

Case Report

Reverse 'Hot Cross Bun' Sign in Primary Progressive Aphasia - An Atypical MRI Feature

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ABSTRACT

Primary progressive aphasia (PPA) is thought to be a disorder of focal cortical degeneration which occurs as a result of lobar atrophy of dominant frontal and temporal lobe. We report a case of a 78 year old male patient presenting with progressive language affection predominantly and clinically diagnosed as PPA but magnetic resonance imaging (MRI) brain showed an unusual finding of reverse 'hot cross bun' sign in pons in T2 weighted (T2W) / diffusion weighted image (DWI)/ T2 fluid attenuated inversion recovery (FLAIR) axial views. This is the first case report of reverse 'hot cross bun' sign in a case of PPA to the best of our knowledge.

Keywords: Primary progressive aphasia, reverse 'hot cross bun' sign, 'hot cross bun' sign.

Introduction

Primary progressive aphasia (PPA) is thought to be a disorder of focal cortical degeneration which occurs as a result of lobar atrophy of dominant frontal and temporal lobe. PPA is divided into three variants on the basis of clinical, brain imaging and histopathological evidence. Left perisylvian atrophy is the hallmark radiologic feature. Our patient presented with progressive language affection predominantly and clinically diagnosed as PPA but MRI brain showed an unusual finding of reverse 'hot cross bun' sign. To the best of our knowledge this sign has never been reported earlier in PPA.

Case Presentation

A 78 year old right handed, retired school teacher, hypertensive male presented with one

and half year history of insidious onset, progressive speech difficulty in form of effortful, non-fluent speech with intermittent pauses. He had grammatical errors in spontaneous speech as well as in writing. Comprehension for complex sentences was impaired but preserved for a single word. Naming and repetition were mildly affected. Fund of information and word knowledge were normal. Conversation discourse was moderately impaired. However, grammatical and phonological errors were more common in writing than in spontaneous speech. He had problems with syntax comprehension. Naming and repetition were mildly disturbed with preserved word comprehension and object knowledge. His past history was unremarkable except for hypertension which was well controlled with medications. He was independent for his daily routine activities.

The general and systemic examination

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was unremarkable. On higher mental function testing, his Mini-Mental State Examination (MMSE) was 24/30 and Frontal Assessment Battery (FAB) was 8/18 suggestive of frontal lobe dysfunction. Language function showed intact comprehension along with preserved reading ability with decreased speech output, grammatical errors in writing and perseveration. There was severe word finding difficulty, impaired fluency with wrong usage of grammar intermittently. A diagnosis of primary progressive aphasia was made based on clinical history, neuropsychological and detailed language function testing. His routine investigations including hematological and metabolic profile were normal. Serum TSH and vitamin-B12 were normal.

CT Head and MRI brain with diffusion weighted image (DWI) with contrast was done. There was no evidence of acute/sub-acute or chronic infarct.



Fig. 1: MRI of the brain, T2-weighted axial section, showing presence of reverse 'hot cross bun' sign at mid pons.

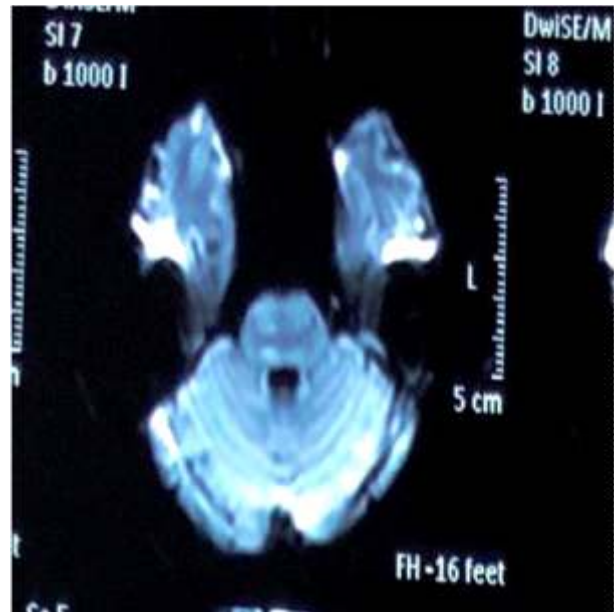


Fig. 2: MRI of the brain, DWI axial section, showing presence of reverse 'hot cross bun' sign at mid pons.

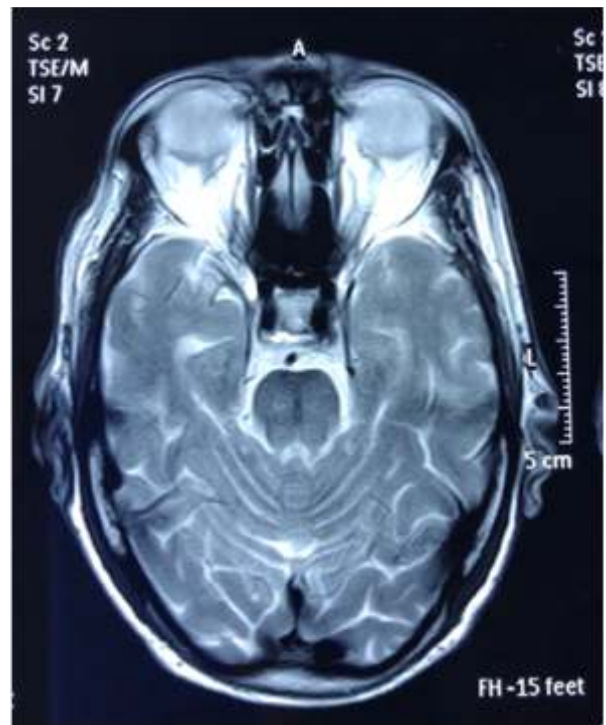


Fig. 3: MRI of the brain, FLAIR axial section, showing presence of reverse 'hot cross bun' sign at mid pons.

MRI brain showed diffuse cerebral atrophy. There was an interesting finding of reverse 'hot cross bun' sign in pons in T2 weighted (T2W) (Fig. 1)/DWI (Fig. 2)/T2 fluid attenuated inversion recovery (FLAIR) (Fig. 3) axial views. Speech therapy was instituted and he was managed with antihypertensive drugs.

Discussion

Primary progressive non-fluent aphasia is a progressive language disorder associated with atrophy of the frontal and temporal regions of dominant hemisphere (1). Primary affliction is in the language domain which differentiates it from the aphasic form of Alzheimer disease. It is important to differentiate these two entities as the former carries a better prognosis. PPA has been diagnosed and divided into different subtype according to Mesulam PPA diagnostic criteria (2). The main components of language involved are spontaneous speech output, anomia, fluency, and grammar. Other domains are relatively unaffected and the patient remains independent for his activities of daily living. There can be a subsequent progression into dementia over a decade. Early stages of the disease may not reveal any abnormal neuroimaging features but left perisylvian atrophy has been described as a hallmark feature.

Our case presented with progressive predominant language affection and we found a unique reverse 'hot cross bun' sign in MRI brain. 'Hot cross bun' sign is a cruciform-shaped T2W hyperintensity through the middle of pons due to degeneration of the cerebellopontine and transverse pontine fibers. Reverse 'hot cross bun' sign is hypointensity in the intersecting lines and hyperintensity in the quadrangles forming a shape of a hot cross bun, which can be best appreciated on T2 and FLAIR image. 'Hot cross bun' sign is seen in multiple system atrophy–cerebellar type, spino-cerebellar ataxia (SCA 1/ 2), secondary Parkinson disease, cerebrotendinous xanthoma (CTX), progressive multifocal leucoencephalopathy (PML) in HIV

seropositive patient and also in neurosarcoidosis (3).

Reverse 'hot cross bun' sign has been described earlier in a patient with pontine infarct (4) and Wilson disease (5) but not in PPA so far.

Conclusion

Our case presented with progressive predominant language affection and we found a unique reverse 'hot cross bun' sign in pons in T2W/DWI/T2 FLAIR axial views of MRI brain. This is the first case report of reverse 'hot cross bun' sign in a case of PPA to the best of our knowledge. The treatment of PPA remains conservative with speech therapy as the mainstay. The prognosis is said to be better in PPA when compared to other neurodegenerative disorders like Alzheimer disease.

Acknowledgment

We have not received substantial contributions from non-contributors and no contributor has been omitted.

Conflict of Interest

We have no conflict of interest to declare.

Source of Funding

None.

Ethical Clearance

Written informed consent was obtained from the patient and relatives of patient for publication of this case, reports and any accompanying images. We are ensuring that, this study manuscript has not been submitted and published elsewhere.

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