MELAS Associated Pathological Hyperemotionalism: A Case Report

Dear Editor,

Mitochondrial myopathy, encephalopathy, lactic acidosis and stroke (MELAS) syndrome is a progressive neurodegenerative disorder with variable clinical presentations. Neuropsychiatric manifestations include mood disturbances, obsessive compulsive features and neurocognitive deficits.[1,2] We report a case of pathological hyperemotionalism associated with MELAS syndrome which has not been documented previously in the literature, according to the best of our knowledge.

T is a 46-year-old Chinese female who was first diagnosed with MELAS in March 2003 after admission to a general hospital for stroke. She presented with left sided numbness and weakness, and prominent drooling of saliva. MRI scan showed a right middle cerebral artery territory infarct and serum lactate was elevated at 5.0 mmol/L. Muscle biopsy from skeletal muscle of right vastusmedialis showed no evidence of micronecrosis, grouped atropy or inflammatory cellular infiltration.

One month after her first stroke, she began to have persistent low mood, with poor sleep and loss of interest in activities. She also had persecutory delusions and suicidal ideation. She was diagnosed with post-stroke depression. Her mental status subsequently improved with a combination of antidepressant, antipsychotic medication and a course of electroconvulsive therapy. Physical recovery was good, and she was soon able to ambulate and function independently.

She was diagnosed with a second stroke in 2008 after she was re-admitted to the same Neurology Unit for recurrent headaches. She was not depressed for some time after her second stroke.

Prior to her recent admission to our psychiatric hospital, she had complaints of low mood sporadically for several weeks, each time lasting 1 to 2 days, and suicidal ideas, but there were no identifiable stressors. She did not have other biological symptoms of depression such as poor sleep or loss of appetite and she did not relate anxiety or compulsive symptoms.

One week into admission, she suddenly became drowsy and had difficulty walking. She was then referred to the general hospital for further investigations. MRI scan of the brain showed an acute left temporo-parieto-occipital infarct for which she was treated conservatively.

Soon later, she was transferred back to our hospital and observed for the first time to have daily episodes of loud crying which lasted approximately 30 minutes each time. These episodes were sudden, unexpected, explosive, uncontrollable and were out of proportion to any possible triggers. These crying episodes occurred daily and could happen up to 3 times a day. In between the crying episodes, she appeared calm and cheerful, did not report any depressed mood, other depressive features (such as loss of appetite, sense of guilt, hopelessness or suicidal ideas) and could not recall those episodes nor related triggers. Bedside cognitive examination did not reveal any frontal release signs or perseveration. Physically, her gait became more steady and she regained power in her upper and lower limbs.

Our impression was that of pathological hyperemotionalism in the context of her recurrent strokes and encephalopathy related to MELAS. The dosage of fluoxetine was carefully increased to 30 mg/day. The hyperemotionalism improved within a fortnight following the medication adjustment.

Up to 32% of MELAS patients have been reported to have depression.[2] However, this is the first reported case of MELAS associated pathological hyperemotionalism. Pathological hyperemotionalism is known to occur in neurological conditions such as stroke. The sudden episodes of crying are experienced by the patient as uncontrollable and not proportional to the patient's feelings. In contrast, patients with mood disorders tend to have pervasive and sustained change in their emotions and exhibit spells of laughter or crying due to underlying mood disturbance.[3] Although pathological hyperemotionalism and mood disorders are different clinical entities, they may coexist in the same patient. In T's case, the pathological hyperemotionalism started following her third stroke.

Recent studies found that a point mutation at nucleotide pair 3243 within the tRNA-Leu (UUR) gene can occur in up to 80% of patients with MELAS. Further advances in scientific knowledge could potentially enlighten us on the relationship between molecular genetics and development of pathological hyperemotionalism in MELAS.

Pathological hyperemotionalism often responds to treatment with selective serotonin reuptake inhibitors (SSRIs). Response to SSRIs can be evidenced within

days of treatment initiation or adjustment as observed in this case, and often occur with much lower dosages than those prescribed in depression.[4,5] This case highlights the need to consider possible occurrence of pathological hyperemotionalism accompanying recurrent strokes in MELAS which can be responsive to treatment.

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