Case Image

Tongue-biting ataxia that appeared to be a psychiatric disorder: a case of neuroacanthocytosis

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A 32-year-old hearing-impaired woman with epilepsy presented with loss of consciousness, seizures, and tongue biting. She was hemodynamically stable and had communication difficulties because of her hearing loss. She required endotracheal intubation to prevent airway obstruction by blood. Head computed tomography showed no atrophy of the bilateral caudate nucleus and lateral ventricular enlargement.

On day 1 after hospitalization, she was extubated and involuntary perioral movements were observed. She had a seizure on day 8. Endotracheal intubation and mechanical ventilation were performed again to prevent involuntary perioral movements and tongue biting (Fig. 1A). She wrote that she did not need her tongue, which suggested a psychiatric disorder. She had visited many hospitals over several years and was prescribed anticonvulsants; the hospitals failed to diagnose a physical illness.

However, the presence of tongue and lip deformities because of self-harm, involuntary perioral movements, truncal ataxia, a history of epilepsy, and the similar symptoms exhibited by only her brother, among relatives, implied acanthocytosis.[1]

Acanthocytes were identified on the peripheral blood smear (Fig. 1B). We identified and excluded symptomatic chorea, drug-induced chorea, metabolic disease, and other neurodegenerative diseases based on age, family history, medications used, blood tests, and imaging findings.

She was referred to a university hospital for a definitive diagnosis of neuroacanthocytosis and deep brain stimulation surgery for her involuntary movements.[2] After surgery, her involuntary movements improved, the gait stabilized, and the tongue biting disappeared.

Although psychiatric disorders may present with self-harm and suspected epilepsy, involuntary movements should prompt consideration of a hereditary disorder.

DISCLOSURE

APPROVAL OF THE Research Protocol: N/A.

Registry and the Registration No. of the Study/Trial: N/A.

Informed Consent: Written informed consent was obtained from the patient.

Animal Studies: N/A.

Conflict of Interest: None declared.

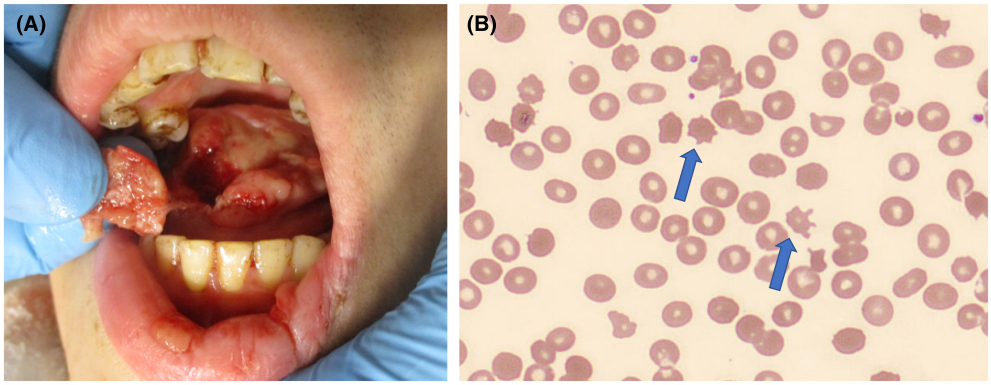

Fig. 1. (A) Tongue bite. (B) Peripheral blood smear showing acanthocytes (arrow).

REFERENCES

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2 Guehl D, Cuny E, Tison F *et al*. Deep brain pallidal stimulation for movement disorders in neuroacanthocytosis. Neurology 2007; 68: 160–1.