

CASE REPORT

Klein Levin Syndrome and Cavum Verge: A Midline Rarity

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ABSTRACT

Klein Levin syndrome (KLS) is rare triad of episodic hypersomnolence, hyperphagia, and hypersexuality. Cavum verge (CV), although, mainly reported as a non-pathological, incidental, neuro-radiological abnormality by itself, may sometimes be associated with behavioral and psychiatric abnormalities. We here present a case of KLS with the magnetic resonance imaging (MRI) finding of Cavum verge. We then discuss the current literature of both the disorders and try to find some heuristic associations, if any, between the two.

Keywords: Cavum verge, Hypersomnia, Klein Levin syndrome.

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INTRODUCTION

First described in 19th century, KLS is rare (1 in millions)¹ triad of episodic hypersomnolence, hyperphagia, and hypersexuality.² With the advent of advanced neuroimaging procedure these highly localized, focal symptoms have been tried to be explained separately according to the neurobiological basis. CV, although, mainly reported as a non-pathological, incidental, neuro-radiological abnormality by itself, may sometimes be associated with behavioral and psychiatric abnormality.³ We here present a case of KLS with the MRI finding of CV. We then discuss the current literature of both the disorders and try to find some heuristic associations, if any, between the two.

CASE REPORT

A 16-year male presented to us with three-week duration of illness characterized by hypersomnia, low mood,

apathy, social withdrawal interspersed with hyperphagia. It started three years ago when after some fever which itself was eventful, with no document available further. Their family members noticed it first, 2 months after the fever episode when he became excessively lethargic and sleepy. In a whole day, he used to get up from the bed once only and used to start eating voraciously. On inquiry, he expressed anhedonia, hopelessness; could not interact with anyone. After 2 to 3 weeks when the episodes subsided he would become completely normal and was doing his normal activities. Gradually, the episodes had been more frequent and longer lasting, and the patient brought to our outpatient department (OPD) at his 7th episodes. No hypersexuality or elevated moods were noted, during the whole duration of illness.

On admission, he was drowsy, having very less speech output and nil eye to eye interaction. Generally, he used to sleep 16 to 20 hours a day regularly. We made a diagnosis of Klein Levin syndrome and investigated him further. All blood parameters, including thyroid function, prolactin level were within normal limits. MRI revealed thalamic infarction with CV (Fig. 1).

DISCUSSION

Earlier believed to be a disorder of hypothalamus, KLS also has been reported due to a lesion of that, as well as temporal lobe and basal ganglia. Several studies utilizing single photon emission computed tomography (CT) scanning during patients' symptomatic periods have demonstrated hypoperfusion in the

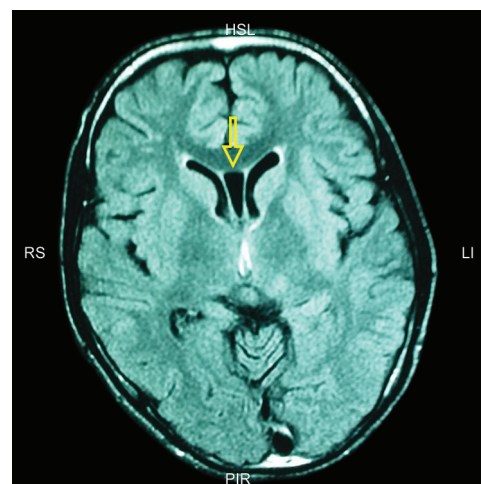


Fig. 1: Cavum verge on MRI

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thalamus, hypothalamus, temporal lobes, orbitofrontal and parasagittal frontal lobes, and, less commonly, the basal ganglia and occipital regions.⁴

The cavum septum pellucidum (CSP) is defined as a crevice-like space of variable width between the left and right transparent septum. If CSP extends posteriorly above the third ventricle and is bounded by the splenium posteriorly, it is called a CV, which is defined as a horizontal cleft between the commisura fornicis and the corpus callosum.⁵ CSP and CV are normal incident variants and are sometimes called the 5th and 6th cerebral ventricles.⁶ The size of normal variant cavum is approximately 1 to 4 mm. CSP is considered abnormally large if it is > 6 mm in size. Enlarged CSP is often seen in psychiatric patients and considered as evidence of disturbed midline development of the brain, and particularly of the limbic system.⁷

It is previously reported to be associated with neuropsychiatric symptoms like headaches,⁸ seizures, dementia, personality changes,⁹ and schizophrenia.³ Regarding our case, it can be hypothesized that being in close proximity with hypothalamus it is quite logical to be associated with such symptoms. Moreover, abnormal development of the limbic system was also made the patient vulnerable, which when undergone severe fever and viral load in his body those KLS symptoms flared up.

It is of immense importance to correlate the clinical symptoms, and when even other areas normal we should be vigilant at midline region lest we overlook this minute and rare finding.

REFERENCES

1. Ramdurg S. Kleine–Levin syndrome: etiology, diagnosis, and treatment. *Annals of Indian Academy of Neurology*. 2010 Oct;13(4):241-246.
2. American Academy of Sleep Medicine. *International Classification of Sleep Disorders: Diagnostic and Coding Manual*. 2nd ed. Westchester, IL: American Academy of Sleep Medicine; 2005.
3. Galarza M, Merlo AB, Ingrassia A, Albanese EF, Albanese AM. Cavum septum pellucidum and its increased prevalence in schizophrenia: a neuroembryological classification. *The Journal of neuropsychiatry and clinical neurosciences*. 2004 Feb;16(1):41-46.
4. Arnulf I, Lin L, Gadoth N, File J, Lecendreux M, Franco P, et al. Kleine–Levin syndrome: a systematic study of 108 patients. *Annals of Neurology: Official Journal of the American Neurological Association and the Child Neurology Society*. 2008 Apr;63(4):482-493.
5. Sarwar M. The septum pellucidum: normal and abnormal. *American Journal of Neuroradiology*. 1989 Sep 1;10(5):989-1005.
6. Macpherson P, Teasdale E. CT demonstration of a 5th ventricle—a finding to KO boxers?. *Neuroradiology*. 1988 Dec 1;30(6):506-510.
7. Flashman LA, Roth RM, Pixley HS, Cleavinger HB, McAllister TW, Vidaver R, Saykin AJ. Cavum septum pellucidum in schizophrenia: clinical and neuropsychological correlates. *Psychiatry Research: Neuroimaging*. 2007 Feb 28;154(2):147-155.
8. Chen JJ, Chen DL. Chronic daily headache in a patient with cavum septum pellucidum and cavum verge. *Ghana medical journal*. 2013;47(1):46-49.
9. Miyamori T, Miyamori K, Hasegawa T, Tokuda K, Yamamoto Y. Expanded cavum septi pellucidi and cavum vergae associated with behavioral symptoms relieved by a stereotactic procedure: case report. *Surgical neurology*. 1995 Nov 1;44(5):471-475.