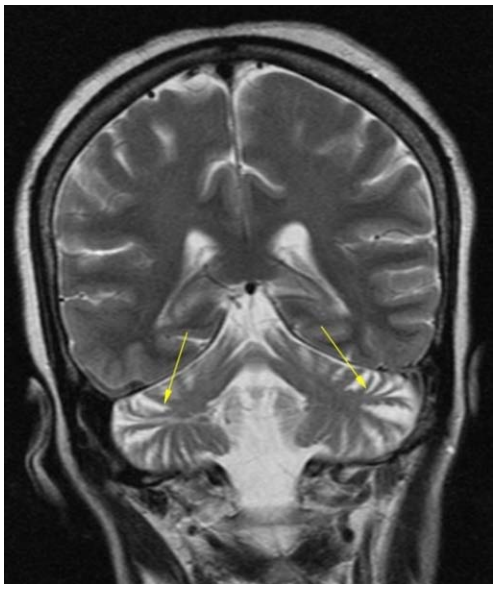
Clinical history:

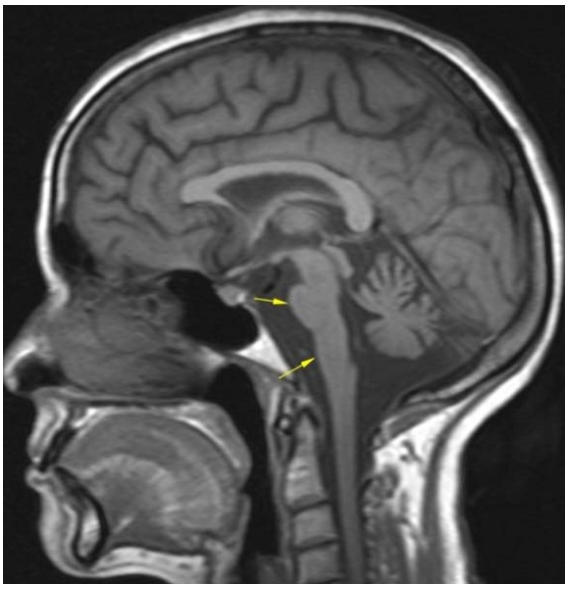
A 50-years old female presented with features of ataxic gait and imbalance.
No history of seizures or hypertension.
No history of any long term drug intake.
MRI Brain was suggested for further evaluation.

Imaging findings on MRI:

- Diffuse cerebellar and pontine atrophy with relative sparing of the supratentorial structures.
- Prominent fourth ventricle , due to the cerebellar atrophy.
- Cruciform high signal intensity on T2W images and FLAIR sequence in the pons consistent with “Hot Cross Bun” sign - reflects degeneration of transverse pontocerebellar fibres.
- Severe thinning of both middle cerebellar peduncles which also showed increased signal intensity on T2WI.



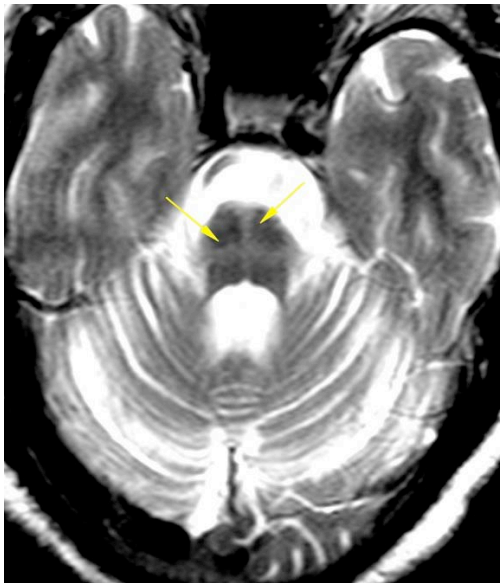
T2Wi coronal: severe atrophy of cerebellum, with relative sparing of supratentorial structures.



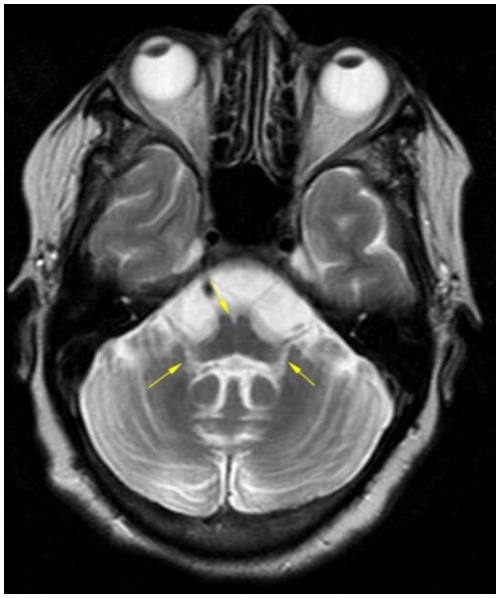
T1Wi sagittal : atrophy of pons and olivary eminences of medulla

Final Diagnosis:

Olivopontocerebellar Atrophy.



T2Wi axial: Cruciform hyperintensity –'Hot cross bun sign' in pons due to demyelination of transverse pontocerebellar fibres.



T2Wi axial: Atrophy of the medulla and middle cerebellar peduncles with high signal intensity in both pedeuncles.

Discussion:

Olivopontocerebellar atrophy (OPCA) is a term used to define neuronal degeneration which specifically involves the cerebellum, pons and inferior olivary nucleus. The comprehensive term of OPCA includes several neurodegenerative syndromes including the commoner sporadic forms and relatively uncommon familial (genetic) forms. Amongst the sporadic OPCA, a significant fraction of cases are a subset of multiple system atrophy (MSA). MSA encompasses the disorders striatonigral degeneration, olivoponto cerebellar atrophy (OPCA) and the Shy-Drager syndrome. MSA is considered synonymous with striatonigral degeneration when parkinsonian symptoms predominates (MSA-p), with olivoponto cerebellar atrophy (OPCA) when cerebellar signs predominate (MSA-c), and with Shy-Drager syndrome when autonomic symptoms are dominant. Inherited OPCA encompass a wide variety of ataxias including hereditary spinocerebellar ataxia, Machado-Joseph disease and autosomal dominant cerebellar atrophy. Familial cases usually manifest by second to third decades while the sporadic cases present in fifth to sixth decades of life.

Patients frequently complain of cerebellar symptoms such as abnormal gait, ataxia, nystagmus or cerebellar dysarthria. Extrapyramidal signs, parkinsonism, dementia, ophthalmoplegia and retinopathy may be present.
MRI is considered the imaging study of choice. MRI characteristically demonstrates atrophy of the cerebellum and brainstem with relative sparing of the supratentorial cerebral parenchyma. Sagittal MRI is especially useful as it distinctly shows atrophy of the olivary eminences of the medulla oblongata. This results in flattening of the ventral brainstem secondary to obliteration of the angle

usually present on the ventral frontier between the pons and medulla oblongata. The fourth ventricle and cerebellopontine angle cisterns become prominent. There may be associated demyelination of the transverse pontocerebellar fibers.

Demyelination of the transverse pontocerebellar fibers is responsible for the "hot cross bun" sign, which is considered to be highly specific for multiple system atrophy. Higher imaging modalities like Positron emission tomography (PET) scan shows reduced metabolism in the brain stem and cerebellum.

The OPCAs are progressive neurodegenerative disorders that have no definitive treatment. Eventually, many patients become wheelchair bound. Severe dysarthria, anarthria, and dysphagia are not uncommon as the disease progresses.

- Morbidity increases significantly, including falls and aspiration pneumonia and enteral feeding becomes necessary for many patients.
- Death commonly results from aspiration pneumonia.
- The duration of familial OPCA is approximately 15 years. The duration of sporadic OPCA is approximately 6 years.

Differentials:

- Cortical Basal Ganglionic Degeneration.
- Friedreich's Ataxia.
- Paraneoplastic Cerebellar Degeneration- Ovarian cancer is one of the malignancies associated with this syndrome, and the paraneoplastic syndrome may manifest in the early and curable stage of cancer. Anti-Purkinje cell antibodies are the diagnostic marker for this entity.Small cell cancer of the lung is also associated with this syndrome.
- Parkinson Disease.
- Progressive Supranuclear Palsy.

Message:

- **MRI is the imaging study of choice in patients with olivopontocerebellar atrophy (OPCA) because CT scanning does not provide adequate resolution of the pons and cerebellum.**
- **Brain MRI is also useful in patients presenting with spinocerebellar syndromes to exclude other diagnoses like multiple sclerosis, cerebrovascular disease and malignancy.**
- **"Hot cross bun sign" is sensitive for OPCA in an appropriate clinical settings.**

Regards,

Dr.Deepa S.Nadkarni / Dr.Shaikh M.Mazhar

N.B: This case is authentic and from the archives of Radiance Diagnostics. For any queries / suggestions/ feedback write to us at radiance@radiancediagnostics.in