A case of subacute sclerosing panencephalitis in a 23-year-old recent immigrant to Australia

"This case … highlights a terrible consequence of a vaccine-preventable disease"

A 23-year-old woman presented with a generalised tonic–clonic seizure on a background of 9 months of progressive neurological decline (characterised by involuntary jerks, monocular visual disturbance and reduced speech) resulting in falls, impaired ability to perform activities of daily living and urinary incontinence. Examination showed right-sided myoclonus, bilateral parkinsonism, primitive reflexes present, akinetic mutism and retinal scarring apparent on fundoscopy.

Two years previously, she had migrated to Australia from the Philippines; her family reported that her neurodevelopment was normal and that she had received routine childhood vaccinations.

The results of computed tomography and angiography of the brain were normal. Magnetic resonance imaging of the brain showed cerebral volume loss and extensive white matter changes with ill-defined subcortical T2 hyperintensities. An electroencephalogram showed non-specific focal epilepsy disorder involving frontal regions. Results of extensive investigations for autoimmune, hereditary and infective causes were unremarkable with the exception of cerebrospinal fluid (CSF) analysis, which revealed unmatched oligoclonal IgG. Enzyme immunoassay of CSF and serum (with corrected optical density values of 2.24 and 4.21, respectively) gave a strongly positive result for measles IgG. Intrathecal measles antibody production was confirmed by the concurrent absence of varicella-zoster virus IgG in the CSF, despite it being detected in the serum.

A raised CSF:serum ratio of measles antibodies, oligoclonal IgG in CSF and clinical features of progressive mental deterioration with myoclonus fulfilled Dyken's diagnostic criteria of probable subacute sclerosing panencephalitis (SSPE), outlined in the Box. Supportive measures were undertaken and isoprinosine treatment for SSPE was initiated. The patient's neurological condition stabilised but there was no improvement in her condition.

SSPE is a fatal, progressive neurodegenerative disease caused by persistent infection with an altered measles virus. Although rare, SSPE should be considered in the differential diagnosis of subacute neurological deterioration and myoclonus, especially in incompletely vaccinated patients. SSPE is rare following measles infection, with an incidence of 4–22 cases per million measles cases.[1] SSPE is rarer in adults, who account for 1–12.7% of cases.[2] There is no association with the attenuated measles vaccine and SSPE; no vaccine strains have ever been isolated from tissue specimens of patients with SSPE.[3] Vaccination unfortunately does not confer 100% protection against measles infection (and hence against developing SSPE, which may occur after subclinical measles infection). A single dose of MMR vaccine is 95% effective, and two doses are 99% effective for measles protection.

This case is important as it highlights a terrible consequence of a vaccine-preventable disease. Achieving whole-population vaccination is instrumental in preventing measles infections through the development of herd immunity. However, 96–99% of a population are required to be vaccinated to prevent sustained measles transmission. In Australia in 2012, only 91.9% of children aged 5 years had received two doses of measles vaccine.[4] Lower immunisation rates have been observed in certain areas, such as parts of the New South Wales north coast. In Australia, there were 154 confirmed measles cases reported in 2013 and 335 cases in 2014.[5] Our case highlights one of the possible consequences if measles vaccination rates are not improved.

Dyken's criteria for diagnosis of subacute sclerosing panencephalitis[1]

Criterion	Description
1. Characteristic clinical features	Progressive, subacute mental deterioration with typical signs like myoclonus
2. Electroencephalogram	Periodic, stereotyped high-voltage discharges
3. Cerebrospinal fluid (CSF)	Raised γ-globulin level or oligoclonal pattern
4. Measles antibodies	Raised titre in serum (≥1:256) and/or CSF (≥1:4) with a CSF:serum ratio <1:200
5. Brain biopsy or autopsy	Showing typical histopathology and/or culturing altered measles virus and/or detection of measles RNA by polymerase chain reaction

Definitive diagnosis: criterion number 5, plus 3 other criteria. **Probable diagnosis:** 3 or more of the 5 criteria. ◆

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