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A Rare Case of Marchiafava Bignami Syndrome

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Abstract: ***Marchiafava-Bignami disease (MBD) is a rare Neuro degenerative disease characterized by Demyelination of corpus callosum. Clinical Diagnosis of MBD is challenging due to its Nonspecific neurological manifestations. It's promptly diagnosed by brain Magnetic Resonance Imaging (MRI). Prompt treatment with high dose Thiamine could be lifesaving. Here we report a 45 year old male with chronic alcoholism who was diagnosed to have MBD and successfully treated with high doses of intravenous thiamine Administration.***

Keywords: Marchiafava-Bignami disease (MBD), Corpus Callosum, Chronic alcoholism, Thiamine

1. Introduction

MARCHIAFAVA BIGNAMI SYNDROME (MBD) is a rare neurological disorder associated with chronic heavy alcohol consumption and malnutrition, characterized by corpus callosal degeneration and necrosis. Particularly it involves middle two-third of corpus callosal-splenium but damage may extend into neighbouring white matter and sometimes as far as subcortical regions.

2. Case History

A 45years male came to emergency dept with complain of impaired balance and unable to walk.He was having history of of lethargy, drowsiness, giddiness, gait ataxia, tingling and numbness in both feet, decreased appetite and partial memory loss. On asking further, no other past history of hypertension, DM, CVA, COPD, TB. He was having vegetarian diet, but decreased appetite. Bowel bladder and sleep was normal. He was chronic alcoholic and nicotine smoker.

3. Clinical Examination

Patient is conscious but drowsy, well oriented to time place person. He was having 76 pulse/min, 108/66mmHg, 98% sPo2 at room air. His respiratory and cardiovascular systems were unremarkable. On neurological examination:

Power = 4/5 in all limbs
Tone = slightly increased due to spacticity Planter = flexors

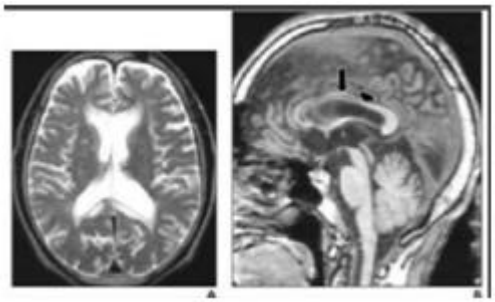
Deep tendon reflex: knees were exaggerated but ankles were sluggish. Biceps, triceps and supinator reflexs were normal.

Tremors were present.

Gait was wide base and truncal ataxic and patient was unable to walk without support. On 2nd day he became aggressive and also had seizure episode. Speech was normal but patient went in stateof confusion.

4. Investigation

Hb: 6.4 Hb, MCV-104,TC- 4500, PLT- 225000.P/S: MACROCYTIC ANEMIA
S. Creat: 0.7 mg/dlRbs: 102mg/dl
Na+: 130.4 mEq/lK+: 2.9 mEq/l
Vit B12 level: 33.45 pg/mL Urine routine was normal. MRI Brain shows as below:



Central atrophy of corpus callosum seen as SANDWICH SIGN on saggital section in MRI. (hyperintense lesion showing cytotoxic damage in spleniumof corpus callosum)

On screening spine, MRI spine wasnormal.

Diagnosis
It is a disease of radiological diagnosis. MRI brain with whole spine screening was done and it showed hyperintense area involving splenium of corpus callosum s/o cytotoxic lesion (partial degeneration of corpus callosum) and no abnormality of spinal cord was detected which confirmed the diagnosis Marchiafava Bignami

5. Treatment

Patient was treated with iv thiamine and vitB12 injections for 7 days and the symptoms were improved in form of decreased numbness and tingling sensation, patient became fully orientedto time, place and person, gait was improved and was discharged on supportive treatment of vitamin B complex, iron and folic acid and advised for follow up.

6. Discussion

Most cases have been reported in those OVER 45 YEARS OF AGE, MALES and ALCOHOLICS Most commonly it arises due to alcohol consumption which leads to thiamine deficiency and consequently corpus callosal degeneration leading to Marchiafava Bignami Syndrome. Recently ithas been found that there are 2 subtypes of the disease- Type A is characterized by major impairment of consciousness, stupor and coma predominate. On MRI it involves entire corpus callosum and sometimes associated with upper motor neuron lesion. Type B shows at most slight impairment of consciousness, partial callosal lesions on MRI and a favorable outcome

7. Conclusion

No specific treatment is available but management of alcohol related problems and intravenous injections combined with vitamin B12 and thiamine have been reported to be beneficial. It should be differentiated from 1) paraneoplastic encephalomyelitis 2) pick disease. Neurologist, psychiatrist and psychlologist are required for combined efforts for treatment had alcohol withdrawal. Along with Thiamine, vit B12 and immunosuppresents, role of Amantadine is under study.

References

[1] Harrison's 19th edition
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