The aetiology of flaccid paralysis in West Nile virus infection

We read with interest the recent article by Park et al., describing a syndrome of acute anterior radiculitis associated with West Nile virus (WNV) infection. Although admittedly there is still much to learn about the clinical spectrum of disease associated with WNV, we were troubled by several of the assertions raised by the article, and the conclusions drawn.

Recent evidence has suggested that the majority of patients developing acute asymmetrical weakness in the setting of WNV infection suffer from damage to spinal anterior horn cells, resulting in a poliomyelitis-like syndrome. This has been supported by electrophysiological data, and by pathology demonstrating the destruction of anterior spinal grey matter. Park et al. assert that “the mechanism of weakness associated with WNV infection continues to be unclear”, and they subsequently “propose an alternate explanation for the associated weakness”. This alternative explanation of “acute anterior radiculitis” is based on MRI findings that showed intradural lumbosacral nerve root enhancement in a patient with unilateral leg weakness. However, the authors do not describe MRI findings in the anterior spinal cord, and the MRI images provided are at the L1–L2 and L2–L3 levels, which lie caudal to the cord segments giving rise to lumbar roots; MRI of the thoracic spine is needed to adequately visualise lumbar cord segments. This is an important omission, since it is unclear whether the authors visualised the anterior lumbar cord before proposing an alternative explanation for WNV associated weakness. In addition, the authors’ contention that their case displayed ventral nerve root involvement can be challenged, since it is difficult to distinguish intradural ventral nerve root enhancement from posterior root enhancement. Furthermore, nerve root enhancement in and of itself is a relatively non-specific finding which may be seen with meningeal inflammation in general, and may not have a clinical correlate. Accordingly, the authors’ argument that the MRI findings call into question the pathophysiology of weakness, and provide evidence for an anterior radiculopathy, rather than a poliomyelitis, as the aetiology of weakness, is speculative.

It is true that definitive MRI changes have thus far been absent in many cases of WNV poliomyelitis, although clear documentation of such changes has been reported. However, neuroimaging data in patients with poliomyelitis due to wild-type poliovirus or other neurotropic viruses has not been consistently gathered, and the incidence of such MRI changes is unknown. It is likely that specific MRI changes may be missed because of variations in the stage of disease at the time of imaging. Finally, the transient reversible muscle weakness seen in this case differs clinically from that in individuals with WNV poliomyelitis-like syndrome, who develop chronic profound weakness.

We agree that WNV infection may be associated with a myriad of clinical and pathological features, and transient weakness due to anterior radiculitis certainly may be among those features. However, generalisation based on one rather atypical case, and invoked to propose an alternative explanation for the flaccid paralysis seen in WNV infection, is problematic. It is our opinion that poliomyelitis clearly has been established as the aetiology of most cases of acute, asymmetrical paralysis seen in WNV infection.

The article by Park et al. raises an important issue, however, about the utility of MRI in the diagnosis of WNV; such questions about MRI findings have been raised in WNV poliomyelitis-like syndrome and in WNV encephalitis: it may be addressed by serial MRI assessments of patients during and following acute illness.

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References

Authors’ reply

Our understanding of the West Nile Virus (WNV) infection and its neurological manifestations has rapidly expanded in recent months. The comments submitted by Dr Leis and colleagues raise concerns regarding the case we presented. The data they cite as contrary evidence to our conclusions were published after the submission of our report (accepted for publication 10 February 2003). Since that time, pathological evidence in human cases has emerged that implicates the spinal cord anterior horn cell in the pathogenesis of WNV associated flaccid paralysis. Nonetheless, the clinical, electrophysiological, and MRI findings presented in our case are still valid.

In our specific case, the patient clearly exhibited signs and symptoms of an acute lower extremity motor paralysis that was supported by electrophysiological studies. It is important to reiterate here that such studies are incapable of distinguishing whether the pathology is located in the anterior horn, ventral root, or motor axon. As such, the conclusions and pathological data that preceded our report were potentially premature in implicating only the anterior horn cell, despite the clinical presentation of a poliomyelitis-like syndrome.

The MRI study we presented showed enhancement of the ventral nerve roots. In the published image no signal change was seen within the adjacent spinal cord itself. Although the nerve roots may enhance with a non-specific meningeal process, the area of signal abnormality in our patient correlated well with the clinical and electrophysiological findings. In addition, signal change was not seen in other areas of the MRI scan. Previous MRI studies of poliovirus infection, as well as non-poliovirus infection that involved the anterior horn cell, demonstrated signal changes and enhancement in the region of the anterior horn cell within the spinal cord. As a result, we concluded that because no such enhancement was seen in the anterior horn cell region, the suspected pathogenesis in our case study may have extended from the anterior horn to include the ventral nerve root.

It has now been demonstrated by neuropathological studies that the anterior horn cell is affected in cases of flaccid paralysis caused by the WNV; however, lymphocytic infiltration of the nerve root was also seen. At this time, we conclude that perhaps the nerve root, in addition to, or independent of, the anterior horn cell, can also be involved in acute flaccid monoparesis caused by the WNV.

As our understanding of WNV infection and its neurological manifestations continues to evolve, it remains important to consider varied presentations of the disease, even if at times they appear contradictory. Unfortunately, MRI is an insensitive tool for assessing WNV infection. It is our hope that future prospective and pathological studies will continue to advance our understanding of the pathophysiological mechanisms that underlie the WNV infection.

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References

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PostScript

Spontaneous retinal venous pulsations can be present with a swollen optic disc

I read with interest the article “Spontaneous retinal venous pulsation: aetiology and signifi-
cance” by Jacks and Miller.1 Their explanation for these pulsations is essentially no dif-
ferent from that put forward by Levine in 1998.2 They then go on to discuss the clinical
importance of spontaneous retinal venous pulsations (SVPs). They refer to the finding
of Levin that the presence of SVPs is an indication of an intracranial pressure below
190 mm H2O.3 However, they conclude with-
out justification that “presence of SVPs allows the examiner to conclude that the
patient does not have optic disc swelling”.4

We cannot conclude that because an individ-
ual has SVPs there is no true disc swelling.
Shortly after their article was published, a 53
year old man was referred to us by his opt-
ometrist with “raised discs”. On examination,
he was found to have markedly elevated
intracranial pressure (ICP). Such people
cannot show SVPs, persons with mild papillo-
edema, particularly individuals with pseudo-
tumour cerebri, have significant fluctuations
in intracranial pressure (ICP). Such people
may indeed show SVPs during the time
throughout which their ICP is normal. Thus,
the decision as to whether or not an elevated
disc is truly swollen should never be made
entirely on the presence or absence of SVPs,
but on the entire clinical picture.

Author’s reply

We thank McKee and Ahad for their letter in
which they question our statement that “the
presence of spontaneous venous pulsations
(SVPs) allows the examiner to conclude that
the patient does not have optic disc swell-
ing”. Although we believe this statement to
be generally true, and agree with McCulley et
al that most discs with optic disc swelling do
not show SVPs, persons with mild papillo-
edema, particularly individuals with pseudo-
tumour cerebri, may have significant fluctuations
in intracranial pressure (ICP). Such people
may indeed show SVPs during the time
throughout which their ICP is normal. Thus,
the decision as to whether or not an elevated
disc is truly swollen should never be made
entirely on the presence or absence of SVPs,
but on the entire clinical picture.

Reference


BOOK REVIEWS

Fetal and neonatal brain injury: mechanisms, management and the risks of practice

Edited by David K Stevenson, William E Benitz, and Philip Sunshine. Published by Cambridge University Press, Cambridge, 2003, pp 886,
£140.00 (hardback). ISBN 0-521-80691-7

This book is now in its third edition and is subdivided into six parts to clearly
form a comprehensive review of the aetiology, pathogenesis, and management of the brain
injured neonate. The text covers epidemiol-
yogy; pathophysiology and pathogenesis; preg-
nancy; complications of labour and delivery; diagnosis of asphyxia; specific conditions
associated with fetal and neonatal brain injury; and assessment and management.

The editors have worked hard to “cross-
link” the basic science with the bedside needs5
and have produced a text with clear explana-
tions of the complex issues surrounding the
management of the brain injured neonate. They combine a broad vision with attention
to detail to produce a very useful text.

There is due emphasis given to current
issues, such as the role of antenatal infection
in causing cerebral injury and hypothermic
neural rescue, and also an eye to the future
and issues such as that on near infrared
imaging. There is a useful review of the
potential for neuroprotective therapy, and up
to date contributions on all the standard
issues such as long term outcome, treatment
of seizures, and drug misuse.

How does the book compare with its competitors? The standard text for most
workers is probably Jo Volpe’s magisterial
single author textbook, and in comparison to
this the new volume fares well. There is less
basic science—particularly in neuroanatomy
and cell biology—but there is a wider clinical
scope. I shall keep both volumes on my
shelves and will expect to find complimentary
information in both on any given topic.

D Edwards

Vascular cognitive impairment: preventable dementia

Edited by John V Bowler and Vladimir
Hachinski. Published by Oxford University
Press, Oxford, 2003, pp 337, £79.50 (hard-

This book is an authoritative account of vascular cognitive impairment written by a
host of international figures in the field of cerebrovascular disease. The title, Vascular
cognitive impairment, is significant. The editors
regard the more widely used term vascular
dementia as having outlived its usefulness.
The latter presupposes problems in memory,
which are not invariably present, and it
defines people relatively late in the course
of disease, preventing early diagnosis and
treatment. The editors propose adoption of
the concept of vascular cognitive impairment
and argue for a wholesale revision of current
diagnostic criteria.

Despite the title, most of the book is
dedicated to vascular dementia and its causes
and consequences. This emphasis reflects the fact that the chapters, which include the
themes of subtypes, cognitive assessment,
neuroimaging, histopathology, genetics, and
treatment, are predominantly reviews or
meta-analyses of published literature. A feature that consistently emerges is the
clinical and pathogenic heterogeneity
of vascular dementia. Moreover, the reader
becomes aware of the obscuration that arises
from treating vascular dementia as a uniform
entity. For example, whereas cognitive stu-
dies X and Y reveal better performance on
tests A and B in patients with vascular
dementia than patients with Alzheimer’s
disease, study Z shows the reverse finding.
The group data obstruct identification of
distinct profiles of impairment relevant to
individual patients. It becomes evident
why a radical overhaul of thinking about vascular
cognitive impairment is required.

My quibble is that like many multi-author
texts there is information overlap across
chapters. There are also occasional errors
overlooked at the proof reading stage.
However, in general the book provides an
useful, state of the art text on vascular
cognitive impairment. Particularly enjoyable
are the editors’ lucidly written introductory
and concluding chapters, which have a strong
personal flavour, and a sense of mission.

J Snowden