

Acute encephalopathy associated with severe fever with thrombocytopenia syndrome

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DESCRIPTION

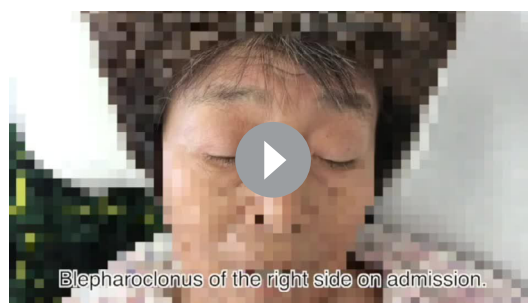
In July 2020, a 72-year-old Japanese woman with hypertension and dyslipidaemia presented with a 5-day history of fatigue and anorexia, followed by fever, slurred speech and involuntary movement of the upper limbs. She had not travelled abroad recently. She lived at the edge of a copse located in Okayama, a western prefecture of Japan and worked in the fields every day. On examination, she was disoriented (Glasgow Coma Scale, E4V3M6) and presented with postural tremor as well as myoclonus of the upper extremities ([video 1](#)) with blepharoclonus ([video 2](#)). Physical investigation did not detect any skin rashes or eschars. Her blood test results revealed the following: white cell count, 2790/ μL (normal: 3300–8600); atypical lymphocytes, 17.3%; platelet, $7.4 \times 10^4/\mu\text{L}$ ($15.8\text{--}34.8 \times 10^4$); aspartate aminotransferase, 128 U/L (13–30); creatine kinase, 1404 U/L (41–153); ferritin, 3810 ng/mL (6.2–138.0) and C reactive protein, 0.23 mg/dL (<0.15). The clinical course was suspicious of severe fever with thrombocytopenia syndrome (SFTS). Her serum sample was sent for a PCR test for SFTS virus (SFTSV), which tested positive. Although MRI of the head was unremarkable, and cerebrospinal fluid examination showed only mildly elevated protein (87 mg/dL,

normal: 10–40) with normal cell counts (2/ μL), electroencephalography revealed the presence of theta waves, indicating acute encephalopathy associated with SFTS. She was treated with high-dose methylprednisolone therapy (1000 mg/day for 3 days) for neurological complications. The next day, her neurological symptoms had improved markedly, and the slurred oral communication and involuntary eye movement ameliorated within a few days. She was discharged on the 13th hospitalisation day with only a mild residual tremor ([video 3](#)).

SFTS, a potentially fatal tickborne disease caused by SFTSV, is endemic to East Asia, including China, South Korea, Japan, Taiwan and Vietnam. SFTS can potentially cause multiple organ complications and has a mortality rate of 30%. Neurological complications are relatively common (71%) in Japanese patients with SFTS,¹ the symptoms of which include disorientation, muscle weakness, tremor, dysarthria and seizures lasting between 5 days and 2 weeks.² Imaging studies using CT and MRI usually show no abnormalities in the head, while mild abnormalities in protein, glucose and cell count can be observed during cerebrospinal fluid examination.^{3,4} Elevated levels of cytokines, such as interleukin-8 and monocyte chemoattractant protein-1, have been reported in the spinal fluid of patients, suggesting that SFTSV-associated cytokine storm may contribute to the development of encephalopathy.⁵ Although glucocorticoid may be commonly administered for virus-associated encephalopathy with the expectation of inhibiting cytokine production and function,⁶ its effectiveness is yet to be established; some literatures reporting effectiveness of the steroid therapy in acute encephalopathy^{7,8} and others not.⁹ While it has been reported that patients treated with glucocorticoid recovered without neurological sequelae,^{3,4} the neurological impairment remained in the present case. Since neurological complications are poor prognostic factors for the quality



Video 1 Disorientation, myoclonus of the upper extremities and postural tremor on admission



Video 2 Blepharoclonus of the right side on admission



Video 3 Residual mild tremor in both hands at discharge



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Images in...

of the remainder of the patient's life, the neurological findings should be noted in patients with SFTS.

Patient's perspective

I do not remember the week around the time of my admission to the hospital. I have trouble writing now because my hands are still shaky.

Learning points

- ▶ A patient with severe fever with thrombocytopenia syndrome (SFTS) can develop acute encephalopathy, thereby leading to various neurological manifestations.
- ▶ SFTS can leave neurological sequelae, and follow-up should be performed after recovery.

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