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An interesting case of Neuroacanthocytosis with head banging: A close differential of Huntington's Disease Like (HDL)

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N euroacanthocytosis (NA) is a rare progressive degenerative disorder of the basal ganglia associated with red blood cell acanthocytes. It includes choreoacanthocytosis (ChAc) and Macleod syndrome (MLS) as core NA syndromes. SIBs are usually manifestated by lip and tonguebiting and finger chewing but head scratching and head banging are also mentioned in literature. We report a case of ChAc who presented to us with head banging. A 17 year old male presented to us with 4 years history of self-injurious behaviour (SIB) in the form of repeated head banging to wall and other hard surfaces along with other self-mutilating behaviour like lip biting, tongue biting and chewing fingers. From past 2 years patient also developed other symptoms like abnormal involuntary movements, slurred speech with oro-mandibular dyskinetic movements with lingual dystonia during speech and food intake. He also had choreo-athetoid movements which were generalized involving both the upper and lower limbs. Peripheral blood Smear showed 18

% acanthocytes. MRI brain revealed significant caudate atrophy without cortical atrophy. He was treated symptomatically with tetrabenazine, anticholinergics and baclofen and advised regular follow up. Though Huntington's disease like (HDL-2), and pantothenase kinase- associated degeneration (PKAN), can present with similar neurological manifestations with acanthocytes, early presentations with autosomal recessive inheritance, significant self-injurious behaviour (SIB) and predominant caudate atrophy along with specific genetic testing differentiates ChAc from others.

Speaker Biography

Raghavendra Bakki Sannegowda is currently affiliated to Associate Professor, Department of Neurology, Father Muller Medical College, INDIA, continuing research in the specialized scientific area of Neurology. He is serving as an honorary author for Journal of Neurological Disorders & other reputed journals and has authored several articles along with chapters in different books related to Neurology, Neuroscience.

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