Posterior Reversible Encephalopathy Syndrome as the initial clinical manifestation of Neuro-Behcet's Disease: A case report./h3

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Neuro-Bechet (NBD) is a presentation of Behcet's Disease where the central nervous system (CNS) is affected; this insult is too, heterogeneous in its features. Specifically, NBD largely consists of two clinical entities affecting the CNS: the more common form of meningoenchephalitis arising from parenchymal insult, and a phenotype consisting of cerebral sinus thromboses' sequlae (non-parenchymal NBD). Among variations reported in the literature, reversible posterior cerebral venulitis has been previously reported as a potential phenotype of the non-parenchymal NBD variant. In a similar manner, Posterior Reversible Encephalopathy Syndrome (PRES) is a heterogeneous clinicoradiological entity typically comprised of a clinical symptom including headache, seizures and visual disturbances, combined with MRI findings indicative of reversible posterior leukoencehpalopathy and vasogenic edema. As PRES became increasingly recognized, atypical radiological phenotypes where also described; the unilateral and reversible diffusion restriction variants. We present here the report of a case of a 72 year old female patient with a personal history of inflammatory bowel disease and Adamantiades-Behcet's Disease (ABD) that presented a clinicoragiological syndrome in the spectrum of atypical PRES, coinciding with the complication of ABD with the non-parencymal variant of NBD. Though autoimmune disease has been proposed as an etiological factor of PRES, we are the first to our knowledge to specifically report a case of PRES in the setting of a first-onset NBD. Furthermore, we argue that in ABD, PRES may represent an intermediate, benign phenotype of NBD where endothelial dysfunction is transient, and thus the full spectrum of non-parencymal NBD may not be developed.