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Case Report

Unusual presentation of cerebral venous thrombosis with bilateral thalamic venous infarct and Degerine–Roussy syndrome

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Roussy in 1906 in their famous paper “Le syndrome thalamique.”² It was later named as “Degerine–Roussy syndrome” or “Thalamic pain syndrome”.

Case report

A 24-year male patient went to High Altitude Area (HAA) (>9000 ft) and after about 3 months; he presented with recurrent episodes of vomiting without any fever, pain abdomen, headache or weakness. Patient was managed for dyspepsia. However he was noticed to have abnormal behaviour in the sense that he used to wince with pain on being touched at any part of his limbs with no sign of inflammation or any other sensory/motor deficit. He also had loss of memory for recent events as he could not recollect location of his workplace, location where the symptoms started. Neurological examination revealed non specific agonizing burning pain all over the body (bilateral) on touch and pressure with no focal motor or sensory deficit and there were no signs of any Upper motor neuron lesion. Cerebrospinal Fluid (CSF) analysis was done to exclude the possibility of meningoencephalitis and was found to be normal. He had recovered from dyspepsia however he persisted to have amnesia for recent events as on being asked about the place where his problem started, patient denied his stay and other routine activities in HAA. On being asked about his routine activities in HAA; he used to confabulate and tell the schedule

Introduction

Cerebral venous thrombosis (CVT) is an uncommon disorder in the general population with annual incidence of 3–4 cases per million¹ and presents with headache in 90% of cases. Dejerine–Roussy syndrome occurs in cases of thalamic infarct and commonly presents with contralateral numbness followed by burning and tingling sensations of varying severity. Thalamic pain was first described by the French neurologist Déjerine and the Swiss-French neuropathologist

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Fig. 1 – NCCT axial image: distended hyperdense straight sinus (white arrow).

which he followed before getting inducted to HAA. Patient was referred to higher centre for imaging and further evaluation.

On Computerized Tomography (CT) scan head patient had mild distension and hyper density of straight sinus and internal cerebral veins (Fig. 1). Magnetic Resonance Imaging (MRI) confirmed these findings with straight sinus, vein of Galen and internal cerebral veins revealing loss of flow void and hyper intensity on T1 weighted as well as T2 weighted/ Fluid Attenuated Inversion Recovery (FLAIR) images (Fig. 2). Abnormal signal intensity were also observed in ventromedial thalami and splenium of corpus callosum appearing hypo intense on T1 weighted images and hyper intense on T2 weighted/FLAIR images with restriction of diffusion on Diffusion Weighted Images (DWI) and corresponding Apparent Diffusion Coefficient (ADC) maps. Based on these findings impression of cerebral venous thrombosis of straight sinus,

vein of Galen and internal cerebral veins with venous infarct bilateral thalami/splenium of corpus callosum was made.

Subsequently patient was treated as a case of CVT with low molecular weight heparin, warfarin and gabapentin was added for thalamic pain relief. Pain has reduced significantly however he persists to deny any memory loss or headache in the past and is not able to recollect the date and place of onset of his illness (Fig. 3).

Discussion

CVT is a difficult diagnosis to make in a peripheral setup without imaging facilities however our patient presented with recurrent vomiting and abnormal behaviour (as perceived and described on initial clinical evaluation) without headache, which is the most common presenting complaint of CVT. The absence of signs of upper motor neuron lesion, focal neurological deficit or features of meningism with normal CSF analysis made the possibility of any organic lesion less likely. Loss of memory of recent events made it difficult to elicit history about chief complaints and exact mode of onset of complaints. The finding of agonizing burning type of pain on pressure bilaterally all over the body in absence of fever, features of inflammation and not taking meals on time made the possibility of functional behavioural abnormality as the only probable explanation left for his presentation till the time neuroimaging revealed CVT with bilateral thalamic infarct. Burning pain on touching and pressing is referred to as thalamic pain and is best known component of Dagerine-Roussy syndrome.^{3,4} Recent memory deficit, amnesia and confabulation as seen in this case is most consistent with bilateral anterior and medial thalamic lesions and may be associated with unilateral lesions.^{5,6} In this case presence of bilateral venous thalamic infarct on MRI is very unusual and peculiar finding explaining most of his presenting clinical features which initially appeared to be subjective and functional.

Important differential for deep venous thrombosis leading to thalamic infarct is artery of Percheron infarct occurring due

Fig. 2 – MRI axial images: absent flow void and hyperintensity of straight sinus (white arrow) and internal cerebral veins (white open arrow). Bilateral ventromedial thalamic infarct (white star) appearing hypointense on T1 weighted image and hyperintense on T2 weighted and FLAIR images.

Fig. 3 – DWI images (a) with corresponding ADC maps (b) showing restriction of diffusion in bilateral thalami (white arrow) and splenium of corpus callosum (white open arrow) suggestive of infarct.

to occlusion of variant P1 common trunk (Percheron) with bilateral supply. Imaging in this condition will reveal normal flow in the deep veins with infarcts in bilateral paramedian thalamic and/or midbrain.⁷

Conclusion

In this case, the unusual coincidence of isolated bilateral thalamic infarct and no other parenchymal involvement led to atypical ambiguous clinical presentation of anterograde amnesia and bilateral thalamic pain with no focal deficits on neurological examination. CVT may affect people living in High altitude areas and should always be considered and evaluated in any case presenting with abnormal behaviour even in absence of usual symptoms like headache, seizures, features of raised intracranial tension or neurological deficits.

Conflicts of interest

The authors have none to declare.

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