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WERNEKINCK COMMISSURE SYNDROME -A RARE CAUSE OF ACUTE ATAXIA

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ABSTRACT Common causes of acute onset ataxia are posterior circulation strokes ,demyelinating ,infectious and toxin induced. When stroke is the etiology we expect bilateral cerebellar hemispheric infarcts to be the cause .However a rare syndrome involving infarction of the paramedian caudal midbrain resulting bilateral ataxia along with INO and a delayed palatal myoclonus has been described with the synonym Wernekinck's commissural syndrome .It results from the occlusion of the interpeduncular perforator branches which originate from a common trunk supplied by multiple arteries. We describe the third case of Wernekinck commissure syndrome from India.

KEYWORDS : neurology, neuropathology, ischemic stroke, tegmentum mesencephali, mesencephalon, movement disorders, stroke, rare diseases

INTRODUCTION

A 43 year old female presented with complaints of sudden onset giddiness ,imbalance while walking and double vision 15 days back .She went to a nearby hospital and was given symptomatic treatment for the same .The diplopia improved within a few days but gait imbalance worsened .On examination her higher mental functions were normal except for cerebellar dysarthria .She had restricted vertical up gaze with gaze evoked nystagmus and horizontal gaze evoked nystagmus without any restriction of eye movements .Pupils were equally reactive to both direct and indirect light .Accommodation reflex was also normal .Other cranial nerves including 9,10,11,12 were normal .Motor and sensory examination was normal .Testing of coordination and cerebellar signs revealed severe stance and gait ataxia ,bilateral disdiadochokinesia. Rebound phenomenon was negative. She was provisionally diagnosed as a probable midbrain stroke in view of vertical nystagmus and bilateral cerebellar ataxia.

MRI brain with MRA showed a small area of diffusion restriction in the caudal paramedian midbrain ventral to aqueduct .MRA was normal .She was also found to be diabetic with a HBA1C of 9 with mildly elevated sugars.CV doppler and 2D ECHO were normal .

She was diagnosed as a case of acute cerebrovascular event ,midbrain infarct with wernekinck's commissure syndrome based on the clinical and radiology findings. She was started on antiplatelets and statin and received gait training and speech therapy along with optimization of sugars .She slowly improved and was walking independently at the time of discharge with only mild residual ataxia and mild dysarthria

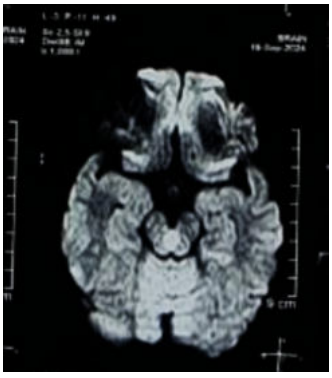

Figure 1: Diffusion weighted MRI image of caudal midbrain showing an oval shaped infarct .

Sources: department of radiology, Dhanalakshmi srinivasan

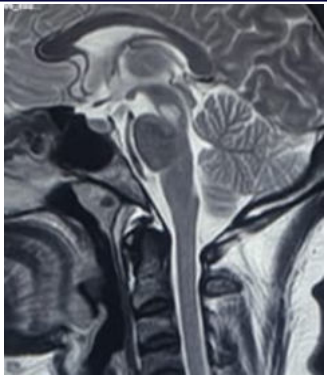

Figure 2 sagittal view showing caudal midbrain lesion . Sources: department of radiology ,Dhanalakshmi Sreenivasan

DISCUSSION :

Causes of acute onset bilateral ataxia include infarcts ,demyelination and infective cerebellitis and very commonly toxin induced .Posterior circulation strokes are a common cause of ataxia However, midbrain infarcts are very rare (0.7-2.3) [1]owing to their unique blood supply .The vascular territories are conventionally divided into the following

(a)Anteromedial or paramedian
(b)Anterolateral (c)Lateral (d)Dorsal .

Anteromedial territory infarcts are the ones associated with caudal midbrain syndromes and the region receives its blood supply from interpeduncular perforator branches especially the inferior paramedian mesencephalic arteries [IPMA][2]. They come from the last 5 mm of the basilar artery, the initial 7 mm of the two Superior Cerebellar Artery (SCA) and the initial segment (segment P1) of the Posterior Cerebral Artery (PCA) .The IPMA's supply the paramedian midbrain tegmentum and have intracerebral and extracerebral components ,which determines the radiological presentation of the infarct. Bilateral infarcts are very rare and result from occlusion of a variant where both IPMA's originate from a common trunk. The Wernekinck's commissure is a part of caudal midbrain that also consists of the MLF ,trochlear nerve nucleus ,the central tegmental tract and the reticular formation. It contains two tracts

a) The crossed ascending dentatorubrothalamic tract –[superior cerebellar peduncle]
b) Crossed descending dentatorubro olivary tract [central tegmental tract] .
Hence ,the syndrome includes three core features

1. Bilateral cerebellar dysfunction-dentatorubrothalamic pathways
2. Eye movement abnormalities -internuclear ophthalmoplegia ,due to involvement of the MLF
3. Delayed palatal myoclonus and tremor -secondary olivary

hypertrophy due to damage to dentatoolivary fibers leading to delayed transsynaptic degeneration of bilateral inferior olivary nuclei [3]

Our patient had the first two classical symptoms of which the INO resolved by third day of admission .She did not have palatal myoclonus or Holmes tremor ,however these symptoms are known to occur after a latent period ranging from months to years [4].

Only two other cases have been reported from India till date . The first case was diagnosed in a patient with preexisting movement disorder with only bilateral ataxia without eye movement abnormalities[5] ;whereas the second case was a classical syndrome with left caudal midbrain infarct [6].However both these were unilateral infarcts as opposed to our case which had bilateral oval shaped infarct .

A large number of tracts and nuclei are arranged in a close-knit fashion in the brainstem leading to small insults resulting in severe symptoms. Bilateral cerebellar ataxia usually localizes to cerebellar hemisphere ,but it is interesting to see how a small infarct in the paramedian brainstem can produce the same syndrome .Presence of oculomotor signs suggestive of INO clinically localizes this lesion to the midbrain.

CONCLUSION

Common syndromes are common .However improved knowledge of uncommon presentations do help us arrive at a clinical-radiological correlation enabling us to predict the course of the disease better .

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