The shrimp sign in ataxic cerebellar progressive multifocal leukoencephalopathy

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An 80-year-old woman presented to the emergency department with vertigo, right hand incoordination, and postural instability five months after rituximab treatment for chronic lymphocytic leukemia. Physical examination revealed dysarthria, horizontal nystagmus, gait ataxia, and right upper and lower limb dysmetria. Brain magnetic resonance imaging showed a T₂ hyperintense (Figure, A) and T₁ hypointense (Figure, B) lesion in the right cerebellar white matter that demarcates the dentate nucleus (Figure, C), with pontine and middle cerebellar peduncle extension. This imaging finding corresponds to the 'shrimp sign': white matter lesion that preserves the curvilinear-shaped dentate nucleus giving a shrimp-like appearance.¹ HIV serological testing was negative, and JC virus DNA was detected in the cerebrospinal fluid, establishing the diagnosis of HIV-negative progressive multifocal leukoencephalopathy (PML). Rituximab was discontinued and she showed no radiological and clinical progression after one-year follow-up. The 'shrimp sign' is a reliable indicator of infratentorial PML in patients with cerebellar ataxia.¹

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Figure Legend.

Non-enhanced axial brain MRI shows a shrimp-like T2 hyperintense and T1 hypointense lesion (arrows in A and B, respectively) in the right cerebellar white matter, that demarcates the dentate nucleus (delimited with a dashed line in A and B). Dentate nucleus is best identified on susceptibility-weighted images (arrowhead in C).
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