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Optic Neuritis with Residual Tunnel Vision in Perchloroethylene ToxicityMarco Onofri^a, Astrid Thomas^a, Cristina Paci^a,
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Trichloroethylene and perchloroethylene (PCE) are the major constituents of commercially available solutions used in dry cleaning shops. We describe the appearance of severe bilateral optic neuritis (ON) with unexpectedly high levels of PCE metabolites in blood and chloroform in urine in a 57-year-old woman owner of a small dry cleaning shop where she worked for about 20 years using PCE. Environmental concentrations collected for 1 h 2 times a day (morning and afternoon) biannually had always been inside the limits (25–50 ppm, 170–339 mg/m³) allowed by current laws [1]. One day before the development of symptoms she had spent more than 9 h ironing clothes and fabrics immediately after withdrawal from the dry cleaning machine. In a control performed 2 weeks after acute toxicity, in the breathing zone where the ironing table was located, air samples collected for 30 min on charcoal and analyzed by desorption with carbon disulfide gas chromatography, showed peaks of 439 mg/m³ (64 ppm) near the basket containing freshly dry-cleaned fabrics, and a concentration in steam rising to 1,716 mg/m³ (252 ppm) during ironing. The onset of symptoms was apparently abrupt. Following one night of malaise, she woke up with severe headache. She was totally blind except for residual light perception. She complained of intense retroorbital pain, and eye movements induced the appearance of bright, luminous phosphenes and increment of retroorbital pain. Pupils were in mid-position, light reflex was slow but bilaterally present. A partial right third nerve palsy was noticed for 2 days, but completely disappeared on the 5th day. At the fundus observation the eye ground appeared normal, there were no signs of edema, ischemia or inflammation, intraocular pressure was 16 mm Hg. A first magnetic resonance imaging (MRI) of the head and orbits, with and without gadolinium, was performed on day 2, and was normal. A second MRI performed with long time echo short tau inversion recovery (LTE-STIR) imaging of orbits and chiasmal portion of optic nerves was performed on day 10, showing increment of intraorbital signals of both optic nerves [2]. T2 weighted axial images obtained at the same time were normal. A third MRI with gadolinium was performed on day 75, and was also normal (fig. 1). Three weeks after the onset of ON she could see only the central 2–3° radius of the visual fields, as tested with Humphrey Field Analyzer, exploring with 80 points the central 10°, 15° and 30° of visual fields (programs 10.1, 10.5), with confrontation campimetry, with tangent screen perimetry. The size of the residual visual fields did not change in the 1-year

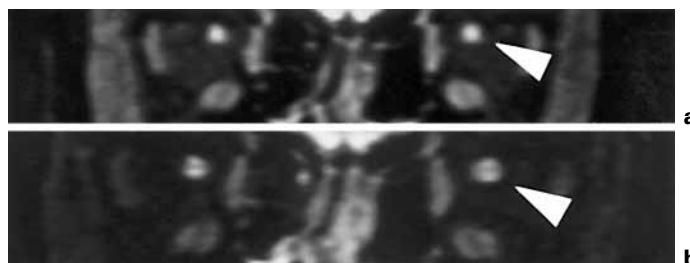


Fig. 1. **a** MRI performed 10 days after acute onset of bilateral optic neuritis. Coronal LTE-STIR image shows increased signal intensity bilaterally, with disappearance of the dural ring (arrowhead). **b** STIR MRI performed 70 days later, notice the hypointense nerve with a normal dural ring (arrowhead).

follow-up (fig. 2). Ophthalmological examination was repeated 8 weeks after the onset of ON, only retinal fluorangiography evidenced a slight papillary hypofluorescence in both eyes. Visual acuity was 8/10 in the right eye, 9/10 in the left eye. Color perception, tested with Ishihara plates and Fansworth 100 Hue Test, was abnormal in the red-green axis for both eyes. Electroretinograms (ERGs) with stimuli of 17–34 cd/m² (5–10 fL), pattern ERGs and visual evoked potentials (VEPs) to checkerboard patterns stimuli of 30', 15' and 7.5' (min of arc) were recorded, 7, 15, 28 days, 2, 5, 12 months after ON onset. Flash ERGs were normal in all recording sessions. PERGs amplitudes were normal until the 5th month follow-up, when a 22% amplitude decrement in the right eye and 31% in the left eye was recorded, persisting at the 1-year follow-up. VEPs were not delayed, the amplitude of N1-P1 deflections to 30' and 15' stimuli were 3.7–4.2, 5.4–6.2 μ V (normal mean 10.6 \pm 2.2, 11.6 \pm 2.1) 7.5 min of arc checks did not elicit reproducible VEPs in all recording sessions. The content of PCE or metabolites in a urine sample, tested by gas chromatography-mass spectroscopy [3], was performed 48 h after the onset of symptoms: a peak was found at 83% (40 mg/l) which indicated the presence of chloroform, a product of decomposition of trichloroacetic acid when heated at 80 °C for 20 min. The chloroform peak exceeded the limit ranges of the gas chromatographer. Blood samples collected 48 h after the onset of ON were tested with gas chromatography head space analysis [3] and showed the presence of PCE at a concentration of 1.08 mg/g 48 h after acute ON onset and 0.65 mg/g 32 h later. Plasmapheresis was not performed as PCE and chloroform would not have been filtered, because the molecules are transported with chylomicrons measuring about 140 daltons, therefore too small to be removed. Leber's optic neuropathy was ruled out by peripheral blood DNA analysis (mtDNA mutation at nucleotide 3460, 11778, 14484, 19257) [4]. One year after the onset of ON there was no evidence of clinical or electrophysiological improvement.

In our patient we could exclude a vascular origin of visual disturbances because the field deficit was not altitudinal, as in ischemic

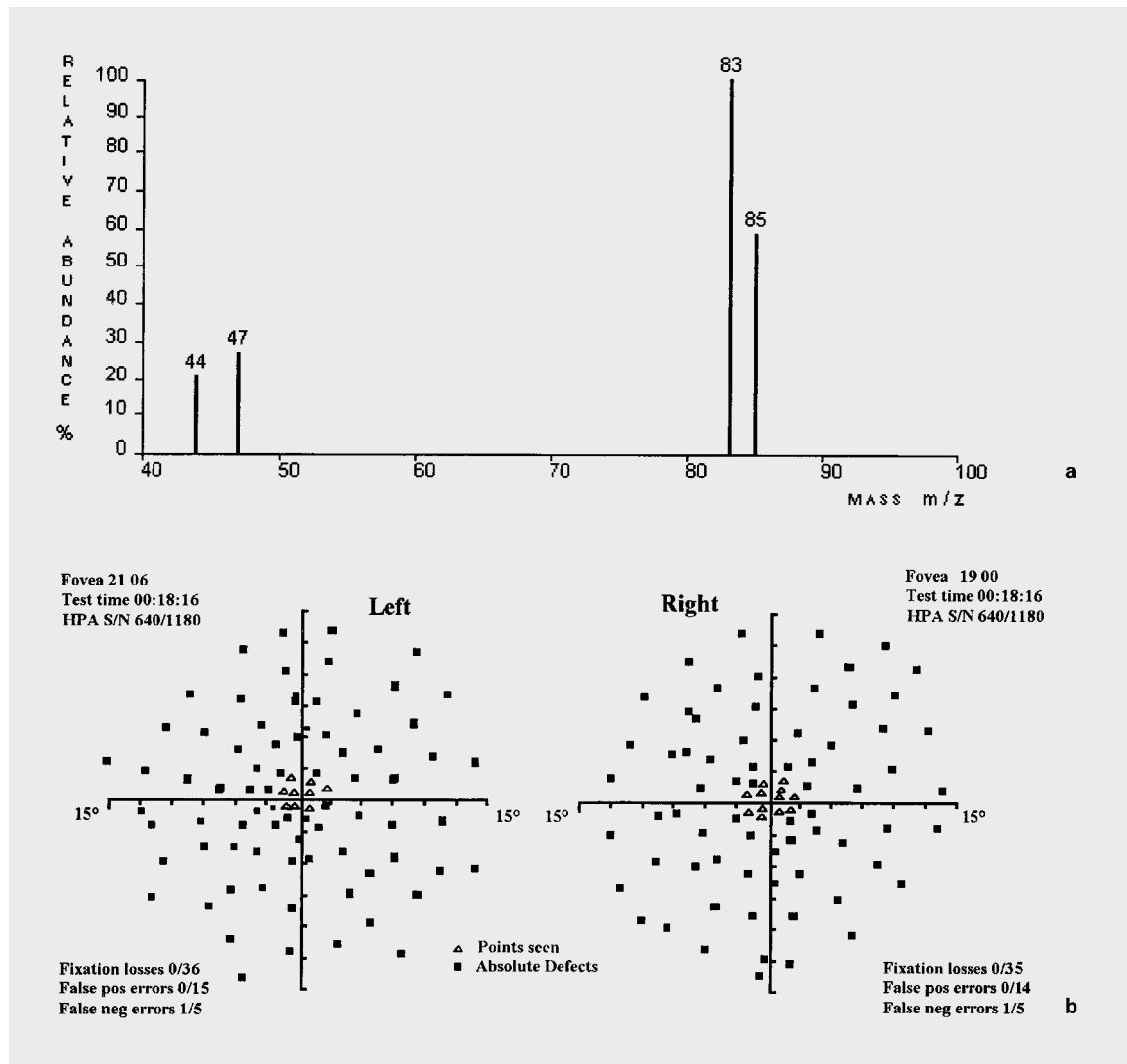


Fig. 2. a Gas chromatography spectrum analysis of CHCl_3 (chloroform) obtained from urine samples 2 days after acute symptoms. **b** Visual fields obtained 6 months after acute onset. Notice that only the central 2D radius vision is preserved.

optic neuropathy, and because of the normal MRI, excluding posterior cerebral artery occlusion. Retinopathy was excluded by fluorangiography, Leber's type ON was unlikely because of the paracentral peripheral type of field defect and because of absent genetic markers.

Nonorganic (functional) origin of the tubular vision was excluded by results of VEPs, of LTE-STIR MRI, and by the findings of consistent field defects with different perimetry and campimetry testing methods. Even though peripheral visual field constriction has been reported in typical presumed ON [5], because of bilateral symmetric presentation at onset and absence of visual field changes and VEP delays during follow-up, we concluded that it is likely that inhalation of solvents induced ON in our patient. The timing of the onset, and clinical and laboratory findings, suggest a relationship between ironing of PCE-cleaned fabrics and the ON here described. By measuring PCE levels in vapors produced by ironing of freshly dry-cleaned fab-

ric, we could show that there may be abnormally high levels of this solvent due to ironing, while normal levels might be picked up in other areas of the same room. As dry cleaning shops are mostly small entrepreneurial activities, it is likely that the owners, like our patient, will not pay sufficient attention to the actual risks of inhaling PCE vapors, and we suggest that a reminder of PCE toxicity might be useful. A previous report of major PCE toxicity described the sudden death of a 2-year-old boy after inhalation of PCE retained in freshly dry-cleaned curtains [6]. As a concluding remark we suggest that the reason why the central field was partly preserved in our patient, depends on the different size of optic nerve fibers in the central and eccentric retinal projection [7]: experimental studies showed that the toxic effect of organic solvents depends on the fibers size [8] and is more evident in large myelinated fibers.

References

- 1 American Conference of Governmental Industrial Hygienists: Threshold Limit Value for Chemical Substance and Physical Agents and Biological Exposure Indices for 1993–1994. Cincinnati, ACGIH 1993.
- 2 Onofrij M, Tartaro A, Thomas A, Gambi D, Fulgente T, Delli Pizzi C, Bonomo L: Long time echo STIR sequence magnetic resonance imaging of optic nerves in optic neuritis. *Neuroradiology* 1996;38:66–69.
- 3 Brugnone F, Perbellini L, Giulari C, Cerpelloni M, Soave M: Blood and urine concentrations of chemical pollutants in the general population. *Med Lav* 1994;85:370–389.
- 4 Johns RD: Genotype-specific phenotypes in Leber's hereditary optic neuropathy. *Clin Neurosci* 1994;2146–2150.
- 5 Optic Neuritis Study Group: The 5-year risk of MS after optic neuritis. Experience of optic neuritis treatment trial. *Neurology* 1997;49:1404–1413.
- 6 Garnier R, Bedouin J, Pepin G, Gaillard Y: Coin-operated dry cleaning machines may be responsible for acute tetrachloroethylene poisoning: Report of 26 cases including one death. *J Tox Clin Toxicol* 1996;34:191–197.
- 7 Potts AM, Hodges D, Shelman CB, Fritz KJ, Levy NS, Mangall Y: Morphology of the primate optic nerve. II Total fiber size distribution and fiber density distribution. *Invest Ophthalmol Vis Sci* 1972;11:989–1003.
- 8 Barret L, Torch S, Usson Y, Gonthier B, Saxod R: A morphometric evaluation of the effects of trichloroethylene and dichloroacethylene on the mental nerve. Preliminary results. *Neurosci Lett* 1991;131:141–144.

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Isolated Dysphagia Due to a Medullary Infarction: A New Lacunar Syndrome

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Disturbance of swallowing or dysphagia is common in stroke, occurring in about 30% of a mixed stroke population [1]. Although the localization of dysphagia in this setting has been generally attributed to bilateral corticobulbar disruption or a brainstem lesion, occurrence of dysphagia in unilateral cerebral lesions [2] and even without a demonstrable lesion [3] makes localization uncertain. In contrast to the poor localization of upper motor neuron (UMN) dysphagia, sudden isolated dysphagia of the lower motor neuron (LMN) type can be more consistently localized to the lateral medulla, as exemplified by the following 3 cases.

A 65-year-old woman with chronic hypertension developed sudden dysphagia on the morning of admission. Two days earlier, she had transient right face and body numbness, which she noted while washing her face. The sensory disturbance lasted 1 h with complete resolution. She denied any associated weakness or clumsiness. Examination was normal except for decreased gag with pooling of saliva. Her speech was mildly dysarthric. She was unable to initiate swallowing on command or use a straw properly. Her gag was decreased bilat-

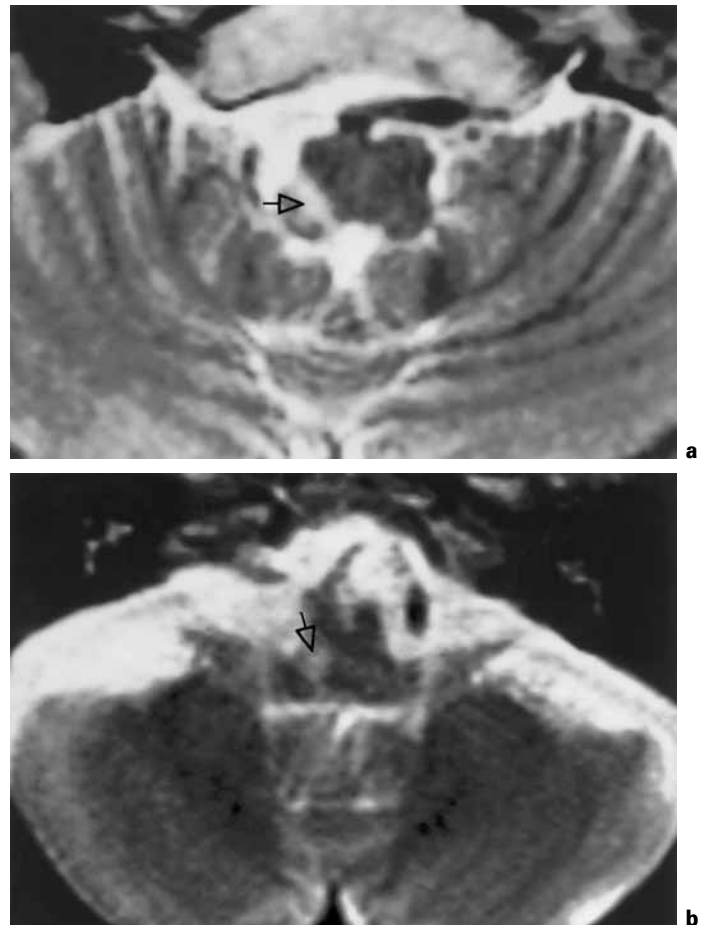


Fig. 1. Patient 1 (a) and patient 2 (b) are shown. T2-weighted MRI showing a small slit-like infarction in a similar region, corresponding to nucleus ambiguus or its fascicle in the lateral medulla. Each lesion is marked with an arrow.

erally with symmetric elevation of the uvula. The tongue protruded in the midline with good lateral movements on command. There was no evidence of limb or ocular disturbance. Indirect laryngoscopic examination confirmed normal vocal cord function. An MRI showed a small slit-like infarct in the right lateral medulla (fig. 1a). She recovered completely from the swallowing difficulty over a 3-week period.

A hypertensive 64-year-old man had sudden onset of dysphagia. Two years earlier, he underwent evacuation of a small right basal ganglia hemorrhage with full recovery. Examination showed decreased gag reflexes bilaterally with drooping of soft palate on the right, and mild dysarthria. An MRI showed a small linear infarction in the lateral portion of right medulla (fig. 1b). Nasogastric tube feeding was used. Two weeks later he was able to resume a regular diet.

A 72-year-old man with a history of diabetes, hypertension and hyperlipidemia had a sudden onset of difficulty with swallowing. Examination was normal except for decreased gag reflexes bilaterally. Mild dysarthria was noted, and the uvula was midline, with normal tongue mobility. No vocal cord paralysis was detected on laryngeal examination. An MRI showed a small infarction in the dorso-

lateral portion of left medulla. Swallowing difficulty gradually resolved over the ensuing 3 months.

All patients complained of isolated sudden dysphagia as the primary reason for seeking medical attention. Videofluoroscopy confirmed pharyngeal retention of dye and mild nasal regurgitation in all 3 cases, consistent with acute LMN dysfunction. There was no evidence of limb or ocular motility disturbance. Subsequent evaluation to look for evidence of large vessel disease or a source of embolus was unrewarding. Carotid and vertebral ultrasound, echocardiograms and MR angiography were all normal.

Despite the lack of complaints other than isolated dysphagia, examination by an experienced staff neurologist (BCL) disclosed mild dysarthria in all with impairment of gag reflexes and oropharyngeal mobility in most. Although predominant dysarthria as a manifestation of small vessel disease is rare and usually occurs in the context of UMN lesions [4, 5], in our patients dysphagia was the major reason for seeking medical care, and all had lesions compatible with LMN involvement.

In pseudobulbar or suprabulbar palsy, lower cranial nerve dysfunction occurs secondary to an UMN lesion. Interruption of bilateral corticonuclear pathways results in UMN-type bulbar symptoms [6], and if corticospinal tracts are involved, spastic hemiparesis may be superimposed. Typical underlying lesions include small vessel infarcts or hemorrhages against the background of chronic hypertension [6]. We are not aware of any published reports of sudden isolated dysphagia as a forme fruste of pseudobulbar palsy, but this may occur. Our cases suggest that a sudden onset of isolated dysphagia in patients with risk factors for small vessel disease is more likely due to an LMN infarction near the nucleus ambiguus or its fascicle at the medullary level. Buchholz [3] already emphasized lower brainstem stroke as the probable cause when he presented 10 patients with isolated dysphagia as the major complaint. Nine patients had either chronic hypertension or diabetes, but brainstem lesions were not visualized on MRI scans in any of his patients.

The lacunar syndrome refers to a clinical presentation 'where the mechanism of infarction involves transient or permanent occlusion of a single penetrating artery with a high degree of probability' [7]. Kim et al. [8] noted that nearly 25% of patients meeting the clinical diagnosis of the Wallenberg syndrome had either normal MR angiography or conventional angiography. Presumably some had penetrating artery disease not visualized on angiography. Our cases with sudden onset of stereotypic isolated dysphagia may represent a partial Wallenberg syndrome. This limited clinical manifestation, along with an MRI lesion occupying only a small portion of the lateral medulla, is compatible with an occlusion of a penetrating artery. We prefer the term 'isolated dysphagia due to a medullary lacune' for these cases.

In summary, acute onset of isolated dysphagia may occur as a manifestation of a lacunar syndrome. The lesion is most likely in the lateral medulla. These patients may be at an increased risk of acute aspiration, but gradual improvement of swallowing can be expected.

References

- 1 Horner J, Massey EW, Riski JE, Lathrop DL, Chase KN: Aspiration following stroke: Clinical correlates and outcome. *Neurology* 1988;38:1359-1362.
- 2 Meadows JC: Dysphagia in unilateral cerebral lesions. *J Neurol Neurosurg Psychiatry* 1973;36:853-860.
- 3 Buchholz DW: Clinically probable brainstem stroke presenting primarily as dysphagia and nonvisualized by MRI. *Dysphagia* 1993;8:235-238.
- 4 Arboix A, Massons J, Oliveres M, Titus F: Isolated dysarthria. *Stroke* 1991; 22:531.

- 5 Kim JS: Pure dysarthria, isolated facial paresis, or dysarthria-facial paresis syndrome. *Stroke* 1994;25:1994-1998.
- 6 Besson G, Bogousslavsky J, Regeli F, Maeder P: Acute pseudobulbar or suprabulbar palsy. *Arch Neurol* 1991;48:501-507.
- 7 Donnan GA, Norrving B, Bamford JM, Bogousslavsky J: Subcortical infarction: Classification and terminology. *Cerebrovasc Dis* 1993;3:248-251.
- 8 Kim JS, Lee JH, Lee MC: Patterns of sensory dysfunction in lateral medullary infarction. *Neurology* 1997;49:1557-1563.

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Volitional Monocular Lilliputian Visual Hallucinations and Synesthesia

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Depression, mania, bipolar affective illness with rapid cycling, and paranoid schizophreniform psychoses are common sources of distress in individuals with multiple sclerosis (MS) exhibiting demyelinating plaques over the temporal lobes [1]. Dementia may occur in MS subjects. A previously reported patient with complex visual hallucinations of religious content and dementia had a large, left frontal plaque and multiple temporal and occipital lobe plaques located over the periventricular white matter [2]. The authors suggested the existence of a disconnection syndrome caused by demyelination of the long subcortical white matter association bundles of the cerebral hemispheres [2]. A patient with MS and complex behavioral manifestations, initially confused with psychiatric illness, is reported. She had no previous history of psychiatric disorders. Despite her isolated complex hallucinations and unusual experiential phenomena, she had no clinical evidence of dementia or intercurrent psychosis.

A 54-year old female with no personal or family history of neurologic or psychiatric illnesses suffered a grand mal seizure without aura 7 years prior to her neurologic consultation. She was healthy until her first seizure, and no concurrent illness was identified on initial evaluation. She had no family history of epilepsy and was taking no prescription or illicit drugs. After her first seizure, she had ataxia and retrograde amnesia for a few weeks. Her initial MRI of the brain showed white matter lesions of the cerebral hemispheres. Oligoclonal bands were recovered from her cerebrospinal fluid (CSF). CSF cell count and protein content were normal, and cultures for bacteria and viruses were negative. Her EEG showed bilateral temporal theta but no specific epileptic discharges. She was diagnosed with multiple sclerosis and temporal lobe epilepsy. A second grand mal seizure took place 6 months later, and treatment was initiated with carbamazepine. Her second seizure was followed in 2 days by visual hallucinations of small objects while remaining fully conscious. She also noticed that the last words of a sentence, or a full sentence said to her, would repeat within her head over and over

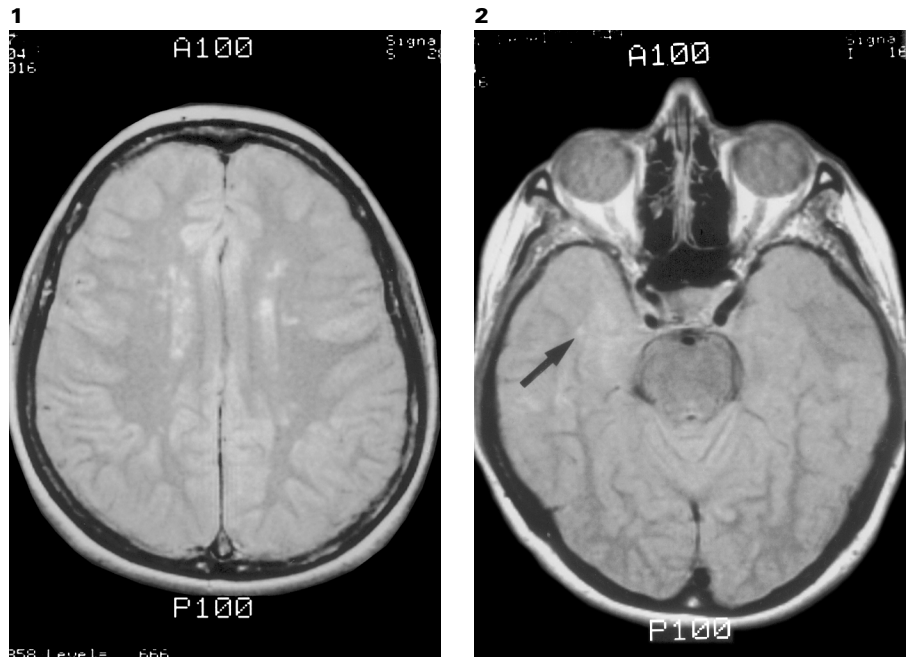


Fig. 1. Flame-shaped irregular subcortical periventricular white matter lesions of the cerebral hemispheres are evident (T₂-weighted images).

Fig. 2. A probable small area of demyelination of the right temporal lobe is indicated by the arrow (T₂-weighted images).

again, sometimes for as long as 30 min (echoacousia and palinacousis). Spontaneous auditory hallucinations of music, bells ringing, clocks ticking, dogs barking, cats meowing or birds chirping followed. Within months, she noticed that objects or the faces of people she stared at persisted in her sight for minutes at a time (palinopsia). If she shifted her eyes to the wall, the faces or objects appeared on the wall (visual allesthesia). At one time her face appeared '25 years younger' when she looked at herself in the mirror (facial rejuvenation). Over time, auditory hallucinations became very vivid and continuous ('racing'), to the extent that she had a very hard time falling asleep and required hypnotics. She woke up every morning to loud music within her head. She was referred to psychiatry for treatment, despite that with her hallucinations she did not experience any other alterations of consciousness, or exhibit any bizarre behavior. She was empirically treated with various tranquilizers, including haloperidol, for several years. Grand mal seizures never recurred, even after carbamazepine was discontinued, but her hallucinations did not mitigate with any treatment. Three years after the onset of her illness, she developed the unusual ability of voluntarily bringing on audiovisual TV-like hallucinations of small objects, animals, or people that either made sounds, spoke, or sang to her. For instance, if she thought of a specific famous country singer, the singer immediately appeared over her left eye singing (synesthesia). Her visual hallucinations were always stationary and confined to the upper temporal quadrant of her left eye. If she thought of an old classmate, she would see him or her clearly, the way the person was during their school years, talking to her. What the person said was appropriate and was not threatening or anxiety provoking. In other words, the content of what was said 'made sense' within the context of her retrieved memory. She was unable to hold conversations with her hallucinatory persons and had clear recollection of her hallucinations. Her general physical examination was normal. On mental status examination she was alert and oriented to time, space and person. She was appropriate, pleasant, cooperative and well-groomed. Mood and affect were normal. She exhibited no

flight of ideas, hesitant, slow or bizarre thinking. Her answers were relevant and her statements logical; she had no delusions, ideas of reference, or ideas of influence. Speech was fluent, repetition was intact, and comprehension of spoken language was preserved. She was able to read aloud and silently, write spontaneously and to dictation, and draw figures normally. Mathematical calculations, including serial subtractions and ability to abstract, were normal. Short- and long-term memory were normal, as well as her attention span. There was no confabulation. Fund of knowledge, judgment, and insight of illness were good. She could recognize objects placed in her hands and had no difficulties with fine finger motor control. She could imitate gestures, and exhibited no neglect, apraxia, aprosody or prosopagnosia. She could initiate her visual hallucinations as described above on many occasions during her repeated neurologic examinations while otherwise acting appropriately. She exhibited no mood changes, alteration of consciousness, facial transfiguration, or anxiety during her visual phenomena. No stereotyped or involuntary movements were noted at those times. On neurologic examination, corrected visual acuity, visual fields, and pupillary responses were normal. Ocular movements were full and symmetric in all directions of gaze. No nystagmus was observed. Hearing was normal. Testing of the additional cranial nerves, equilibrium, muscle tone, muscle strength and sensory examination for all modalities was normal. Deep tendon reflexes were symmetric and she had no grasp, rooting, or palmomental reflexes. She had bilateral Babinski signs. There was no gait disturbance and no tremors or dystonia. A repeat MRI of the brain revealed occipital periventricular areas of demyelination, subcortical white matter hyperintensities, and one small area of demyelination over the right temporal lobe (fig. 1, 2). There was no hydrocephalus and no brain stem lesions. Short and middle latency auditory evoked potentials were normal. Her blink reflexes showed a delay of the R1 component. Facial electromyogram showed polyphasic motor units of long duration over the frontalis, mentalis and orbicularis oris muscles, indicative of chronic denervation with reinnerva-

tion. Baseline EEG was normal. Her video-EEG monitoring (no sphenoidal electrodes were employed) and brain SPECT during her typical voluntarily induced visual hallucinations failed to demonstrate epileptic discharges or blood perfusion abnormalities from the normal resting awake state. Treatment with clonazepam helped her to sleep, but did not change her hallucinations. She reported no changes in her symptoms on follow-up visits.

Complex partial seizures of purely cognitive content in MS patients can be confused with intermittent behavioral manifestations of the disease due to a relapse. Therefore, it is important that epileptic events are ruled out in patients with MS who exhibit unusual recurrent symptoms, especially in the absence of motor manifestations. Although this patient had two grand mal seizures at the onset of her illness, her EEG was normal with no paroxysmal activity recorded at rest or during her spontaneous or volitional visual hallucinations. Furthermore, a brain SPECT that was completed during her volitional and spontaneous experiential phenomena was unchanged from the normal resting state while awake. Her visual symptoms must be distinguished from complex visual hallucinations and palinopsia secondary to disorders of the visual pathways. The case herein discussed had bilateral white matter hyperintensities on MRI, but no detectable abnormalities on her visual field testings.

Dysmetropsia is the disorder of visual perception characterized by an apparent modification of the size of perceived objects. Objects appear either enlarged (macropsia) or shrunk (micropsia). Monocular micropsia is a symptom of retinal edema, while hemimicropsia refers to a selective disturbance in the perception of the size of an object, contralateral to a hemispheric lesion, in the absence of visual field deficits [3]. On the other hand, lilliputian or smaller than life-size colorful visual hallucinations in a clear sensorium are typical of peduncular hallucinosis in patients with lesions of the mesencephalon [4]. This patient's complex symptoms are unique because her lilliputian imagery was confined to the temporal superior quadrant of her left visual field, was faithfully reproduced voluntarily, and she had no detectable abnormalities on her visual field testing and no brain stem, thalamic, or optic nerve lesions. Although the temporal lobe demyelinating lesion may have interrupted conduction through callosal fibers (fig. 1) or through the uncinate fasciculus (fig. 2), she had none of the clinical signs of callosal disconnection and there was no evidence of transcortical aphasia. Finally, synesthesia (experiential phenomena perceived in two or more senses simultaneously) of temporal lobe epileptic origin was ruled out in this patient by her normal ictal EEG [5]. It is a common observation in clinical practice that patients with episodic painful or epileptic disorders have an inherent permissive ability to 'prime' or 'cue' their episodes, either consciously or unconsciously. In multiple sclerosis, amplification of abnormal signals arising from the areas of demyelination perhaps may be 'gated ad lib' in some exceptional cases, as the one detailed here [6]. The discovery of Bumgartner and Epstein [7] that normal individuals can voluntarily alter their visual evoked potentials without shifting their eyes supports this inherent quality of the human brain to freely modify ongoing electrophysiologic activities. The ultimate basic pathophysiology of this patient's experiential phenomena is uncertain; it may be speculated that 'cross talking' of demyelinated subcortical white matter fibers caused her symptoms, since her ictal EEG and SPECT studies were normal, revealing no epilepsy or abnormalities of brain perfusion. It is unclear what role was played by the commissural occipital fibers of the tapetum lining the walls of the posterior ventricular horns in the expression or modulation of this patient's visual hallucinations.

Very recently, a patient was described with TV-like 'picture within a picture' lilliputian visual hallucinations that were embedded in her left inferior visual field, and emerged from an otherwise normal visual background. She was aware of the hallucinatory nature of her experience but had no synesthesia and no volitional control over her hallucinations. A left inferior quadrantanopsia was present and her brain MRI revealed a parieto-occipital stroke [8].

References

- 1 Honer WG, Hurwitz T, Li DKB, Palmer MSC, Paty DW: Temporal lobe involvement in multiple sclerosis patients with psychiatric disorders. *Arch Neurol* 1987;44:187-190.
- 2 Fontaine B, Seilhean D, Tourbah A, Daumas-Duport C, Duyckaerts C, Benoit N, Devaux B, Hauw J-J, Rancurel G, Lyon-Caen O: Dementia in two histologically confirmed cases of multiple sclerosis: One case with isolated dementia and one case with psychiatric symptoms. *J Neurol Neurosurg Psychiatry* 1994;57:353-359.
- 3 Cohen L, Gray F, Meyrignac C, Dehaene S, Degos J-D: Selective deficit of visual size perception: Two cases of hemimicropsia. *J Neurol Neurosurg Psychiatry* 1994;57:73-78.
- 4 Lhermitte J: Syndrome de la calotte du pédoncle cérébral. Les troubles psycho-sensoriels dans les lésions du mesocéphale. *Rev Neurol (Paris)* 1922; 38:1359-1365.
- 5 Jacome DE, Gumnit RJ: Audioalgesic and audiovisuoalgesic synesthesias: Epileptic manifestation. *Neurology* 1979;29:1050-1053.
- 6 Eisen A, Purves S, Hoirsch M: Central nervous system amplification: Its potential in the diagnosis of early multiple sclerosis. *Neurology* 1982;32: 359-364.
- 7 Bumgartner J, Epstein CM: Voluntary alteration of visual evoked potentials. *Ann Neurol* 1982;12:475-478.
- 8 Benegas NM, Liu GT, Volpe NJ, Galetta SL: 'Picture within a picture' visual hallucinations. *Neurology* 1996;47:1347-1348.

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Ganglion Cyst of the Peroneal Nerve: A Differential Diagnosis of Peroneal Nerve Entrapment Neuropathy

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A 51-year-old man presented with a 7-month history of intermittent pain along the lateral side of the left leg and 2 days prior to admission he was unable to lift his left foot. No pain or other sensory symptoms were noted. No evidence of external compression of the peroneal nerve at the fibular head and no weight loss were reported.

Physical examination revealed a complete deficit of dorsiflexion of the left foot but eversion of the foot in plantar flexion was normal and no sensory deficits were found. No abnormal mass was palpable around the peroneal head or in the popliteal fossa. Pressure exerted in the popliteal fossa produced local pain with weak radiation towards the toes (positive Tinel sign). Neurography of the deep peroneal nerve using the 'inching technique' demonstrated a conduc-

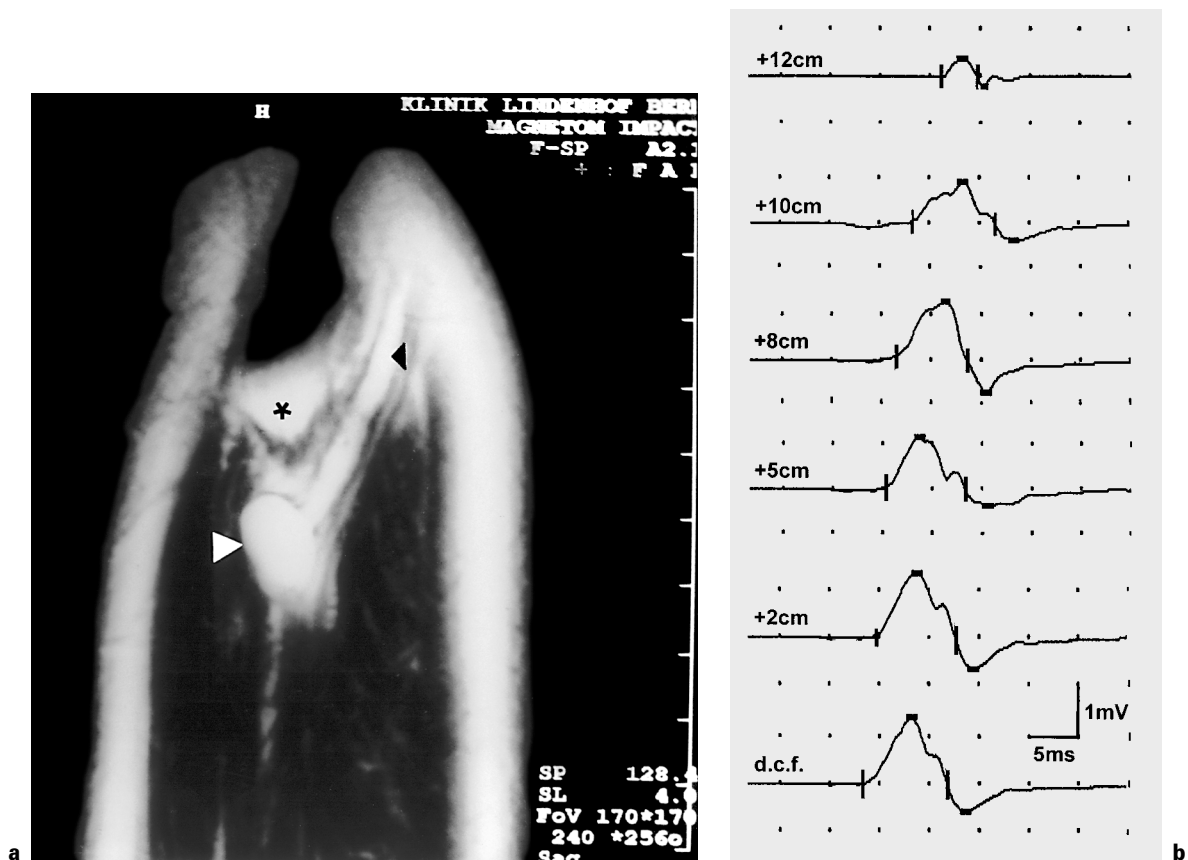


Fig. 1. **a** Sagittal T₂-weighted MR scan (TR 3,600/TE 120) laterally through the fibular head (*) demonstrates the cystic mass (▷) along the posterior aspect of the anterior tibial and extensor digitorum longus muscles and its extension along the peroneal nerve up to the popliteal fossa (◄). **b** Peroneal nerve neurography with 'inching technique'. Note the partial block at 10 cm which is more pronounced at 12 cm above the standard stimulation site which is located 2–3 cm distal of the fibular head. d.c.f. = Distal capitulum fibulae.

tion block (amplitude reduction to 30% compared to distal stimulation) localized 10–12 cm proximal to the fibular head (fig. 1b) in the popliteal fossa, corresponding to the site of the positive Tinel sign. Magnetic resonance imaging (MRI) of the left knee showed a polycystic mass localized close to the proximal tibiofibular joint (fig. 1a).

The diagnosis of a ganglion cyst of the common peroneal nerve was made, and surgical decompression was performed. The common peroneal nerve showed marked thickening beginning 3 cm above the level of the femorotibial joint and extending distally as a glassy, irregular mass intraneurally in the deep peroneal nerve sheath. Under magnification by means of the operating microscope the intraneural 'sausage-like' extension of the ganglion was located exclusively within the nerve fascicles contributing to the deep peroneal branches, while the fascicles destined for the superficial peroneal branches were unaffected. The total craniocaudal length of the mass was 12 cm. Histological investigation confirmed the diagnosis of a ganglionic cyst. At follow-up a year later, the patient showed a minimal deficit of dorsiflexion of the left foot without any significant gait difficulties.

The clinical picture of our case resembled a peroneal entrapment neuropathy at the fibular head and no mass was palpable. However, the history was negative for factors causing external nerve compres-

sion at the fibular head, such as habitual leg-crossing or kneeling for a long time. Clinical examination showed a sparing of the superficial peroneal nerve and a positive Tinel sign atypically located in the popliteal fossa. Neurography confirmed an atypical site of the conduction block in the popliteal fossa, 12 cm proximal to the fibular head, which was evidence against a common entrapment neuropathy and prompted an MRI investigation.

Peripheral nerve lesions due to intraneural ganglions are rare clinical findings but are well described in the literature [1–5]. Their most frequent site by far is the peroneal nerve [3]. The clinical onset of a peroneal nerve lesion due to a ganglion cyst is marked by pain on the anterolateral surface of the leg followed by weakness of the muscles of the anterior compartment of the leg. Paralysis of the peroneal muscles is less common and typically, there is little or no sensory deficit indicating that the superficial peroneal nerve is spared. According to Parkes [4] the intraneural extension of the ganglion originating from the proximal tibiofibular joint distally and proximally along the deep peroneal nerve is due to a small recurrent articular branch from the deep peroneal nerve innervating this joint. This explains the predominant involvement of the muscles supplied by the deep peroneal nerve and the relative sparing of the superficial peroneal branches.

However compression of the common peroneal nerve may also present as complete paresis of both branches [6], with a clinical picture not different from the much more frequent entrapment neuropathy at the fibular head.

Clinical evidence of a local mass and confirmation of the diagnosis by ultrasonography, CT or MRI should prompt early surgical decompression, as this offers the best prognosis for recovery [7]. In the absence of a palpable mass, which is sometimes the case with ganglion cysts, the diagnosis should nevertheless be considered whenever the history is negative for factors causing external nerve compression or when the clinical presentation is atypical of the self-limited entrapment neuropathy at the fibular head. In this situation it is important to perform neurography and MRI [8, 9].

References

- 1 Brooks DM: Nerve compression by simple ganglia: A review of thirteen collected cases. *J Bone Joint Surg* 1952;34-B:391–400.
- 2 Barrie HJ, Barrington TW, Colwill JC, Simmons EH: Ganglion migrans of the proximal tibiofibular joint causing lesions in the subcutaneous tissue, muscle, bone, or peroneal nerve: Report of three cases and review of the literature. *Clin Orthop* 1980;149:211–215.

- 3 Nucci F, Artico M, Santoro A, Bardella L, Delfini R, Bosco S, Palma L: Intraneural synovial cyst of the peroneal nerve: Report of two cases and review of the literature. *Neurosurgery* 1990;26:339–344.
- 4 Parkes A: Intraneural ganglion of the lateral popliteal nerve. *J Bone Joint Surg* 1961;43-B:784–790.
- 5 Hartwell AS: Cystic tumour of median nerve. Operation: Restoration of function. *Boston Med Surg J* 1901;144:582–583.
- 6 Stack RE, Bianco AJ, MacCarthy CS: Compression of the common peroneal nerve by ganglion cysts. *J Bone Joint Surg* 1965;47-A:773–778.
- 7 Ghossain M, Mohasseb G, Dagher F, Ghossain A: Compression du nerf sciatique poplité externe par un kyste synovial. *Neurochirurgie* 1987;33:412–414.
- 8 Coakley FV, Finlay DB, Harper WM, Allen MJ: Direct and indirect MRI findings in ganglion cysts of the common peroneal nerve. *Clin Radiol* 1995; 50:168–169.
- 9 Leon J, Marano G: MRI of peroneal nerve entrapment due to a ganglion cyst. *Magn Reson Imaging* 1987;5:307–309.

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