Facial Palsy in Cerebral Venous Thrombosis
Transcranial Stimulation and Pathophysiological Considerations

J. Straub, MD; M.R. Magistris, MD; J. Delavelle, MD; T. Landis, MD

Background—Cranial nerve palsy in cerebral sinovenous thrombosis (CVT) is rare, its pathophysiology remains unclear, and data from electrophysiological examinations in such patients are missing.

Case Description—We report the case of a 17-year-old woman with familial protein S deficiency who was admitted with extensive multiple CVT. Two weeks after onset of symptoms, she developed isolated right peripheral facial palsy, and MR venography showed segmental occlusion of the ipsilateral transverse sinus. Complete recovery of facial palsy occurred concomitant with recanalization of the transverse sinus. Facial neurography, including transcranial magnetic stimulation of the facial nerve and related motor cortex, ruled out a coincidental idiopathic palsy and revealed conduction block proximal to the facial canal.

Conclusions—Facial palsy in our patient was caused by transient neurapraxia in the intracranial segment of the nerve. We suggest that elevated venous transmural pressure in the nerve’s satellite vein, which belongs to the affected drainage territory of the transverse sinus, might have caused venous blood-brain barrier dysfunction in the intrinsic vascular system of the nerve, with leakage of fluids and ions into the endoneurial space and thus an increase in interstitial resistance. ([Stroke. 2000;31:1766-1769.])

Key Words: cerebral thrombosis ■ cranial nerves ■ neural conduction ■ protein S deficiency ■ transcranial magnetic stimulation

The clinical picture of cerebral venous thrombosis (CVT) is variable, and a wide spectrum of neurological symptoms has been described.1–5 Cranial nerve palsy in CVT is rare and is often thought to result from cavernous sinus thrombosis or elevated intracranial pressure.2,4–8 In the early literature, several cranial nerve syndromes in CVT have been identified and attributed to extension of thrombosis into contiguous venous tributaries,2 presumably leading to direct pressure palsy of the nerves lying in proximity to the clot. It is only recently that Kuehnen et al9 have drawn new attention to cranial nerve syndromes in thrombosis of the transverse and sigmoid sinus and suggested that local stasis in the cranial nerve veins draining into the transverse sinus might cause temporary nerve dysfunction.9 This case report of isolated facial palsy in CVT related to protein S deficiency refines their hypothesis, as electrophysiological examination has permitted more insight into the pathogenesis of facial palsy in CVT.

Case Report
We describe a 17-year-old woman with familial protein S deficiency who began taking oral contraceptives in August 1997. Four months later, on December 25, she developed a severe headache, with photophobia and phonophobia, nausea, and neck stiffness, and was hospitalized 2 days later. Cerebrospinal fluid analysis was normal, but the initial pressure was not measured. After transient improvement, the headache worsened despite analgesic treatment, episodes of vomiting occurred, and bilateral papilledema was noted. Cerebral MRI showed extensive thrombosis in the territory of the left internal and proximal right internal cerebral veins, the vein of Galen, and the straight, superior longitudinal, left transverse, and sigmoid sinuses. High-dose intravenous heparin and coumarin were initiated, and oral contraceptives were discontinued. The patient was transferred to our hospital on January 6, with intense nuchal pain and rigidity, papilledema with only minimal reduction of visual acuity, and hyperreflexia. On January 8 she developed right peripheral facial palsy with lagophthalmus and dry eye without keratitis, diminished taste, and hyperacusis of the right ear. The palsy was mild and more marked on the upper part of the face (frontalis and orbicularis oculi muscles). Bedside otorhinolaryngological examination ruled out otitis and did not reveal eighth nerve palsy; audiometry was not performed. Repeat MRI showed persistence of thrombosis in the above-mentioned sinuses, particularly segmental occlusion of the right transverse sinus (Figure 1). There was no contrast enhancement of the facial nerve and no temporal bone abnormality (Figure 2). On day 4 after palsy onset, electrophysiological examination was performed on a Viking IV electromyography apparatus.
(Nicolet), with recordings from mentalis and nasalis muscles using surface electrodes. 10,11 For both muscles, supramaximal electrical stimulation (ElStim) was performed in the fossa stylomastoidea, and magnetic stimulation with a MagStim 200 stimulator (Magstim Company Ltd) was applied to the ipsilateral mastoidal region (MagStim), permitting excitation of the facial nerve at its entrance into the acoustico-facial meatus. 12 For the nasalis muscle, stimulation was also applied to the face-associated motor cortex (CxStim). The ElStim and MagStim evoked responses were within normal range on both mentalis and nasalis muscles, whereas the CxStim evoked response (from nasalis muscle) was markedly reduced in amplitude compared with the unaffected side (Figure 3). These findings are suggestive of a partial conduction block of the facial nerve, proximal to its entrance into the facial canal; normal response to MagStim speaks against a coincidental idiopathic facial palsy or pressure palsy within the canal. 10,11 The blink reflex R1 and R2 responses (recorded from orbicularis oculi muscles with stimuli applied to the supraorbital nerve) showed normal latencies but were reduced in amplitude on the palsy side.

Figure 1. Sagittal (A) and coronal (B) cerebral MR venography showing thrombosis in the territory of the left internal and proximal right internal cerebral veins, the vein of Galen (A), and straight and superior longitudinal sinuses, as well as the left transverse (B) and the junction of right transverse and sigmoid sinuses (B arrow).

Figure 2. Axial T1-weighted gadolinium MRI shows no abnormal contrast enhancement of the facial nerve in its intracranial segment (above) or in the fallopian canal (below).

Figure 3. Electrophysiological data. Recordings from nasalis muscle, on the palsy side (RIGHT) and control healthy side (LEFT). Electrical stimuli are applied at the fossa stylomastoidae (ElStim); magnetic transcranial stimuli are on the mastoid (MagStim) and over the contralateral motor cortex (CxStim). On day 4 (from facial palsy onset), latencies are normal; amplitudes are within normal range (although slightly reduced on the palsy side); MagStim evokes a normal response, thus discarding a coincidental idiopathic facial palsy or pressure palsy within the canal; CxStim-to-ElStim amplitude ratio is decreased on the palsy side (14%) compared with the healthy side (43%) suggesting proximal conduction block. On day 18, palsy has subsided and ElStim and MagStim are unchanged, whereas latency to CxStim is delayed and responses seem dispersed; CxStim-to-ElStim amplitude ratio is still decreased (18%).
Figure 4. Enlargement of arrow in Figure 1B showing transmural thrombosis at the junction of right transverse and sigmoid sinus (above) and control MR venography showing its recanalization (below).

Under anticoagulant and analgesic treatment the patient recovered slowly, and MR venography on January 15 showed partial recanalization of the right transverse and most of the other sinuses (Figure 4). The patient was discharged the next day and had completely recovered from facial palsy 10 days later. Because the first electrophysiological examination took place early and therefore could not completely rule out the subsequent occurrence of wallerian degeneration, a second recording was performed. On day 18, when facial palsy had subsided, the ElStim and MagStim evoked responses remained unchanged. The CxStim response was delayed and dispersed; amplitude was still reduced.

Discussion
In our patient, the chronology of events documented by repeat MRI (Figure 4), which showed occlusion and later recanalization of the right transverse sinus concomitant with onset and recovery from facial palsy, led us to believe that the palsy was a direct consequence of the right transverse sinus occlusion. CT of the petrous bones was normal, and MRI showed no abnormal gadolinium enhancement of the nerve, which may be seen in facial palsy of other origin.13 Patients with phenotypic protein S deficiency may develop multiple thromboembolic complications, including CVT14,15; however, there is no mention of increased incidence of nerve palsies in such patients.

Although involvement of all cranial nerves has been described in CVT,2,3,5,9,16 facial palsy in CVT is very rare and little is known about its pathophysiology. The cranial nerve syndromes identified in the earlier literature have been thought to be caused by pressure palsy, either directly by the thrombotic clot lying in proximity to the nerves5 or by raised intracranial pressure.7 Another explanation for cranial nerve palsy in CVT was given by Kuehn et al.,9 who reported 5 cases of cranial nerve palsy as the only feature of isolated transverse sinus thrombosis. The authors hypothesized that the palsy was due to reversible compromised oxygen or glucose consumption caused by impaired drainage of the cranial nerve veins into the transverse sinus, but no electrophysiologic examination was done to support this hypothesis. In our patient, ElStim and MagStim performed on day 4 after facial palsy onset showed normal responses, whereas both CxStim and blink reflex disclosed markedly decreased amplitudes on the affected side. This indicated a normal conduction within the facial canal and thus discarded a coincidental idiopathic facial palsy10,11 or a pressure palsy within the canal, as might have been suspected from the earlier literature5 and knowledge about the ultrastructure of the nerve.17–19 Clinically, the rapid recovery of the nerve function was suggestive of a palsy due to neurapraxia; diminished taste, hyperacusis, and dry eye indicated a supranuclear blocking. This was confirmed by the electrophysiological findings, consistent with a conduction block of the nerve proximal to the site of excitation that is proximal to its intracanalicular segment.12

In our patient, transmural thrombosis of the distal transverse sinus, which involves the drainage site of the superior petrosal sinus and thus affects the drainage territory of the lateral pontine vein (via the superior petrosal vein),20–22 could have raised the intraluminal pressure in the nerve’s satellite vein. Impaired venous drainage of the nerve root’s satellite vein is transmitted to the intrinsic vascular system of the nerve, which consists of segmentally arranged vessels, variable in number and size, that connect with the larger adjacent vessels of the extrinsic system. The postcapillary collecting venules that run in the intraneural septa of the nerve have a lumen lined with endothelial cells connected with tight junctions which represent the venous blood-brain barrier.18,20,22 A rise in venous intraluminal pressure in the extrinsic system can increase the venous transmural pressure in these intraneurial venules and lead to venous blood-brain barrier dysfunction, with leakage of fluids and ions into the endoneurial space.22 Thus, an impairment of the saltatory current flow, with reversible slowing of conduction or even conduction block,23 could account for the neurapraxia proximal to the facial canal observed in our patient. In the fallopian canal, the drainage of the nerve is assured by a widely connected venous network that communicates with the blood vessels of the entire temporal bone. Main vessels empty into the lateral, superior, or inferior petrosal sinuses, the pterygoid plexus of veins, and the middle meningeal veins17,24 and thus provided efficient drainage of the intracanalicular nerve portion. Because the satellite veins of the more caudal cranial nerves drain via the inferior petrosal vein and petrosal sinus distal to the occlusion site into the sigmoid
sinus or directly into the jugular vein, they also remained unaffected. We suggest that some of the cranial nerve syndromes reported earlier might have been a consequence of similar electrophysiological changes rather than of direct compression or reversible compromised oxygen and glucose consumption.

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References
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