and play a role in carcinogenesis.¹⁰ The attendant liver damage contributes further to malignant liver deposit formation.

We conclude that, in patients with porphyria cutanea tarda, the search for an occult neoplasm may be of diagnostic and therapeutic value.

Final diagnosis

Porphyria cutanea tarda in a patient with Hodgkin's lymphoma.

Keywords: porphyria cutanea tarda, lymphoma, uroporphyrin decarboxylase

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A treatable cause of lymphocytic meningo-encephalitis

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A 27-year-old woman presented with a one-day history of headache, photophobia, neck stiffness and drowsiness. She had been previously well apart from minor illnesses and chicken pox aged 18 years. On examination she was pyrexial and photophobic with neck stiffness but no rash. Although drowsy she was rousable and without focal neurology. Cerebrospinal fluid (CSF) examination revealed an excess of lymphocytes, $32 \times 10^6/l$ (normal range $< 5 \times 10^6/l$) with a normal protein of 471 mg/l (100–600 mg/l), and glucose of 2.8 mmol/l (2.2–3.9 mmol/l). No bacteria were seen on Gram stain, or on subsequent culture. With treatment she steadily improved and was discharged seven days after presentation.

Questions

- 1 Name five viruses which can cause acute lymphocytic meningo-encephalitis.
- 2 What treatments are available?

Answers

QUESTION 1

The common viruses causing acute meningo-encephalitis are listed in box 1.

QUESTION 2

Aciclovir is available as treatment for herpes virus infection (10 mg/kg intravenously tid). Hyperimmune immunoglobulin can be used for varicella-zoster infection. Other treatments for individual infections include vidarabine (for herpes simplex encephalitis).

Discussion

In this patient, virus culture of CSF, throat swabs and faeces were negative. Serological studies for enterovirus, mumps and herpes simplex did not show evidence of recent infection. However acute and convalescent serology demonstrated a rise in varicella-zoster virus (VZV) complement fixation titre from 64 to 256 with VZV IgM only in the convalescent sample. This was consistent with the illness being due to VZV infection. The clear history of chicken-pox means this was reactivated VZV infection of the central nervous system presenting without the typical zoster rash.

Common viruses causing acute meningo-encephalitis

- herpes viruses: herpes simplex, varicella-zoster, Epstein-Barr
- enteroviruses: echo viruses (> 30 types), Coxsackie A and B (polio virus)
- paramyxovirus: mumps, measles
- (adenoviruses and arboviruses)

Box 1

Clinical features of meningitis and encephalitis

Meningitis

- headache
- fever
- neck stiffness
- photophobia
- irritability
- nausea

Encephalitis

- headache
- drowsiness
- confusion
- coma
- fits
- focal neurology
- personality change

Box 2

- 1 Price RW. Herpes virus infections of the nervous system. In: Wyngaarden JB, Smith LH, Jr, eds. *Cecil textbook of medicine*, 18th edn. London: WB Saunders, 1988; pp 2195–8.
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Learning points

- varicella-zoster virus is a cause of lymphocytic meningo-encephalitis
- reactivation of varicella-zoster can present without a rash
- a low threshold for the use of aciclovir in patients presenting with a lymphocytic meningo-encephalitis is to be recommended

Box 3

Reactivated VZV infection is relatively common (3.4/1000 annually¹) and in 1% of cases there is neurological involvement. This manifests as a meningo-encephalitis, or as a vasculitis in addition to the later neurological condition of post-herpetic neuralgia.² The clinical presentation is relatively non-specific, with features of headache, photophobia, neck stiffness, altered consciousness and hallucinations.² In contrast to herpes simplex encephalitis, fits and temporal lobe signs are uncommon. The zosteriform rash is the usual clue to its presence but as this case demonstrates, reactivation can occur without the rash.³ Lumbar puncture generally shows a moderate lymphocytosis with raised protein and normal or slightly reduced glucose. The electroencephalogram can show diffuse slow wave changes. Microbiological tests include detection of specific VZV antigens within the CSF although virus culture is more difficult. While not routinely available, a specific polymerase chain reaction does exist to detect virus in the CSF. Standard acute and convalescent serology will diagnose recent infection but this does not help treatment decisions during the acute illness.

The importance of considering the diagnosis on presentation is the availability of specific anti-viral therapy. Aciclovir (10 mg/kg tid iv) is of proven benefit in herpes simplex encephalitis and there exist reliable reports of benefit in VZV infection.⁴ The difficulty in setting up clinical trials means absolute proof of efficacy is not available.

The prognosis for central nervous system infection with VZV varies from excellent in pure meningeal involvement to a mortality of 25% for the most severe encephalitic forms.⁵ A low threshold for the use of aciclovir in patients presenting with a lymphocytic meningo-encephalitis is to be recommended.

Final diagnosis

Meningo-encephalitis due to reactivated varicella-zoster virus infection.

Keywords: meningo-encephalitis, varicella-zoster virus, shingles, aciclovir

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