
NEURODEGENERATION ASSOCIATED WITH GENETIC DEFECTS IN PHOSPHOLIPASE A₂

To the Editor: In their article, Gregory et al.¹ describe 2 main pathologic features of infantile neuroaxonal dystrophy and idiopathic neurodegeneration with brain iron accumulation: axonal spheroids and iron deposits. The authors describe these 2 elements as unrelated and induced by the defective phospholipase A₂.

However, I believe that the accumulated iron plays a role in spheroid formation. As the authors note, the association of spheroids and iron accumulation appears in another disease, pantothenate kinase-associated neurodegeneration (formerly Hallervorden-Spatz syndrome).² Iron-positive macrophages were described in another rare disease, hereditary diffuse leukoencephalopathy with spheroids.³ Moreover, the combination of axonal spheroids and iron deposits is seen in non-metabolic disorders, such as superficial siderosis of the CNS. In this disorder, chronic or intermittent spilling of blood into the subarachnoid space and spread of heme by the CSF causes infiltration of iron and ferritin in the cortex associated with abundant axonal spheroids.⁴ Spheroids were described in the vicinity of hemosiderin-laden macrophages surrounding arteriovenous malformations, and even in hemorrhagic infarctions.⁵ Thus, accumulation of iron alone may induce spheroid formation.

The spheroids of the hereditary diseases may differ from those of superficial siderosis of the CNS. However, the strong association of spheroids and iron in hereditary diseases and acquired disorders suggests a causative relation, and perhaps the toxic effect of iron has a role in spheroid formation.

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Editor’s Note: The authors of the article were offered the opportunity to respond but declined.

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CORRECTION

Association between late-life body mass index and dementia: The Kame Project

In the article “Association between late-life body mass index and dementia: The Kame Project” by T.F. Hughes et al. (Neurology 2009;72:1741–1746), reference 6 (Atti et al.) should not be referenced in the Introduction, page 1741, paragraph 1, sentence 2. All other uses of this reference are correct. The authors regret the error.