studies showing that visual cortical areas process tactile information in Braille readers.⁹ Dissociation between verbal and musical alexia in Braille has been reported following left middle cerebral artery infarction in a professional organist blind from childhood.³

Braille alexia, the tactile homologue of pure alexia (alexia without agraphia), a category specific visual agnosia, may result from disruption of different, possibly overlapping, psychoperceptual mechanisms, some analogous to those postulated in pure alexia.¹⁰ It may reflect problems integrating tactile information over the temporal or spatial domains, associative forms of agnosia (or tactile simultanagnosia). A frontal-parietal network may contribute to the integration of perception with action over time,¹¹ and right hemisphere lesions may be associated with impaired integration of spatial information from multiple stimuli.¹² Alternatively, Braille alexia may reflect perceptual impairment, an apperceptive form of agnosia. As Braille characters are close to the limits of normal perceptual resolution, impaired light touch perception following damage to the primary sensorimotor cortex or its connections may result in degraded tactile identification and slowed Braille reading speed.

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doi: 10.1136/jnnp.2006.106922

Competing interests: None.

References

- 1 Gloning I, Gloning K, Weingarten K, *et al.* Über einen Fall mit Alexie der Brailleschrift. *Wien Z Nervenheilkd Grenzgeb* 1954;**10**:260–73.
- 2 Birchmeier AK. Aphasic dyslexia of Braille in a congenitally blind man. *Neuropsychologia* 1985;**23**:177–93.
- 3 Signoret J-L, van Eeckhout P, Poncet M, *et al.* Aphasie sans amésie chez un organiste aveugle. *Rev Neurol Paris* 1987;**143**:172–81.
- 4 Perrier D, Belin C, Larmande P. Trouble de la lecture du Braille par lésion droite chez une patiente devenue aveugle. *Neuropsychologia* 1988;**26**:179–85.
- 5 Hamilton R, Keenan JP, Catala M, *et al.* Alexia for Braille following bilateral occipital stroke in an early blind woman. *Neuroreport* 2000;**11**:237–40.
- 6 Maeda K, Yasuda H, Haneda M, *et al.* Braille alexia during visual hallucination in a blind man with selective calcarine atrophy. *Psychiatry Clin Neurosci* 2003;**57**:227–9.
- 7 Rudel RG, Denckla MB, Spalten E. The functional asymmetry of Braille letter learning in normal, sighted children. *Neurology* 1974;**24**:733–8.
- 8 Larner AJ. "Dementia unmasked": atypical, acute aphasic, presentations of neurodegenerative dementing disease. *Clin Neurol Neurosurg* 2005;**108**:8–10.
- 9 Sadato N. How the blind see Braille: lessons from functional magnetic resonance imaging. *Neuroscientist* 2005;**11**:577–82.
- 10 Farah MJ. Visual agnosia. *Disorders of object recognition and what they tell us about normal vision*. Cambridge: MIT Press, 1995.
- 11 Quintana J, Fuster JM. From perception to action: temporal integrative functions of prefrontal and parietal neurons. *Cereb Cortex* 1999;**9**:213–21.
- 12 Carmon A, Benton AL. Tactile perception of direction and number in patients with unilateral cerebral disease. *Neurology* 1969;**19**:525–32.

Acute myelopathy selectively involving lumbar anterior horns following intranasal insufflation of ecstasy and heroin

We report a patient who developed acute myelopathy after intranasal insufflation of amphetamines and heroin. The functional prognosis was very poor; after 4 months, she remained paraplegic. MRI imaging showed selective T2 hyperintensity and intense enhancement confined to the spinal anterior horns and lumbar nerve roots and plexus. This unique MRI pattern, together with neurophysiological data, suggests that the pathological process at the first primary affected spinal anterior horns (SAH), conditioning motoneuron cell death, and then nerve roots and lumbar plexus as a consequence of wallerian degeneration

Case report

A 17-year-old girl was admitted to the emergency department in a drowsy state and unable to walk after an overdose of intranasal insufflated heroin and amphetamines. After a few hours, drowsiness progressed to stupor, and progressive weakness in all four limbs, mainly involving the lower limbs, developed. At that time, laboratory data showed massive rhabdomyolysis (creatinine phosphokinase 36 880 mg/dl) with acute renal failure (ARF), and hepatic failure; medical therapy was promptly started. The patient's past medical history was unremarkable except for habitual use of amphetamines (ecstasy) and cannabinoids since the age of 12 years. The previous week she had insufflated heroin about once a day; the previous night she reported a double dose of heroin consumption, and a high dose (approximately 1 g) of intranasal insufflation of amphetamines. The next day the patient was alert and cooperative but complained about diffuse muscle pain and tenderness, prevailing

in the lower limbs, where weakness rapidly worsened to flaccid, areflexic paralysis.

Urinary chromatography detected amphetamines (3-4-methylenedioxymethamphetamine (MDMA) 7680 ng/ml, 3-4-methylenedioxymphetamine 2000 ng/ml) and opiates (>2000 ng/ml, morphine 228 ng/ml). CSF protein content was slightly increased with no intrathecal Ig synthesis; glucose, chloride and cells were within the normal range. All virological analyses, including HIV, were negative. Rhabdomyolysis related ARF completely normalised by day 10, with resumption of spontaneous diuresis; from that time on, no more myalgias were reported. Muscle weakness promptly improved in the upper limbs to normal strength while flaccid paralysis persisted in the lower limbs. The patient had no sensory symptoms or signs, and no sphincteric abnormalities were observed.

Motor evoked potentials recorded from foot muscles were absent from day 9 onwards; both sural nerve sensory action potentials (SAPs) and tibial somatosensory evoked potentials (SSEPs) were normal. On day 16, electromyography revealed complete unexcitability of the leg motor nerves, together with fibrillation potentials without any voluntary electromyographic activity in all lower limbs and paraspinal muscles innervated by L3–S1 roots. Quantitative sensory test, skin sympathetic reflex and urodynamic test were all within the normal range.

MRI performed at the beginning of symptoms was negative. One month later, MRI showed selective T2 hyperintensity of the anterior horns and signal alteration of lumbar nerve roots, associated with volume increase and intense gadolinium enhancement (fig 1).

Intravenous high dose steroid therapy was started with no clinical benefit. MRI signal alteration and enhancement of the nerve roots almost completely disappeared while signal alteration of the anterior horns remained, although it was reduced in intensity. Four months later, the patient was still paraplegic.

Figure 1 MRI imaging. (A) Post-gadolinium (standard dose) T1 weighted MRI image, coronal view: linear enhancement along the nerve roots is evident. (B) T2 weighted MRI image, coronal plane: widespread T2 hyperintensity along nerve roots is more evident on the right side. (C) Post-gadolinium (standard dose) T1 weighted MRI image, axial view: selective anterior horn intense enhancement is seen. (D) T2 weighted MRI image, axial plane: selective anterior horn T2 hyperintensity is noted.

Discussion

Rhabdomyolysis, ARF, acute hepatotoxicity and transverse myelitis (TM) are well known complications of intravenous heroin assumption.^{1,2} A single case of acute myelopathy as a consequence of heroin inhalation has been reported following, in common with other cases, a period of abstinence preceding the event; moreover, recovery was almost complete 1 month later.¹ No cases of acute MDMA myelopathy are known; rhabdomyolysis is known to complicate MDMA consumption.³

This case particularly highlights selective involvement of the lumbar motoneurons, as supported by neurophysiological and neuro-radiological evidence. Unexcitability of motor fibres and complete denervation in the lower limbs and paraspinal muscles, with spared SAPs and SSEPs, suggest damage of SAH and/or ventral root axons. MRI confirmed selective T2 hyperintensity confined to the SAH and nerve roots, with intense enhancement in both, spreading from D10 up to the conus (fig 1). This unique MRI pattern has never been reported previously to our knowledge, and differs from MRI patterns observed in TM and radiculitis. In TM, MRI can sometimes show an inflammatory lesion localised within the cord, usually not involving more than 3–4 vertebral segments (more often at the dorsal level), hyperintense in T2 weighted images and with nerve root sparing. In the acute phase, enhancement is generally present, but not involving the nerve roots.⁴ In radiculitis (eg, Guillain-Barré syndrome), MRI signal alterations are commonly localised to spinal anterior roots, without any involvement of the spine.⁵ Moreover, the MRI pattern did not suggest a spinal infarction. The anterior spinal artery derives from a single anterior radiculo-medullary artery (Adamkiewicz artery) that arises from L3 upwards; spinal infarction is rare and occurs mostly in the cervical spine. MRI abnormalities on sagittal T2 weighted images in our patient are not consistent with typical "pencil-like" hyperintensities and cord enlargement.

Therefore, taken together, the clinical, neuroradiological and neurophysiological data suggest that the pathological process primarily affected SAH conditioning motoneuron cell death, and then nerve roots and lumbar plexus as a consequence of wallerian degeneration.

Suggested pathogenetic mechanisms of heroin associated acute myelopathy include hypotension watershed zones ischaemia, vasculitis and hypersensitivity reaction. Among the most credited hypothesis is the possibility of a direct toxic effect of heroin.^{1,2} In our case, we speculate that a pathogenetic role played by MDMA cannot be excluded and may contribute towards an explanation of some of the atypical features such as the lack of a clear period of abstinence from heroin, as described in most cases of opiate toxic myelopathy, and the unusually high dose of MDMA inhaled. Moreover, it has been proved that the lumbar motoneurons express high levels of serotonin receptors (mediating MDMA effects) whereas opiates receptors are three times more represented in the dorsal horns compared with the ventral horns of the spinal cord.⁶

This case increases our knowledge of the possible severe acute toxic effects of intranasal insufflation of heroin and ecstasy, often considered by the public as "safe" drugs. The selective involvement of the anterior horns of the spinal cord indicates a poor functional prognosis and suggests a specific, but still

unknown, pathogenetic effect on the spinal motoneurons, possibly toxic or immunopathological in nature.

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doi: 10.1136/jnnp.2006.109207

Competing interests: None.

References

- McCreary M, Emermann C, Hanna J, *et al*. Acute myelopathy following intranasal insufflation of heroin: a case report. *Neurology* 2000;**55**:316–17.
- Buttner A, Mall G, Penning R, *et al*. The neuropathology of heroin abuse. *Forensic Sci Int* 2000;**113**:435–42.
- Liechti ME, Kunz I, Kupferschmidt H. Acute medical problems due to Ecstasy use. Case-series of emergency department visits. *Swiss Med Wkly* 2005;**135**:652–7.
- Choi KH, Lee KS, Chung SO, *et al*. Idiopathic transverse myelitis: MR characteristics. *AJNR Am J Neuroradiol* 1996;**17**:1151–60.
- Byun WM, Park WK, Park BH, *et al*. Guillain-Barré syndrome: MR imaging findings of the spine in eight patients. *Radiology* 1998;**208**:137–41.
- Morales M, Battenberg E, Bloom F. Distribution of neurons expressing immunoreactivity for the 5HT3 receptor subtype in the rat brain and spinal cord. *J Comp Neurol* 1998;**385**:385–401.

Unusual case of tick borne encephalitis with isolated myeloradiculitis

Tick borne encephalitis (TBE) virus causes the most important arthropod transmitted disease in central Europe. In endemic areas, TBE has an incidence of 1.2 per 10 000 and a mortality of approximately 1%. The TBE virus is a neurotropic human pathogen. The most common presentations are meningitis (49%), meningoencephalitis (41%) and meningoencephalomyelitis (10%).¹ Patients with concomitant spinal cord involvement are thought to be affected more severely and mechanical ventilation is often necessary.²

We present an unusual TBE case with an isolated myelitis without signs of meningitis or meningoencephalitis.

Case report

A 43-year-old man from the Black Forest area, Germany, who had not had TBE vaccination developed symptoms of gastroenteritis with fever (diarrhoea, nausea and vomiting) lasting for approximately 1 week. Two days after recovery from the gastrointestinal symptoms he experienced severe pain in his shoulders followed by proximal pareses of his brachial muscles without signs of meningism. As the symptoms were not ameliorating during the

following days, the patient was admitted to hospital. At that time he had severe pareses of the upper extremities and mild pareses of the lower extremities. All tendon reflexes appeared very brisk but pyramidal signs were absent. Sensory testing was normal. The patient recalled a tick bite several weeks ago, after having collected mushrooms in the forest.

The following laboratory findings were abnormal: increased C reactive protein 8 mg/l, leucocytosis 12 200/μl, cerebrospinal fluid with mild pleocytosis (49 cells/μl: 80% lymphocytes, 20% monocytes), a clearly increased total protein content (2070 mg/l), local IgM synthesis of 83.5% and an increased IgG index of 0.81. Serological testing of the CSF and serum showed positive IgM and IgG antibody reactivity against TBE virus. A virus specific IgG CSF/serum index of 3.69 (normal value <2) confirmed high intrathecal specific antibody production. Coinfection with *Borrelia burgdorferi* was excluded. Moreover, there was no evidence of further possible aetiologies, such as infection by *Treponema pallidum*, herpes simplex virus 1 and 2, varicella zoster virus, enterovirus, HIV, Epstein-Barr virus or cytomegalovirus, or evidence of a vasculitis or collagenosis.

On electromyography and electroneurography, axonal lesions were seen in the left deltoid (C5/6), the left triceps (C7/8) and the left abductor digiti minimi manus (C8/Th1). There was normal neurography of the right and left radial nerve, the left ulnar and the left median nerve, and thus a brachial plexus lesion was excluded. In summary, there was evidence of a polyradiculitic or anterior horn lesion from C5 to Th1 (maximum at C5 and C7).

Motor evoked potentials showed delayed central motor conduction to both legs and the right arm. Tibial somatosensory evoked potentials were pathological on the left side, also with a central delay. The clinical and electrophysiological pattern was compatible with an incomplete myelitis involving the anterior horn cells.

Spinal MRI performed 25 days after symptom onset was normal. To exclude mild encephalitis, cranial MRI and electroencephalography were performed. Both examinations showed normal findings.

The patient recovered slowly and incompletely over the following weeks. The pareses were slightly improved, in particular the pareses in the distal upper extremities. The weakness of the proximal arm muscles remained almost unchanged.

Discussion

The most frequent symptoms of TBE are meningitis, meningoencephalitis or meningoencephalomyelitis. Polio-like syndromes with polyradiculitic manifestations can complicate the disease.^{3–5} These reported cases of myeloradiculitis in TBE virus infections, however, showed additional signs of meningoencephalitis or cranial nerve involvement.⁵ To the best of our knowledge, only one case with an isolated anterior horn lesion of the C3 to the Th1 level has been reported.⁶ Even in this case, cranial MRI showed two cerebral lesions.

The prognosis of these cases with polio-like symptoms seems generally to be poor, with only slow and minor recovery, and persistent severe amyotrophy and pareses.^{4,6}

We have presented the second case of a patient with an isolated myelitis caused by a TBE virus infection. In contrast with the patient described by Beer *et al*,⁶ our patient