Teaching NeuroImages: Dyke-Davidoff-Masson syndrome

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Figure 1 Coronal and axial MRI

Coronal and axial MRI (A–C) demonstrate asymmetric left-sided cerebral hemiatrophy and encephalomalacia with associated overlying compensatory left calvarial thickening. There is also enlargement of the left frontal sinus on coronal MRI (D).

A 50-year-old man with epilepsy from age 6 presented with seizures and personality change. Neuroimaging demonstrated left cerebral hemiatrophy, calvarial thickening, and increased frontal sinus pneumatization. There was a small, left frontal lobe hemorrhage (not shown), presumably from unwitnessed trauma. The appearances (figure 1) were compatible with Dyke-Davidoff Masson (DDM) syndrome.¹ He remains seizure-free on an increased dose of sodium valproate. His personality changes persist.

DDM results from an in utero or childhood cerebral insult resulting in cerebral atrophy with calvarial and sinus remodelling.² There was a subtle sign of limb asymmetry, unnoticed by the patient or family (figure 2).

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Figure 2 Size discrepancy between distal thumbs, possibly from early childhood neurologic insult

Author contributions

Robert Durcan: writing of case, literature review, formatting of document. Shane Smyth: critical analysis of case and description, provision of clinical photograph. Ferdia Bolster: critical analysis of radiologic findings.

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Disclosure

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