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Case Report

Fatal status migrainosus in Chiari 1 malformation

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Received : 19 November 19
Accepted : 22 November 19
Published : 13 December 19

DOI
10.25259/SNI_491_2019

Quick Response Code:

ABSTRACT

Background: Headaches are common in Chiari Type 1 malformation (CM-1). The prevalence of migraine headaches in CM-1 is similar to that of the general population. However, when migraine headaches occur with CM-1, they tend to have an earlier age of onset, are more frequent and certainly more severe than when they occur without CM-1 association. The exact role or impact of CM-1 in migraine headaches has not been fully elucidated.

Case Description: We report a fatal case of status migrainosus in 7 years old with CM-1 and review the literature on the possible associations.

Conclusion: Migraines occurring in association with CM-1 pose a management challenge and can be potentially fatal especially if associated with autonomic symptoms. The exact pathophysiological interaction between these two conditions when they occur simultaneously needs to be further elucidated.

Keywords: Chiari malformations, Fatal migraine, Migraine, Status migrainosus

INTRODUCTION

Fatalities directly due to a migrainosus attack are scarce in the literature. Furthermore, few reports have focused on the association of migraine with Chiari Type 1 malformations (CM-1) in adults and even fewer if any in children. Here, we report an unusual fatal case of 7 years old with status migrainosus (SM) and CM-1.[2,3,5,11,13]

CASE DESCRIPTION

A 7-year-old boy presented with a 3-week history of severe intractable headaches characterized by visual aura, photophobia, phonophobia, nausea, and an unusual sensation in the abdomen. Other symptoms reflected autonomic dysfunction, e.g., pallor, diaphoresis, pulse ranging from 29 to 140, variable blood pressure (70–156 systolic/diastolic 45–120 mmHg), episodic pupillary dilatation, constipation, and had two syncopal episodes. His blood counts, infectious markers, and cerebral spinal fluid studies were all normal. A magnetic resonance imaging scan of the brain revealed multiple punctate areas of restricted diffusion in both heads of the caudate and lentiform nuclei of the basal ganglia consistent with microinfarcts [Figure 1]. In addition, there was an 8 mm descent of the cerebellar tonsils below the foramen magnum consistent with the diagnosis

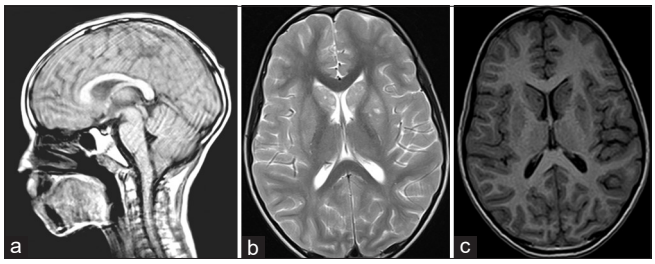

Figure 1: (a-c) From left to right; sagittal T2-weighted magnetic resonance image (MRI) scan showing caudal descent of cerebellar tonsils below the foramen magnum, second image showing axial T2-weighted MRI scan with discrete hyperintense lesions on the head of the caudate lobe and the putamen, third image showing corresponding T2 hypointense areas.

of a CM-1. Cerebral angiography and echocardiography were normal. The electrocardiogram revealed only bradycardia. The diagnosis of SM was established, and he was treated with prochlorperazine 0.15 mg/kg intravenous after preloading with normal saline; the headaches resolved within 15 min, and the child was symptom free for almost 48 h. However, subsequently, the autonomic dysfunction recurred accompanied by severe bradycardia and cardiopulmonary arrest, resulting in the patient's expiration.

DISCUSSION

According to the new International Classification of Headache Disorders 3rd edition 2018, the 7 years old discussed here had 1.2 migraine with aura headaches secondary to Chiari malformation.[13] This patient exhibited as a SM patient, defined as a debilitating migraine attack lasting more than 72 h.[7,13] Of all migraine sufferers, 23–73% exhibit autonomic dysfunction which is characterized by; bradycardia, syncope attacks, diaphoresis, constipation, and blood pressure fluctuations.[9] Basal ganglia infarcts as seen in this 7-years-old are not unusual in patients with migraines.[1,4,6] Although sudden death in an adult patient with intractable migraine and CM-1 has been described, few if any of such cases have been reported in the pediatric population.[8,10,14-16]

CONCLUSION

The combination of pediatric CM-1 with migraines can be lethal, notably more so with autonomic dysfunction.[12]

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Guest IA, Woolf AL. Fatal infarction of the brain in migraine. Br Med J 1964;1:225-6.
2. Harnod T, Lin CL, Kao CH. Survival outcome and mortality rate in patients with migraine: A population-based cohort study. J Headache Pain 2018;19:57.
3. Kaplan Y, Oksuz E. Chronic migraine associated with the Chiari Type 1 malformation. Clin Neurol Neurosurg 2008;110:818-22.
4. Kobari M, Meyer JS, Ichijo M, Imai A, Oravez WT. Hyperperfusion of cerebral cortex, thalamus and basal ganglia during spontaneously occurring migraine headaches. Headache 1989;29:282-9.
5. Kurth T, Gaziano JM, Cook NR, Bubes V, Logroscino G, Diener HC, *et al*. Migraine and risk of cardiovascular disease in men. Arch Intern Med 2007;167:795-801.
6. Maleki N, Becerra L, Nutile L, Pendse G, Brawn J, Bigal M, *et al*. Migraine attacks the Basal Ganglia. Mol Pain 2011;7:71.
7. Marcus DA. Treatment of status migrainosus. Expert Opin Pharmacother 2005;2:549-55.
8. Martinot A, Hue V, Leclerc F, Vallee L, Closset M, Pruvo JP. Sudden death revealing Chiari Type 1 malformation in two children. Intensive Care Med 1993;19:73-4.
9. Miglis MG. Migraine and autonomic dysfunction: Which Is the horse and which is the jockey? Curr Pain Headache Rep 2018;22:19.
10. Monroe DJ, Meehan JT 4th, Schandl CA. Sudden cardiac death in a young man with migraine-associated arrhythmia. J Forensic Sci 2015;60:1633-6.
11. Pascual J, Oterino A, Berciano J. Headache in Type I Chiari malformation. Neurology 1992;42:1519-21.
12. Taylor FR, Larkins MV. Headache and Chiari I malformation: Clinical presentation, diagnosis, and controversies in management. Curr Pain Headache Rep 2002;6:331-7.
13. Vincent M, Wang S. The international classification of headache disorders, 3rd edition. Cephalgia 2018;38:1-211.
14. Yeh WZ, Blizzard L, Taylor BV. What is the actual prevalence of migraine? Brain Behav 2018;8:e00950.
15. Zhang J, Shao Y, Qin Z, Liu N, Zou D, Huang P, *et al*. Sudden unexpected death due to Chiari type I malformation in a road accident case. J Forensic Sci 2013;58:540-3.
16. Ziegler DK, Mallonee W. Chiari-1 malformation, migraine, and sudden death. Headache 1999;39:38-41.

How to cite this article: Jokonya L, Makarawo S, Mduluza-Jokonya TL, Ngwende G. Fatal status migrainosus in Chiari 1 malformation. Surg Neurol Int 2019. Surg Neurol Int 2019;10:243.