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CLAUDE'S SYNDROME ASSOCIATED WITH HYPERHOMOCYSTEINEMIA

Internal Medicine

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ABSTRACT

Claude's syndrome is a distinctive brainstem syndrome characterized by ipsilateral third cranial nerve palsy with contralateral hemiataxia and is due to an intrinsic or extrinsic lesion in the midbrain. We report a case of Claude's syndrome caused by cerebrovascular accident due to Hyperhomocysteinemia. A 21year young male was admitted to our hospital with complaints of ataxia, right ptosis and diplopia. Brain magnetic resonance imaging (MRI) showed an acute non haemorrhagic infarct in midbrain on right side with hypoplastic left transverse sinus. Hyperhomocysteinemia was diagnosed by assessing the Serum Homocysteine level. The patient was treated with Intravenous Mannitol, Aspirin and Multivitamin supplements. The most common cause of Claudes's syndrome is cerebrovascular disease and malignancy. Stroke is one of the most common causes of morbidity and mortality. Hyperhomocysteinemia has emerged as an important independent risk factor.

KEYWORDS

Hyperhomocysteinemia, Claude's syndrome, ataxia.

INTRODUCTION

Hyperhomocysteinemia has a multifactorial origin incorporating genetic, nutritional, pharmacological and pathological factors [1]. Hyperhomocysteinemia is associated with increased risk of vascular diseases [2]. Homocysteine is a Sulphur containing amino acid whose metabolism stands at the intersection of two pathways: re-methylation, which requires Folate and vitamin B12; trans-sulfuration to cystathionine which requires Folate and vitamin B6 [3]. The pathways are coordinated by S- adenosyl methionine which acts as an activator of cysathionine beta synthase (CBS) and as an allosteric inhibitor of MethyleneTetrahydrofolate Reductase (MTHFR).

Meanwhile, Claude's syndrome is a distinctive brainstem syndrome characterized by ipsilateral third cranial nerve palsy with contralateral hemiataxia and is due to an intrinsic or extrinsic lesion in the midbrain.

Case Report

A 21year old young male was admitted to our hospital with complaints of diplopia, right sided ptosis and ataxia since 3 days. His previous medical history was unremarkable and he was a vegetarian by diet. The right eye pupil was 1mm larger than the left side with impaired direct light reflex. Extra ocular movements were normal in the left eye whereas limited movements in the right eye were observed and the gait was ataxic (patient was swaying on left side).

Magnetic Resonance Imaging (MRI) Brain was suggestive of an acute non-hemorrhagic infract in the Midbrain on right side (Figure 1 & 2 suggestive of areas of restricted diffusion with corresponding low ADC values was noted in midbrain on right side appearing hyperintense on FLAIR images).

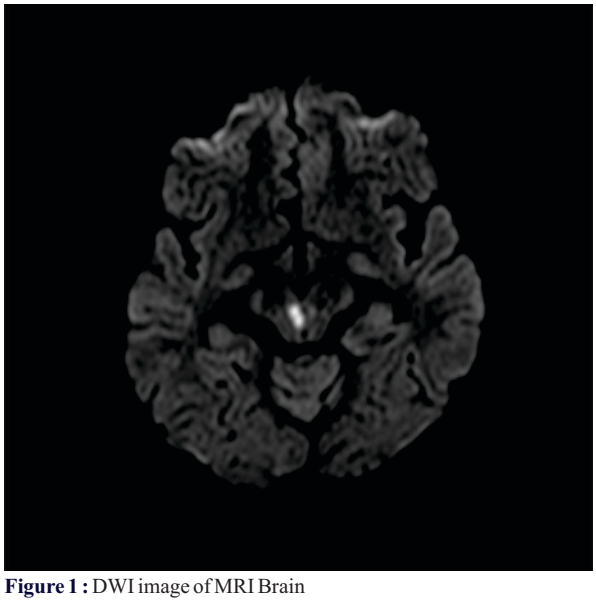

Figure 1 : DWI image of MRI Brain

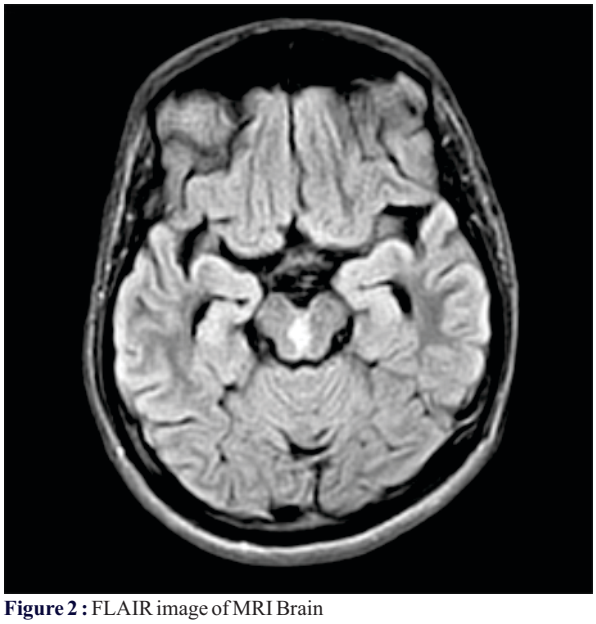

Figure 2 : FLAIR image of MRI Brain

Blood investigations revealed an elevated Homocysteine levels, whereas Lipid Profile, Hemogram, Renal and Liver Function Tests were within normal limits and 2D Echocardiogram was also normal.

The patient was treated with Intravenous Mannitol, Aspirin and multivitamin supplements. Ataxia resolved to some extent after a week of treatment but paresis of the third nerve persisted.

DISCUSSION

Patients with Hyperhomocysteinemia develop arterial thrombotic events, venous thromboembolism and more seldom premature arteriosclerosis [5]. Evidence suggest that an increased concentration of Homocysteine may result in vascular changes through several mechanisms. High levels of Homocysteine induce sustained injury of arterial endothelial cells, proliferation of arterial smooth muscle cells and enhance expression /activity of key participants in vascular inflammation, atherogenesis and vulnerability of established atherosclerotic plaques [6]. These effects are supposed to be mediated through its oxidation and concomitant production of reactive oxygen species. Other effects of Homocysteine include impaired generation and decreased bioavailability of Endothelium Derived Relaxing Factor / Nitric Oxide, oxidation of low –density lipoprotein. The effect of elevated Homocysteine appears multifactorial affecting both vascular wall structure and blood coagulation system [7].

In this case, our patient presented with ataxia, diplopia and right ptosis which was due to an acute infarct in midbrain and the search for risk factor stepped on to Hyperhomocysteinemia.

Hence increased Homocysteine level is an important risk factor for the development of ischemic stroke in all populations especially in younger age group.

CONCLUSION

Hyperhomocysteinemia is an important risk factor in Cerebrovascular accident which is commonly neglected. Meticulous approaches in identifying the most appropriate risk factor can enable us towards taking necessary precautions in preventing such diseases.

REFERENCES

1) Modi M, Prabhakar S, Majumdar S, Khullar M, Lal V, Das C P. Hyperhomocysteinemia as a risk factor for ischemic stroke: An Indian scenario. Neurol India 2005; 53:297-301
2) Arrastia RD. Homocysteine and neurologic disease. Arch Neurol 2000; 57:1422-26
3) Mennen LI, de Courcy GP, Guilland JC, Ducros V, Zarebska M, Bertrais S, Favier A, Hercberg S, Galan P. Relation between homocysteine concentrations and the consumption of different types of alcoholic beverages: the French Supplementation with Antioxidant Vitamins and Minerals Study. The American journal of clinical nutrition. 2003 Aug 1;78(2):334-8.
4) Sawhney IM, Singh G, Lekhra OP, Mathuriya SN, Parihar PS, Prabhakar S. Uncommon presentations of neurocysticercosis. J Neurol Sci. 1998; 154:94–100. [PubMed] [Google Scholar]
5) Refsum H., Ueland P. M., Nygard O., Vollset S. E. (1998). Homocysteine and cardiovascular disease. Annu. Rev. Med.49, 31–62. 10.1146/annurev.med.49.1.31 [PubMed] [CrossRef] [Google Scholar]
6) Petras M., Tatarkova Z., Kovalska M., Mokra D., Dobrota D., Lehotsky J., et al. . (2014). Hyperhomocysteinemia as a risk factor for the neuronal system disorders. J. Physiol. Pharmacol. 65, 15–23. [PubMed] [Google Scholar]
7) Boldyrev A., Bryushkova E., Mashkina A., Vladychenskaya E. (2013). Why is homocysteine toxic for the nervous and immune systems? Curr. Aging Sci. 6, 29–36. 10.2174/1874609812059990007 [PubMed] [CrossRef] [Google Scholar]