ISSN: 2309-5288 (Print)
ISSN: 2309-6152 (Online)
CODEN: JCLMC4

Journal of Coastal Life Medicine

Hypertensive Encephalopathy Presenting as Cadasil

Received: 24 October 2022, **Revised:** 26 November 2022, **Accepted:** 29 December 2022

Chejarla Likhitha[1]*, N N Anand[2], A Karthik Ramalingam[3], Anand Mohan[4]

*[1]Junior resident, Department of General Medicine, Sree Balaji Medical College and Hospital, Chennai, Tamil Nadu, India. likhitha1626@gmail.com

[2] Professor & HOD, Department of General Medicine, Sree Balaji Medical College and Hospital, Chennai, Tamil Nadu, India.

[3]Assistant professor, Department of General Medicine, Sree Balaji Medical College and Hospital, Chennai, Tamil Nadu, India.

[4] Assistant professor, Department of General Medicine, Sree Balaji Medical College and Hospital, Chennai, Tamil Nadu, India.

***CORRESPONDING AUTHOR: Dr. CHEJARLA LIKHITHA**

Junior resident, Department of General Medicine, Sree Balaji Medical College and Hospital, Mobile: 9966229997
Email ID:likhitha1626@gmail.com

Keywords

CADASIL, Hypertensive Encephalopathy, ischemic, haemorrhage, mutation, dementia

Abstract

Hypertensive encephalopathy is a less often clinical scenario of hypertensive emergency, typified by severe resistant hypertension and mental status changes. Besides that, Hypertensive encephalopathy is enkindled by meagre controlled primary or secondary cause of hypertension and is difficult to pin down the etiology, which can concretely affect the treatment outcome.

Furthermore we discuss regarding a young adult male who has a meagrely controlled hypertensive presenting with head ache, vomiting and behavioural changes. On examination patient had impaired recent memory with other systems being intact. Computed tomography showed diffuse cerebral edema and Magnetic resonance showed white matter hyper intensities on T2 weighted images involving subcortical white matter and temporal lobe with relative sparing of posterior lobes which is classical for CADASIL (cerebral autosomal dominant artery disease with leukoencephalopathy and subcortical infarcts). CADASIL is inherited as an autosomal dominant trait, results from a mutation on chromosome 19p13.12 involving the NOTCH 3 gene. Besides classical radiological features suggesting CADASIL, the clinical and single gene testing did not unveil the diagnosis. Despite sometimes having high blood pressure, the blood pressure dropped throughout the second week after admission. Taking the clinical improvement into consideration his diagnosis was changed to hypertensive encephalopathy. By varying clinical appearance and exhibiting seldom evident clinicoradiological separation, hypertensive encephalopathy sets itself apart. Understanding the clinical and radiological signs of hypertensive encephalopathy will aid in guiding therapy and preventing pointless and expensive studies.

1. Introduction:

One neurologic condition that affects people with high blood pressure is hypertensive encephalopathy. A headache, seizures, visual abnormalities, altered mental state, and focal neurologic indications are some of the quickly progressing signs and symptoms that define it. (1) The condition may be deadly if it goes undiagnosed and treatment is postponed, despite the fact that it is often reversible if the hypertension is treated promptly. A diagnosis may be difficult to make, especially in people who also have other conditions, since the clinical signs are sufficiently vague. There are many neurologic diseases that might resemble hypertensive encephalopathy, including stroke, cerebral haemorrhage, venous thrombosis, and encephalitis. (2,3) Additionally, it is unknown whether significant vasospasm or forceful vasodilatation of the cerebral vasculature are responsible for the pathophysiologic alterations of hypertensive encephalopathy.(4)

ISSN: 2309-5288 (Print)
ISSN: 2309-6152 (Online)
CODEN: JCLMC4
Journal of Coastal Life Medicine

The microangiopathy of the cerebral vasculature known as "CADASIL" is hereditary in origin. In the setting of a family history of dementia and/or ischemia, transient ischemic events, headaches with aura, and gradual cognitive decline are the main symptoms. The sequencing of the NOTCH3 gene, whose mutation is to blame for the illness, is used to make the final diagnosis, but MRI plays a crucial role by highlighting the distinctive anomalies even in pre-symptomatic disease.(5,6)

The investigations were carried out to see whether radiological results may be helpful in establishing the diagnosis of hypertensive encephalopathy, in assessing the prognosis of this condition, and in contributing to our knowledge of the aetiology of the disease.(4)

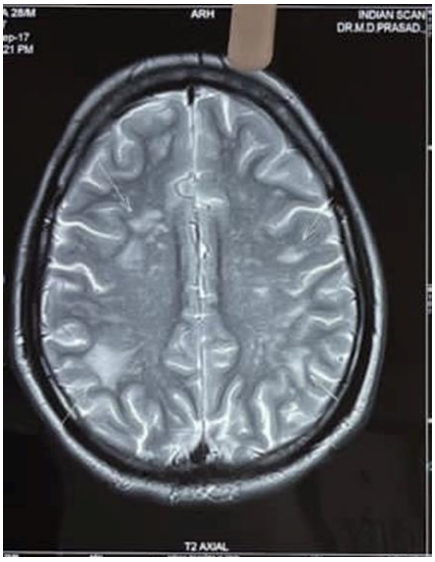
There are many ischemic lesions and a variety of symptoms depending on where the lesions are located in the brain due to this hereditary condition that affects the tiny blood arteries in the brain and causes impaired blood flow to certain regions of the brain. Episodes of migraine are often the initial clinical sign of CADASIL in young people. Recurrent subcortical ischemia events eventually cause neurological decline, frontal lobe dementia, and depressive-type mental illnesses. Spotty or nodular foci of hyposignal in T1- and hypersignal in T2-weighted sequences, including typically confluent areas within the white matter, are characteristic of the basal ganglia and brain's white matter on MRI. (7,8) These hypersignals, which are typically symmetrical and predominant in the periventricular regions and the centrum semiovale, show a more or less severe leukoencephalopathy aspect in conjunction with lacunar infarcts of the basal ganglia and brainstem, and occasionally microscopic bleeding, on gradient-echo sequences. The involvement of the prefrontal and temporal lobes stands out in particular [4].

2. Clinical Case :

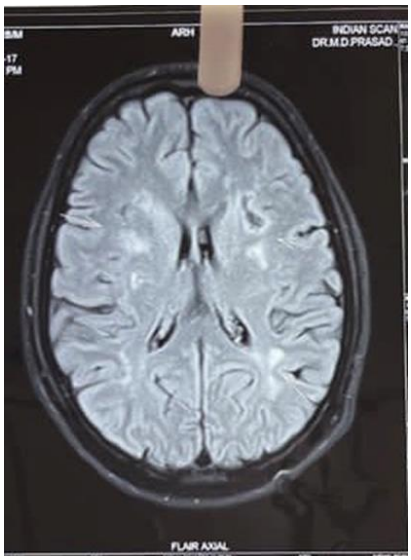
A well-built 32- year male was brought to the OPD with complaints of headache for 2 weeks, vomiting and with a change in behaviour since 2 days. He was apparently normal before two weeks, later he developed headache which is continuous, in occipital region associated with vomitings. His wife has given the history of him eating from the plate even after the food is over, brushing on only one side(hemi-negligence), difficulty identifying the objects, memory impairment, irrelevant talk, and repetition since last two days. At the age of 27(in 2017), he had an episode of deviation of mouth which lasted for few minutes. CT-Brain reports showed acute infarcts and posterior reversible encephalopathy syndrome and was also known to have hypertension for which he is on SIDDHA medication since then. he doesn't have any history of diabetes, seizure disorder, thyroid impairment. On examination, patient was conscious, lethargic, and afebrile. no features of pallor, icterus, lymphadenopathy, or edema were noted. His vital signs showed respiratory rate-28 per min, temperature-97.8 F, pulse rate-92 per minute, blood pressure-210/140 mm of mercury and oxygen saturation-98%. Central nervous system examination of higher mental function showed impaired recent memory and intact remote memory, & intact motor, sensory systems and cranial nerves. deep tendon reflexes were exaggerated with superficial plantar with-drawl bilaterally, cerebellar signs couldn't be elicited while meningeal signs were negative, and other systems were normal.

Computed tomography of brain showed diffuse cerebral edema and Magnetic resonance showed white matter hyper intensities on T2 weighted images involving subcortical white matter and temporal lobe with relative sparing of posterior lobes which is classical for CADASIL

ISSN: 2309-5288 (Print)
ISSN: 2309-6152 (Online)
CODEN: JCLMC4
Journal of Coastal Life Medicine



(a).



(b)

Figure: MRI: (a) Axial T2 - multifocal discrete areas of hyperintensities noted in bilateral fronto parietal sub cortical white matter and centrum semi ovale **(b)** axial T2 flair hyper intensities noted in bilateral gangliocapsular region and left occipital subcortical white matter with few areas of gliosis

Routine lab investigations did not show any significant abnormalities, CT renal angiogram did not unveil any abnormality. ESR, C-RP, ANA ,P-ANCA, C-ANCA were negative.C3,C4 complement levels were normal. NOTCH 3 gene test for CADASIL was done at Centre for cellular and molecular biology, Hyderabad which unveiled any mutation.

Alpha blockers, calcium channel blockers, ACE inhibitors, and other antihypertensive medications were started as part of the first course of treatment.

Even while he sometimes had elevated blood pressure, it steadily decreased over the course of the second week after admission. His diagnosis was revised to hypertensive encephalopathy taking into account the clinical improvement. When the patient was reassessed four weeks following his discharge

ISSN: 2309-5288 (Print)
ISSN: 2309-6152 (Online)
CODEN: JCLMC4
Journal of Coastal Life Medicine

on anti-hypertensive medication, he was otherwise normal except from his continuing memory impairment.(6)

3. Discussion :

16% of patients who report with hypertensive crises have HE symptoms, and the condition is characterised by a heterogeneous clinical presentation and frequent, clearly visible clinicoradiological dissociation (relatively mild symptoms in comparison to extensive imaging abnormalities). (2–4). Headache, altered mental state, convulsions, and focal neurological symptoms are among the frequent clinical indications of HE. (3) Our patient reported of repeated bouts of mild bifrontal headache upon arrival, along with a few modest neurological symptoms, including slurred speech. Initial focused neurology triggered inquiries for potential stroke right away.

Even while several imaging features pointed to a simple ischemic stroke in the context of hypertension, the presence of considerable subcortical white matter and temporal lobe hyper intensity in addition to massive vessel wall abnormalities led us to look for other, less common explanations. In hindsight, it seems that they were a medley of unusual but well-known HE characteristics, maybe with atheromatous modifications causing irregularities in the vessel walls.(5,6)

In our first differential diagnosis, we considered ADEM and cerebral vasculitis. Contrary to PRES, ADEM has a monophasic course that necessitates longer immunosuppressive therapy in order to achieve and maintain recovery. In cases with vasculitis, flare-ups may occur months or even years after the original diagnosis, particularly if the initial intravenous immunosuppressive therapy is not followed by a maintenance programme.

4. Conclusion:

It is now known that severe hypertension has a wide range of clinical and radiological brain consequences. Hypertensive encephalopathy is a condition characterised by a heterogeneous clinical presentation and frequent, clearly visible clinicoradiological dissociation (relatively mild symptoms in comparison to extensive imaging abnormalities)The white matter alterations in the posterior circulation, which are often detected in the parieto-occipital region, are the most frequently documented. (5) Vasogenic oedema favouring the posterior circulation is likely the cause of the clinical and radiological symptoms of HE, which represent a failure of cerebral autoregulation. (4)Prompt detection is crucial since these alterations are often reversible, albeit not always, at least in the early stages.(7,8).By varying clinical appearance and exhibiting seldom evident clinicoradiological separation, hypertensive encephalopathy sets itself apart. Understanding the clinical and radiological signs of hypertensive encephalopathy will aid in guiding therapy and preventing pointless and expensive studies.

References

1. Zampaglione B, Pascale C, Marchisio M, et al.. Hypertensive urgencies and emergencies. Prevalence and clinical presentation. Hypertension 1996;27:144–7. 10.1161/01.HYP.27.1.144 [PubMed] [CrossRef] [Google Scholar]
2. Karakis I, Macdonald JA, Stefanidou M, et al.. Clinical and radiological features of brainstem variant of hypertensive encephalopathy. J Vasc Interv Neurol 2009;2:172–6. [PMC free article] [PubMed] [Google Scholar]
3. Nagata M, Maeda M, Tsukahara H, et al.. Brain stem hypertensive encephalopathy evaluated by line scan diffusion-weighted imaging. Am J Neuroradiol 2004;25:803–6. [PMC free article] [PubMed] [Google Scholar]
4. Fugate JE, Rabinstein AA. Posterior reversible encephalopathy syndrome: clinical and radiological manifestations, pathophysiology, and outstanding questions. Lancet Neurol 2015;14:914–25. 10.1016/S1474-4422(15)00111-8 [PubMed] [CrossRef] [Google Scholar]
5. Hauser RA, Lacey DM, Knight MR. Hypertensive encephalopathy. magnetic resonance imaging demonstration of reversible cortical and white matter lesions. Arch Neurol 1988;45:1078–83. [PubMed] [Google Scholar]
6. Osman Y, Imam YZ, Salem K, et al.. Isolated Brainstem Involvement in a Patient with Hypertensive Encephalopathy. Case Rep Neurol

ISSN: 2309-5288 (Print)
ISSN: 2309-6152 (Online)
CODEN: JCLMC4
Journal of Coastal Life Medicine

Med 2013;2013:1–7. 10.1155/2013/540947 [PMC free article] [PubMed] [CrossRef] [Google Scholar]

7. McKinney AM, Jagadeesan BD, Truwit CL. Central-variant posterior reversible encephalopathy syndrome: brainstem or basal ganglia involvement lacking cortical or subcortical cerebral edema. Am J Roentgenol 2013;201:631–8. 10.2214/AJR.12.9677 [PubMed] [CrossRef] [Google Scholar]
8. Gamanagatti S, Subramanian S. Hypertensive encephalopathy: isolated pons involvement mimicking central pontine myelinolysis. Korean J Radiol 2006;7:218–9. 10.3348/kjr.2006.7.3.218 [PMC free article] [PubMed] [CrossRef] [Google Scholar]