Teaching *NeuroImages*: Cranial nerve hypertrophy in IgG4 anti-neurofascin 155 antibody–positive polyneuropathy

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Figure Brain MRI

Symmetric hypertrophy and enhancement of trigeminal (V1, V2, and V3) and oculomotor (III) nerves on coronal (A, B) and axial (C) 3D fluid-attenuated inversion recovery acquisitions.

A 45-year-old man presented with an 11-year history of severe refractory chronic inflammatory demyelinating polyneuropathy (CIDP). Clinical examination revealed distal weakness of all 4 limbs, ataxia, facial diplegia, dysphonia, and facial sensory disturbance. Several therapeutics were tried: IV immunoglobulins (IVIg), corticosteroids, plasma exchange, cyclophosphamide, and mycophenolate mofetil. Anti-NF155 immunoglobulin G4 (IgG4) antibodies were detected by ELISA (1:2,000). Brain MRI revealed diffuse hypertrophy of multiple cranial nerves (figure).

Anti-NF155 IgG4 antibodies are associated with a younger age at onset, sensory ataxia, tremor, and a poor response to IVIg treatment.¹ Nerve hypertrophy has been reported in up to half of patients with CIDP, particularly in anti-NF155 antibody-positive patients. Hypertrophy of cervical and lumbosacral roots/plexuses has also been observed.² Neurologists should be aware that patients with CIDP and anti-NF155

antibodies may demonstrate diffuse cranial nerve hypertrophy.

AUTHOR CONTRIBUTIONS

Jérôme Franques: study concept and design, acquisition and interpretation of data. Frédérique Chapon: acquisition, analysis, and interpretation of radiologic data. Jérôme Devaux: acquisition, analysis, and interpretation of immunologic data; critical revision of manuscript. Stéphane Mathis: study concept and design, critical revision of manuscript.

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DISCLOSURE

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