial weakness are observed in myasthenia gravis, usually conjoined with ptosis and ocular palsies, also could be combined with the weakness of other bulbar muscles and masseters. Bifacial weakness is also a feature of congenital myopathies (carnitine, nemaline deficiency), but in all these cases the patient has other clinical manifestations and none of these disorders develop facial weakness acutely.

**Trauma:**
Traumatic palsies of facial nerve appear to be relatively frequent. Of cranial nerve injuries in adults, seventh nerve is second in frequency to the olfactory nerve, but involvement of both facial nerves is rare. Fractures of the temporal bone (usually with damage to the middle or internal ear), otitis media and middle ear surgery are relatively uncommon causes.

**Toxin exposure:**
Ingested toxins would seem less likely to produce cranial neuropathy, but ethylene glycol may result in facial diplegia.

Facial paralysis is a rare concomitant of benign intracranial hypertension, where generalized or asymmetrical dull headache is the cardinal symptom. Also blurred vision, dizziness, diplopia or numbness of one side of the face may occur. This condition is more common in young women and frequently is concomitant to obesity. The diagnosis is based on findings of papilledema, elevated CSF pressure, signs of increased intracranial pressure on CT or MRI examinations.

Additional aspects of the history will be helpful in differentiating between the various diseases mentioned above: systemic symptoms – fever, cough, fatigue, sweating, dry mouth, arthritis; similar episodes in anamnesis; headache; family history; trauma; sexual history; contacts with infections; travel/camping; toxin exposure; bad habits – smoking/drugs/alcohol.

**History, Continued**
Patient does not have any episode of motor/sensory disturbance and headache in the past. He develops painless facial diplegia preceded with the episode of sore throat that lasted one day, headache and diplopia that both resolved in couple of minutes, although to exclude benign intracranial hypertension further investigations should be performed.
There is no history of trauma and no family history of neurologic illness. He did not have parotid/mastoid surgery. He doesn’t report any vesicular eruptions at the face or in the external auditory canal at both sides. Patient denied systemic symptoms, infectious contacts, drug use; he is non smoker and had no known toxic exposures. He is married for 15 years, has two children and was sexually active. He denies any occasional sexual contacts with other partners and also denies any sexually transmitted infections in his history. He can not recall any recent bites with/or local papulla or erithema. He lives in the capital and has not traveled recently anywhere.

Commentary II

Absence of family history of inherited diseases, also presenting clinical signs and acute course of the disease exclude Mobius syndrome, faciohuleroscapular dystrophy and congenital myopathies; although sclerosterosis needs further radiographic investigation of the skull. Patient does not report any similar episodes in the past; although he has numbness in the left hand, motor disturbance is more prominent sign; all these make SLE less likely.

No trauma was mentioned and no toxin/drugs exposure. Absence of occasional sexual contacts for the last years makes syphilis and HIV less likely, although both need serology to be done.

Absence of any systemic involvement (fever, arthritis and other) and also no bites and/or local papulla or erithema on the skin makes less likely to have tuberculosis, leprosy, Lyme disease and sarcoidosis, although physical examination should be performed.

In addition to facial diplegia the presenting signs (laryngitis, diplopia and numbness in the left hand) should be considered for further differential diagnosis:

1. Bell’s palsy
2. GBS
3. Neoplasms
4. Ramsay Hunt syndrome
5. Lyme disease
6. Leprosy
7. Diabetes mellitus
8. Infectious mononucleosis
9. Sarcoidosis
10. HIV
11. Sclerosteosis
12. Mononeuritis multiplex
13. Benign intracranial hypertension

Physical examination should be focused on detailed evaluation of cranial nerves, assessment of motor and sensory systems as well as examination of the chest and abdomen.

Physical Examination

Patient is afebrile and fully conscious. Cranial nerve examination reveals bilateral facial weakness, other cranial nerves remained intact. Taste, hearing - normal. Fundoscopy does not show abnormalities. Motor system examination reveals normal strength and muscle tone in all limbs, diminished reflexes in the upper limbs – biceps, triceps and radial-periosteal and absent patellar and Achilles. Babinski sign is absent. Abdominal superficial cutaneous reflexes are preserved. Sensory system testing reveals
diminished light touch and pinprick sensation in the left palm at the area of median nerve distribution; proprioception and vibration are preserved. Coordination testing shows normal finger-to-nose and heel-to-knee movements. Romberg’s sign absent. Gate normal. Meningeal signs absent. Patient does not have any urinary/bladder dysfunction. Skin and visible mucosa with no specific changes, accessible lymph nodes by palpation do not reveal abnormalities; the parotid glands are not enlarged. Auscultation of the heart and lungs is normal; the abdomen is soft and nontender, without organomegaly.

**Commentary III**

The lack of CNS findings makes central (pontine) tumors less likely. Extrapontine tumors need further investigation. No other cranial nerve involvement excludes mononeuritis multiplex. A few sensory deficits indicate on absence of leprosy and makes less likely diabetic neuropathy, although glucose tests should be done. Normal lymph nodes and no signs of organomegaly almost exclude infectious mononucleosis, but preceding episode of laryngitis makes necessary to examine peripheral blood.

The differential diagnosis at this stage includes:

1. Bell's palsy
2. GBS
3. Neoplasms (extrapontine)
4. Ramsay Hunt syndrome
5. Lyme disease
6. Diabetes mellitus
7. Infectious mononucleosis
8. Sarcoidosis
9. HIV
10. Sclerosteosis
11. Benign intracranial hypertension

Further investigations should be focused on laboratory, chest X-Ray, neuroimaging studies and electrophysiologic data.

Laboratory findings and radiologic examination: peripheral blood smear is normal; Audiometry, skull and chest radiography show no abnormalities; syphilis tests and HIV antibody tests are negative. Lyme serology is negative. Blood glucose is normal. MRI of the head is nondiagnostic.

**Commentary IV**

Laboratory data narrowed our differential diagnosis to the following:

1. Bell’s palsy
2. GBS
3. Ramsay Hunt syndrome
4. Benign intracranial hypertension

Electrophysiologic studies and CSF examination should be performed.

**Electrophysiologic Data**

<table>
<thead>
<tr>
<th>NERVE</th>
<th>SIDE</th>
<th>STIM SITE</th>
<th>RECORD</th>
<th>CM</th>
<th>AMPL</th>
<th>LAT</th>
<th>CV</th>
</tr>
</thead>
<tbody>
<tr>
<td>Median</td>
<td>R</td>
<td>Wrist</td>
<td>Digit II</td>
<td>16</td>
<td>10.2(&gt;10)</td>
<td>2.9(&lt;3.5)</td>
<td>55.2(&gt;50)</td>
</tr>
<tr>
<td>NERVE</td>
<td>SIDE</td>
<td>STIM SITE</td>
<td>RECORD</td>
<td>CM</td>
<td>AMPL</td>
<td>LAT</td>
<td>CV</td>
</tr>
<tr>
<td>-------------------</td>
<td>------</td>
<td>-----------------</td>
<td>--------</td>
<td>----</td>
<td>----------</td>
<td>-------</td>
<td>--------</td>
</tr>
<tr>
<td>Facial</td>
<td>R</td>
<td>Anterior mastoid</td>
<td>Orbicularis oris Nasalis</td>
<td>11</td>
<td>1.7 (&gt;1)</td>
<td>4.5 (&lt;3.1)</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>2.2 (&gt;1)</td>
<td>5.0 (&lt;4.2)</td>
<td></td>
</tr>
<tr>
<td>Facial</td>
<td>L</td>
<td>Anterior mastoid</td>
<td>Orbicularis oris Nasalis</td>
<td>11</td>
<td>1.3</td>
<td>5.1</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>1.8</td>
<td>5.4</td>
<td></td>
</tr>
<tr>
<td>Median</td>
<td>R</td>
<td>Wrist</td>
<td>APB</td>
<td>8</td>
<td>8.5(&gt;6)</td>
<td>2.9(&lt;4.2)</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Elbow</td>
<td></td>
<td></td>
<td>7.7</td>
<td>7.2</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>8</td>
<td></td>
<td>25</td>
<td>58.1(&gt;50)</td>
<td></td>
</tr>
<tr>
<td>Median</td>
<td>L</td>
<td>Wrist</td>
<td>APB</td>
<td>8</td>
<td>7.9</td>
<td>4.6</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Elbow</td>
<td></td>
<td></td>
<td>8.0</td>
<td>8.9</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>8</td>
<td></td>
<td>24</td>
<td>55.8</td>
<td></td>
</tr>
<tr>
<td>Ulnar</td>
<td>R</td>
<td>Wrist</td>
<td>ADM</td>
<td>8</td>
<td>8.5(&gt;5.5)</td>
<td>3.2(&lt;3.4)</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>B.Elbow</td>
<td></td>
<td></td>
<td>7.8</td>
<td>7.2</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>A.Elbow</td>
<td>8</td>
<td></td>
<td>22</td>
<td>9.8</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>8</td>
<td></td>
<td>14</td>
<td>55.0(&gt;50)</td>
<td></td>
</tr>
<tr>
<td>Ulnar</td>
<td>L</td>
<td>Wrist</td>
<td>ADM</td>
<td>8</td>
<td>9.0</td>
<td>3.1</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>B.Elbow</td>
<td></td>
<td></td>
<td>8.2</td>
<td>7.0</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>A.Elbow</td>
<td>8</td>
<td></td>
<td>21</td>
<td>9.6</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>8</td>
<td></td>
<td>14</td>
<td>51.2</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>8</td>
<td></td>
<td>21</td>
<td>53.8</td>
<td></td>
</tr>
<tr>
<td>Tibial</td>
<td>R</td>
<td>Ankle</td>
<td>AHB</td>
<td>9</td>
<td>4.6(&gt;4)</td>
<td>6.8(&lt;6)</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Popliteal fossa</td>
<td></td>
<td></td>
<td>4.2</td>
<td>14.0</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>9</td>
<td></td>
<td>40</td>
<td>55.5(&gt;40)</td>
<td></td>
</tr>
<tr>
<td>Tibial</td>
<td>L</td>
<td>Ankle</td>
<td>AHB</td>
<td>9</td>
<td>4.7</td>
<td>7</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Popliteal fossa</td>
<td></td>
<td></td>
<td>4.3</td>
<td>14.4</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>9</td>
<td></td>
<td>39</td>
<td>52.7</td>
<td></td>
</tr>
<tr>
<td>Common peroneal</td>
<td>R</td>
<td>Ankle</td>
<td>EDB</td>
<td>9</td>
<td>3.6(&gt;2)</td>
<td>3.8(&lt;5.5)</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Fibular head</td>
<td></td>
<td></td>
<td>2.7</td>
<td>11.0</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Lateral knee</td>
<td>9</td>
<td></td>
<td>35</td>
<td>48.6(&gt;40)</td>
<td></td>
</tr>
<tr>
<td>Common peroneal</td>
<td>R</td>
<td>Ankle</td>
<td>EDB</td>
<td>9</td>
<td>3.4</td>
<td>3.6</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Fibular head</td>
<td></td>
<td></td>
<td>2.5</td>
<td>10.8</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Lateral knee</td>
<td>9</td>
<td></td>
<td>34</td>
<td>47.2</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>9</td>
<td></td>
<td>2.2</td>
<td>12.9</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>9</td>
<td></td>
<td>2.2</td>
<td>42.8</td>
<td></td>
</tr>
</tbody>
</table>
Electrophysiological data reveal prolonged distal latencies in left median nerve motor and sensory fibers and also in both tibial nerves motor fibers. Nerve conduction velocities and amplitudes are normal in ulnar, tibial and peroneal nerves motor and sensory fibers. In both facial nerves latencies are significantly prolonged, but amplitude is not decreased.

Blink reflexes are not evoked from both sides. Needle EMG is not performed.

Electrophysiologic findings indicate on the left distal median sensory-motor neuropathy, both distal tibial motor neuropathy and bilateral facial neuropathy (with severe demyelination and no axonopathy).

**Diagnostic Impression**

Lumbar puncture is performed and CSF analysis shows normal pressure, 3 lymphocytes per mm3 with markedly elevated protein concentration 127 mg/dl and normal glucose level. Immunoassays of serum of CSF for herpes simplex virus, Varicella zoster virus, Epstein-Barr virus, cytomegalovirus shows no abnormalities; Antiganglioside serology and serum antibody titer to C.jejuni are not performed.

Electrophysiologic data, absence of papilledema, no sings of intracranial hypertension on MRI and normal CSF pressure eliminate benign intracranial hypertension. Absence of pain preceding or concomitant to the facial diplegia, no signs of vesicular rush involving the ear and face and normal antibody levels for herpes viruses, also involvement of other peripheral nerves in addition to both facial nerves makes less likely Ramsay Hunt syndrome and Bell's palsy.

The patient is treated by plasma exchange (50 ml/kg) for 2 weeks. Facial diplegia gradually lessened from the 4-6th day of starting treatment. Clinical examination at the day 45 shows significant improvement at the left side of the facial muscles – patient has only mild weakness of the upper face, and complete recovery at the right face; no sensory loss in the left hand, appearance (diminished but present) of the reflexes in the lower limbs and normal reflexes in the upper limbs. Repeat nerve conduction studies revealed mild demyelination in the left facial nerve's upper branches. Facial diplegia was completely resolved by day 65. A follow-up study 6 month later shows no evidence of recurrence.

**Commentary V**

A diagnosis of the Guillain-Barre syndrome (GBS) variant acute facial diplegia is made. Although unilateral facial paralysis is relatively common with an incidence of 20 to 25 per 100,000 people, bilateral facial palsy occurs in 0.3% to 2.0% of the facial palsy cases.(1) Although Bells’ palsy is one of the most common causes of unilateral facial impairment, it also may be bilateral, but rarely is the involvement on the two sides simultaneous.(2) The sensory deficit in the left hand, prolonged distal latencies in left median and both tibial nerves and diminished/absent brisk reflexes indicate to other possible cause in our case. CSF evaluation proves the diagnosis. Sparing of taste and absence of blink reflexes indicate on the more proximal involvement of facial nerves.

The acute autoimmune inflammatory polyneuropathy -Guillain-Barre syndrome affects children and adults of both sexes in all parts of the world. It is know as nonseasonal and nonepidemic. The reported incidence rate worldwide has varied from 0.4 to 1.7 cases per 100,000 people per year. A mild respiratory or gastrointestinal infection precedes the neurologic manifestation by 1-3 weeks in most patients. Recently the enteric organisms Campylobacter jejuni was described as one of the most frequent preceding infection, although other bacterial and viral infections should also be considered (Lyme disease, Mycoplasma pneumoniae, influenza, Epstein-Barr virus, HIV, cytomegalovirus). Pathogenesis is based on the cell mediated immunologic reaction against peripheral nerves. The initiation cause of the immune reaction is not known. There is the evidence of sencitized T-cells to myelin but antmyelin
antibodies play a major role in initiating the disease. A number of antiganglioside antibodies are detected inconsistently in patients with GBS. Anti-GT1a IgG antibodies with or without GQ1b reactivity have been found in patients with GBS, however anti-GQ1b IgG antibody with anti-GT1a reactivity has been detected in patients who have ataxic GBS or Fisher syndrome.\(^{(3)}\)

The major clinical manifestation of typical pattern of GBS is weakness, which in most patients ascends from legs to trunk, arms and cranial musculature (Landry’s ascending paralysis). The weakness can progress to total motor paralysis and death from respiratory failure within a few days. Sensory loss is variable and mostly occurs in first days. Diminished or absent tendon reflexes are common, although hyperreflexia was also reported in several cases.\(^{(4)}\) Various cranial nerve involvements occur in half of the cases, but usually come later after limbs are affected. Isolated facial diplegia without prominent signs of GBS is considered as rare variant of this disease. In this uncommon manifestation of GBS after exclusion of other underlying causes, described above, the diagnosis is made by electrodiagnostic studies and CSF examination. Albuminocytologic dissociation usually occurs after the first few days of symptoms and is probably a reflection of a widespread inflammatory disease of nerve roots. Nerve conduction studies are considered as early diagnostic indicator and reveal various degrees of demyelination and/or axonopathy. In some cases electrophysiologic evaluations do not correlate with the facial weakness and the recovery rates, where blink reflex tests are helpful to predict the result of facial palsy.\(^{(5)}\)

Absence of axonopathy is good prognostic feature in our case. In acute cases of bifacial weakness identifying GBS is very important, because these patients may benefit from appropriate treatment.

**Bibliography**