

NEUROLOGICAL PICTURE

Bilateral substantia nigra lesions on magnetic resonance imaging in a patient with encephalitis lethargica

A 21 year old man was admitted to hospital presenting with low grade fever (37.7°C), slight meningeal irritation, headache, visual hallucinations, dysarthria, insomnia, gait and limb ataxia, and a postural tremor.

There was no relevant history except for a fall into a river after a fight 2 weeks before admission. There were only a few superficial skin lesions, no medical attention was sought. The patient denied the use of recreational drugs.

Brain CT was normal. A lumbar puncture was performed showing 72 white blood cells/mm³ (84% monocytes and 16% polymorphic neutrophils), and a normal protein (34 mg/dl, normal <45 mg/dl) and glucose content (70 mg/dl, in serum 100 mg/dl). Bacterial, mycobacterial, and viral cultures were negative. Treatment with acyclovir and ceftriaxone was given and discontinued after 5 days, when the results of the cultures and polymerase chain reaction were normal.

Four days after admission, the neurological picture changed into a hypokinetic rigid syndrome with cogwheel rigidity, katatonica, akinetic mutism, dysphagia, and severe sialorrhoea and hyperhidrosis. A few oculogyric crises were noted. A clinical diagnosis of encephalitis lethargica was made, according to the criteria of Howard and Lees.¹ Brain MRI showed bilateral hyperintense signals in the substantia nigra (fig 1) and also lesions in the right striatum (fig 2) and in the right frontal lobe on MRI FLAIR images.

A treatment with levodopa/benserazide and ropinirole was started on day 4 without benefit. The patient developed spasticity and anisocoria. On day 14 treatment was started with 1 g intravenous methylprednisolone daily during 5 days with only a slight improvement of his extrapyramidal symptoms. A more sustained amelioration

occurred after a second 5 day treatment with daily intravenous 1 g methylprednisolone, started on day 23, followed by tapering.

A seroconversion was noted for Cocksackie virus type B3 (titre 0 on day 4, 1/320 on day 25). Other bacterial and viral serologies remained negative or unchanged. The substantia nigra lesions were less intense on a control brain MRI on day 50.

At the latest follow up a slight extrapyramidal syndrome persists with an expressionless face, diminished blinking, a monotonous voice, slight cogwheel rigidity, diminished arm swing, a fast, irregular tremor of arms and lips, and excessive sweating.

The current treatment consists of 200 mg levodopa/50 mg benserazide and 1 mg ropinirole thrice daily.

As far as we know, this is only the second case of bilateral substantia nigra lesions on MRI in a patient with encephalitis lethargica.²

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Figure 1 Bilateral hyperintense lesions of the substantia nigra on MRI flair image on day four.



Figure 2 Hyperintense lesion of the right striatum on MRI flair image on day four.