

# Teaching NeuroImages: Lacunar stroke and polyarteritis nodosa

## Consider ADA2 deficiency (DADA2)

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### Figure 1 Cutaneous feature



Livedo reticularis.

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A 14-year-old boy with a 7-year history of livedo reticularis (figure 1) and initial diagnosis of cutaneous polyarteritis nodosa presented with acute impaired taste and tactile sensation on the tongue. One year prior, he showed left partial oculomotor palsy due to a lacunar midbrain stroke (figure 2A). New imaging (figure 2, B and C) revealed multifocal vertebrobasilar ischemia. Exome sequencing demonstrated compound heterozygosity in ADA2 (an intronic and a G47R<sup>1</sup> mutation), confirming adenosine deaminase 2 deficiency, an early-onset auto-inflammatory disease characterized by features of polyarteritis nodosa and stroke.<sup>2</sup> The treatment was changed to adalimumab, whereas anti-tumor necrosis factor drugs are considered as first-line therapy.<sup>2,3</sup>

### Author contributions

Tayrine da Silva Gonçalves: study concept and design, acquisition of data, analysis or interpretation of the data. César Augusto Pinheiro Ferreira Alves: study concept and design, analysis or interpretation of the data, critical revision of manuscript for intellectual content. José Albino da Paz: study supervision, analysis or interpretation of the data, critical revision of manuscript for intellectual content. Leandro Tavares Lucato: study supervision, analysis or interpretation of the data, critical revision of manuscript for intellectual content.

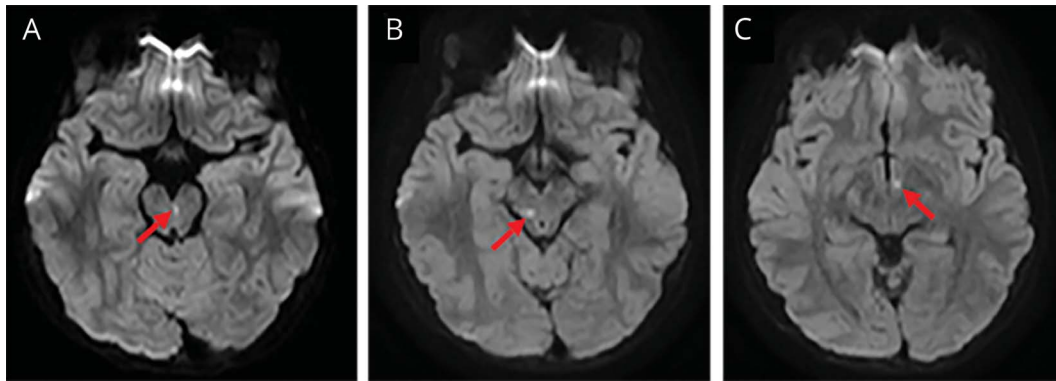
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## Figure 2 MRI findings



Very small acute ischemic foci, showing diffusion-weighted imaging hyperintensity. (A) First stroke involving left paramedian caudal midbrain. (B, C) Second stroke involving right midbrain lateral tegmentum (B) and hypothalamus left wall (C).

### Disclosure

The authors report no disclosures relevant to the manuscript.  
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### References

1. Caorsi R, Penco F, Schena F, Gattorno M. Monogenic polyarteritis: the lesson of ADA2 deficiency. *Pediatr Rheumatol* 2016;14:51.
2. Caorsi R, Penco F, Grossi A, et al. ADA2 deficiency (DADA2) as an unrecognized cause of early onset polyarteritis nodosa and stroke: a multicentre national study. *Ann Rheum Dis* 2017;76:1648–1656.
3. Meyts I, Aksentjevich I. Deficiency of adenosine deaminase 2 (DADA2): updates on the phenotype, genetics, pathogenesis, and treatment. *J Clin Immunol* 2018;38:569–578.

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