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Alexander Disease: An Important Mimicker of Focal Brainstem Glioma-Response

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To the Editor: We were pleased to read the interesting case presented by Ring et al., and like our case, it indicates the importance of considering a diagnosis of Alexander disease in patients with particularly unusual mass lesions on MRI. While our case discussed focal posterior fossa lesions in juvenile Alexander disease, this case highlights the fact that even classic, infantile Alexander disease can give the impression of tumor-like lesions.

Despite the diagnostic issues raised by these two interesting cases, we believe that confirmatory genetic tests for Alexander disease should be undertaken judiciously; it is impractical to sequence the *GFAP* gene in every case where Rosenthal fibers are present in biopsied tissue. Rosenthal fibers are found in diverse reactive and neoplastic processes in the CNS, not just pilocytic astrocytoma and Alexander disease. Given sufficient tissue the pathologist would normally not misinterpret how Rosenthal fibers fit into a pathological process, including Alexander disease; its pathology is distinctive enough to be recognized in all but small biopsies. However, when tissue is limited and the presence of Rosenthal fibers is not be easy to reconcile with the other histopathological abnormalities or the clinical and radiological features, we would certainly recommend supplementary genetic testing for Alexander disease.

This case presented by Ring et al. again highlights the importance for neuro-oncologists to keep Alexander disease in their differential diagnosis for CNS malignancy in children, especially when the MRI findings show bilateral, symmetric lesions within the frontal white matter, deep structures of the brain or the brainstem.

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