Acquired Spinocerebellar Ataxia (Celiac Disease)

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Abstract:
Spinocerebellar ataxias are inherited as autosomal dominant or recessive. But some of the ataxias are treatable and secondary to metabolic, endocrine and nutritional causes. One such disease is celiac disease with variable gastrointestinal skin and neurological manifestations. The sensitivity to gluten occurs and it may produce antibodies and malabsorption of vitamins. The antibodies may further involve in immune reactions to cause ataxia. Cases are reported since 1966 by Cooke and Smith and the incidence is 1 in 105 to 1 in 1150. (1)

Keyword: Acquired Spinocerebellar Ataxia Celiac Disease

Introduction: In celiac disease there is sensitivity to gluten in the diet which causes immune mediated Injury to the intestine resulting in diarrhea and malabsorption. Many parts of the central nervous system are affected including cerebellum, peripheral nerves, clinically manifesting in the form of cerebellar atexia, polyneuropathy, etc.

Case Vignette Mr. S, 18 years old, male, presents with complaints of unsteadiness of gait for the past 2 years. He was swaying on either side while on walking. He developed loose motion on taking wheat based diet. He had weakness in getting up from squatting with tripping of toes and buckling of knees. He also had weakness in holding the slippers and climbing the stairs. He had weakness in turning side to side on the bed he was able to raise the arm above the shoulder. He had slurring of speech and tremor of fingers. He was born to non consangunous parents and full term Hospital delivery. There was no birth asphyxia. His developmental milestones were normal. He studied upto VII std. His scholastic performance was poor. No other siblings are suffering from similar disease. Patient had dilated vessels in both bulbar conjunctivae. There was mild scoliosis and gnacomasia on both sides. He had pes cavus and alopecia. Examination of central nervous system revealed there was wasting in the small muscles of hand and forearm. There was hypotonia in both upper limbs. The power is from 4 to 4- in upper limb. There is areflexia. The finger grip is 80%. In the lower limbs.
The bulk is normal. There is hypotonia. The Celiac disease is an autoimmune dis- 
power is 4- throughout. There is a are- ease of small intestine caused by im-
flexia. The plantar reflex are not ellicit- mune reaction to gladin, a gluten protein 
able. There is horizontal nystagmus, cerebel- found in wheat. Ataxia occurs due to 
lar, dysarthria, trunkal ataxia and gait ataxia concomitant autoimmunity or malab-
with tremor of fingers. Patient had emotional sorbtion. Celiac disease has extra intesti-
blality and sleep disturbances. Other neuro-
val manifestations which are common-
logical examination were normal. Since the est like ataxia, peripheral neuropa thy patient gives a history of loose motion follow-
and other rare manifestation like en-
ing intake of wheat based diet a biopsy of D3 cephalopathy, myopathy, myelopathy, 
segment was taken for analysis. This is posi-
epilepsy, multiple sclerosis, stiffman syn-
tive for celiac disease type 3. (1) There is a drome and myoclonic ataxia. In cealic 
crease number of intra epithelial lympho-
disease the postmortem studies shows 
cytes (2) reduced hyperplasia and loss of vil-
that there is inflammation in cerebellum, 
ous structure without mucosal atrophy. (3) peripheral nervous system and other 
Cuboidal appearance of nuclei that are no parts of CNS. The following antibodies, 
longer oriented basally in surface epithelial the anti gliadin antibody, anti endomy-
cells. (4) Increased lymphocytes and plasma antibody, anti recticulin antibody and an-
cells in lamina propria. Nerve condition study tibodies to many types of transglutami-
of all 4 limbs showed distal symmetrical sen-
nase isoenzymes are positive in addition 
sory & motor axonal neuropathy. MRI brain to positive duodinal biopsy in celiac dis-
showed cerebellar atrophy. Anti Recticular ease. There is also IgA deficiency 
 Antibody : Negative Anti Endomycin Anti-
found out. The gluten sensitivity of celiac 
body : Negative Tissue transglocose Anti-
 disease is strongly heritable bygenetic 
body : Negative Antiglutamatic Anti-
load coming from : MHC Class II asso-
body : Negative Other tests like X-Ray 
ciation of HLA DQ8 and DQ2. This does 
Chest, ECG, ANA, Ant.DSDNA, TSH, FT4, not depend onimmunological trig-
Serum B12, Alfa Feto Protein, S.Copper & ger. Cerebellar ataxia is one of the com-
Cereoloplasmin, GTT, IGA, are Negative. 
In view of cerebellar ataxia, peripheral neuro-
phy, alopecia encephalopathy and GI com-
plaints related to wheat based diet and posi-
tive biopsy report to celiac disease a diagno-
sis of celiac disease manifesting as acquired 
ataxis was made.

Discussion:

With the history, examination and investiga-
tion it is clear that this patient has gluten sen-
sitivity. He had developed ataxic gait and gionopathy, small fiberneuropathy, 
weakness of both lower limbs, trunk and dis-
tal parts of upper limbs with cerebellar dy-
sarthria and tremor of hands. He is positive They are progressive in nature. Immuno-
suppressive agents are indicated with 
strict gluten free diet forganglionopathi-
es.
Other Rare Manifestations:
Gluten ENCEPHALOPATHY presents as migraine like head ache whenever a gluten diet is taken and focal neurological deficits which resolves with gluten free diet. MRI brain shows white matter abnormalities show PET brain image abnormalities and raise in transglutanate antibody titre.

EPILEPSY usually a patient is a young man who has a temporal lobe epilepsy with resistance to drugs. Associates with hippocampal sclerosis.

Myopathy, Myelopathy, Multiple Sclerosis, Myoclonic Ataxia and Stiffness Syndrome are associated with rise in various antibody titre.

Conclusion:
This case is presented for its rarity and to highlight the clinical features of an imported treatable cause of sporadic cerebellar ataxia – the possibility of celiac disease should be considered in every case.

Note: Informed consent was obtained from our patient for publication of information.

References:
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