First episode of acute demyelination of the central nervous system: Should I worry?

A 10-year old boy presents with fever (38.5°C to 39°C), mild headache, abdominal cramps and diarrhea without vomiting over the past few days. His physical examination is normal and you recommend rest, regular diet and oral rehydration solution. Two weeks later, his worried parents bring him back because he remains fatigued, cries easily, is encephalopathic (confused, disoriented), complains of numbness in his arms and has had unsteady gait for the past 24 h. You confirm these findings and admit him to hospital. A magnetic resonance imaging (MRI) brain scan shows multiple white matter lesions consistent with demyelination. A diagnosis of acute disseminated encephalomyelitis (ADEM) is confirmed. He requires treatment with corticosteroid and immunoglobulin and recovers completely. His parents now ask, "what is his risk of multiple sclerosis?"

LEARNING POINTS

- Acute demyelination in childhood is a serious event and is not as rare as previously thought.
  - Between April 1, 2004, and December 31, 2005, the CPSP study on acquired demyelination syndrome (ADS) of the central nervous system confirmed 135 cases.
- The most frequent clinical presentation of the initial episode was acute disseminated encephalomyelitis in 31% of cases, followed by optic neuritis, transverse myelitis, and monosymptomatic and polysymptomatic syndromes.
- All children with ADS are at risk for recurrent demyelination characterizing the chronic autoimmune disease multiple sclerosis (MS).
  - During 20 months of active ADS surveillance, initial preliminary results showed that almost 10% of cases (12 of 135) developed MS within two years.
  - Single-centre studies have shown that 25% of children with ADS will be diagnosed with MS within three to five years.
- After an acute event, practitioners should ensure long-term follow-up with repeat clinical examinations, and laboratory and radiological investigations. Neurological referrals are advised.
- MRI of the brain and/or spinal cord is an essential diagnostic tool in demyelination.
  - During the study period, the proportion of children undergoing brain MRI has increased from 87% to 100%.
- Over 80% of the children required treatment with corticosteroids or immunoglobulin for the demyelinating event.
- Current research into possible etiological factors of demyelination include:
  - one or more environmental agents, such as a virus, triggering the immune system to target the white matter proteins of the brain;
  - a genetic predisposition; and
  - immunological studies.
- National epidemiological data and further research that includes biomarker and comprehensive MRI studies that aim to identify children at highest risk of an MS outcome.

The Canadian Paediatric Surveillance Program (CPSP) is a project of the Canadian Paediatric Society, which undertakes the surveillance of rare diseases and conditions in children. For more information, visit our Web site at <www.cps.ca/cpsp> or <www.cps.ca/pcsp>. Accepted for publication February 22, 2007