Parkinsonism and its types

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Primary parkinsonism

- Most patients (about 80–85%) diagnosed with Parkinson's disease have what is called ***primary parkinsonism or idiopathic Parkinson's disease*** (meaning that the disease ***has no known cause***).

- ***This type tends to respond well to drugs that work by increasing or substituting dopamine molecules in the brain.***

Secondary parkinsonism
(parkinsonian syndrome or atypical parkinsonism)

- a key difference is that patients with secondary parkinsonism ***do not respond well to dopaminergic medications such as levodopa.***

- **Includes:**

1. **drug-induced parkinsonism**
2. **vascular parkinsonism**
3. **normal pressure hydrocephalus (NSA),**
4. **corticobasal degeneration (CBD)**
5. **progressive supranuclear palsy (PSP)**
6. **multiple system atrophy (MSA).**

Parkinson disease

- ***by far the most common cause of the parkinsonian syndrome***, **accounting for approximately *80% of cases***

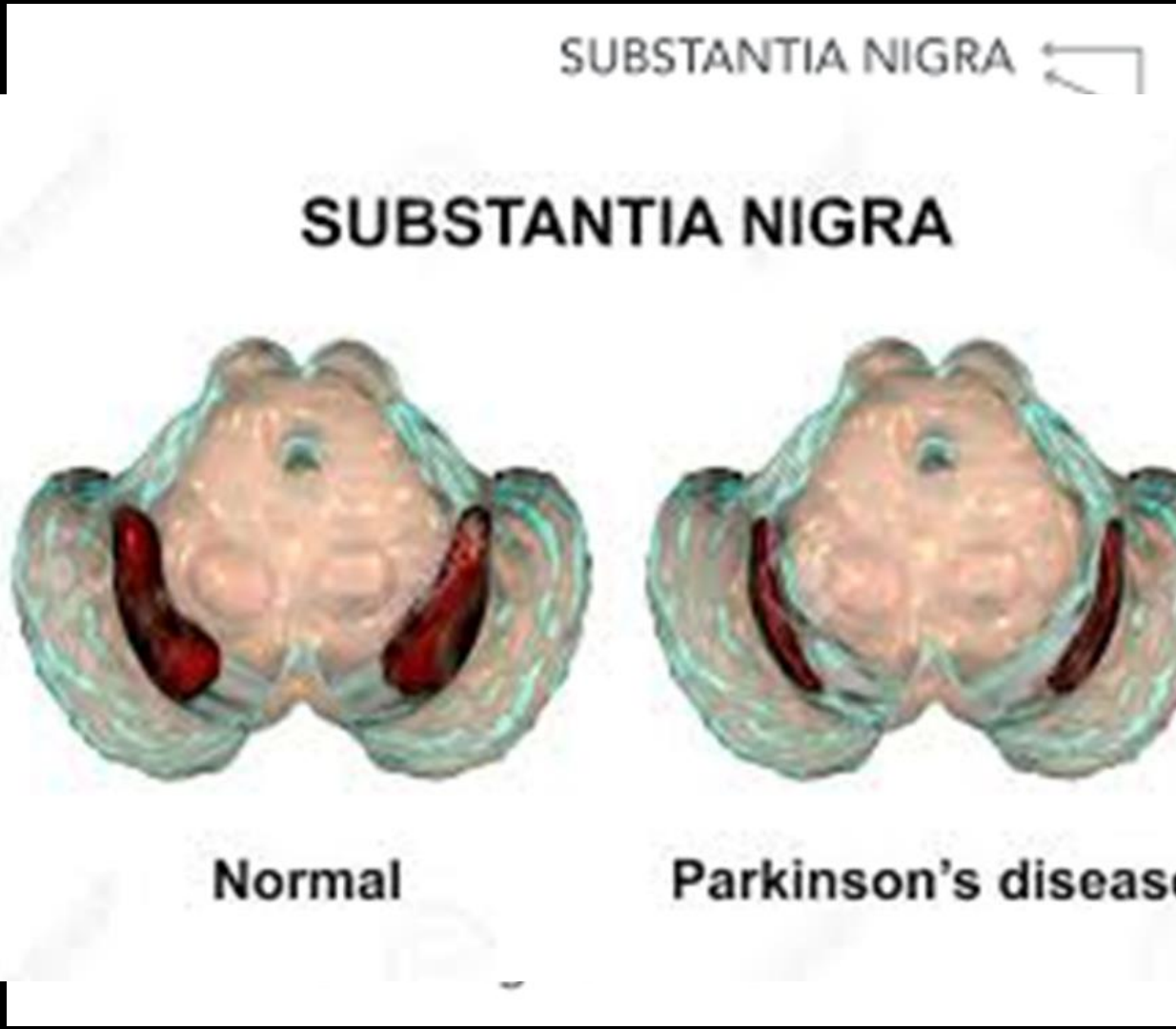
- **In most cases the diagnosis of probable PD can be made on clinical grounds, *and no ancillary investigations are needed.***
- **However, in early PD the *full triad of clinical symptoms and signs (bradykinesia, tremor at rest and rigidity)* may not yet be manifested**

- **The lesion in PD has been localized to the *dopaminergic cells of the pars compacta of the substantia nigra***

- **definitive diagnosis of idiopathic PD, requires *histologic demonstration of intraneuronal Lewy body inclusions* in the substantia nigra (SN) pars compacta.**

- **Substancia nigra is located at midbrain and composed from pars compacta and retinacula**

- **pars compacta is formed by dopaminergic neurons and located medial to pars reticulata (between this part and red nucleus)**
- ***Parkinson's disease is characterized by the death of dopaminergic neurons in this region.***



Imaging findings:

- **CT scans** ***can show nonspecific atrophy with enlarged ventricles and sulci***.

- **Conventional MRI at 1.5 T with routine T2- and T1W imaging *does not reveal disease-specific abnormalities in PD* and, *particularly in the early phases, the MRI appears normal***
- **Its main clinical role is to exclude *1.*subcortical vascular pathology, *2.*rare secondary causes of parkinsonism (e.g., Wilson's disease, NPH, or tumors, granulomas, or calcification of basal ganglia), and *3.*in discriminating atypical parkinsonian syndromes.**

T1W findings:

- **may show loss of normal slight hyperintensity in substantia nigra *due to loss of neuromelanin***

- ***may show mild hyperintensity of compact and reticular parts of the substantia nigra and red nuclei (due to iron accumulation)***

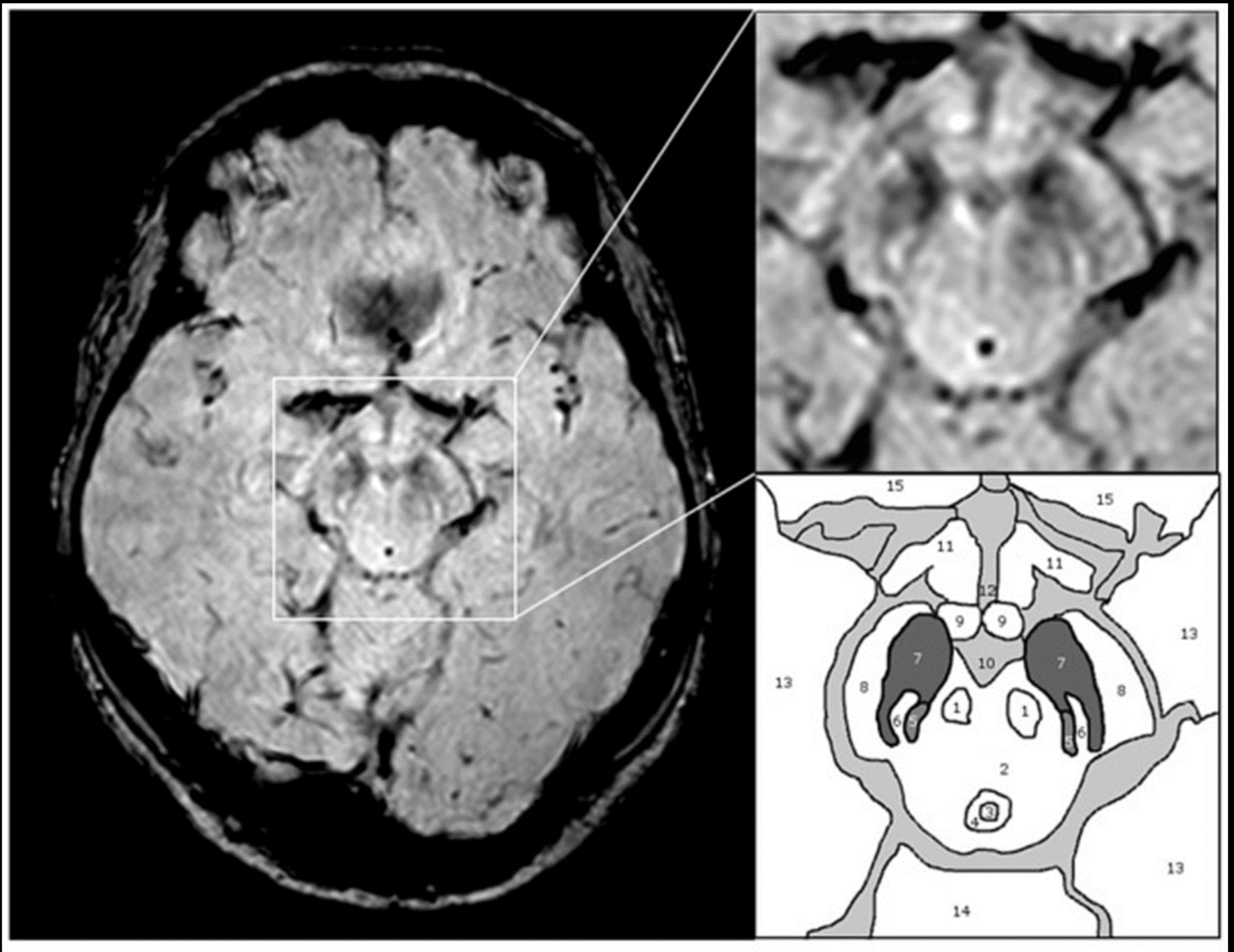
T2*-GRE and SWI findings:

- **swallow tail sign** describes the normal axial imaging appearance of nigrosome-1 within the substantia nigra on high-resolution T2*/SWI weighted MRI.

- **Loss of the normal swallow tail appearance of susceptibility signal pattern** in the substantia nigra on axial imaging is perhaps the ***most promising diagnostic sign***

- ***Nigrosome-1**, within the posterior third of the substantia nigra, returns high signal on axial SWI in a linear or comma shape.* ***It is surrounded anteriorly, laterally and medially by low SWI signal intensity which resembles the distinctive split tail of a swallow***

- ***In Parkinson disease the high SWI signal within nigrosome-1 is absent and hence the swallow tail sign is absent.*** **This is believed to be the result of abnormal iron metabolism**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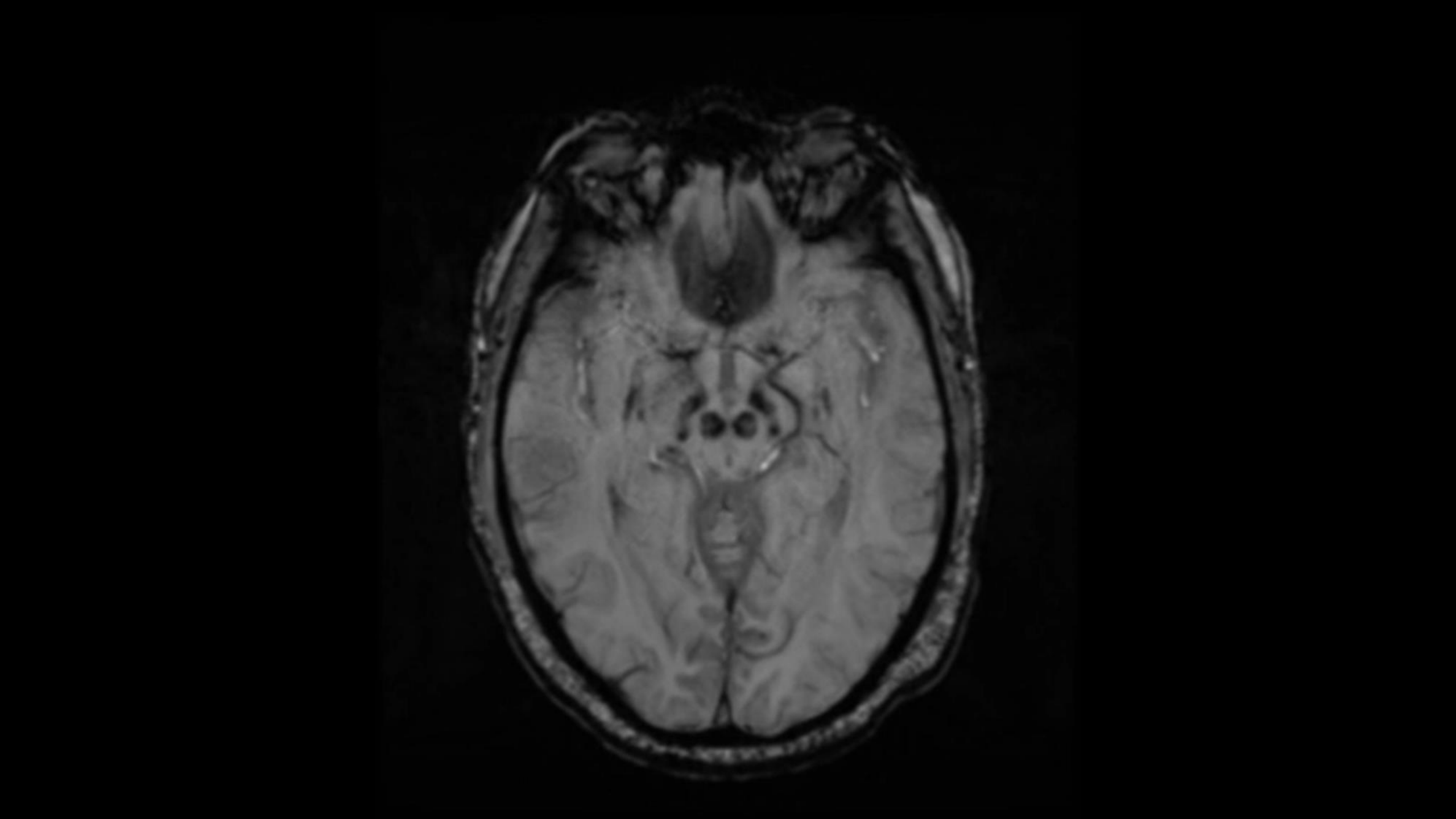
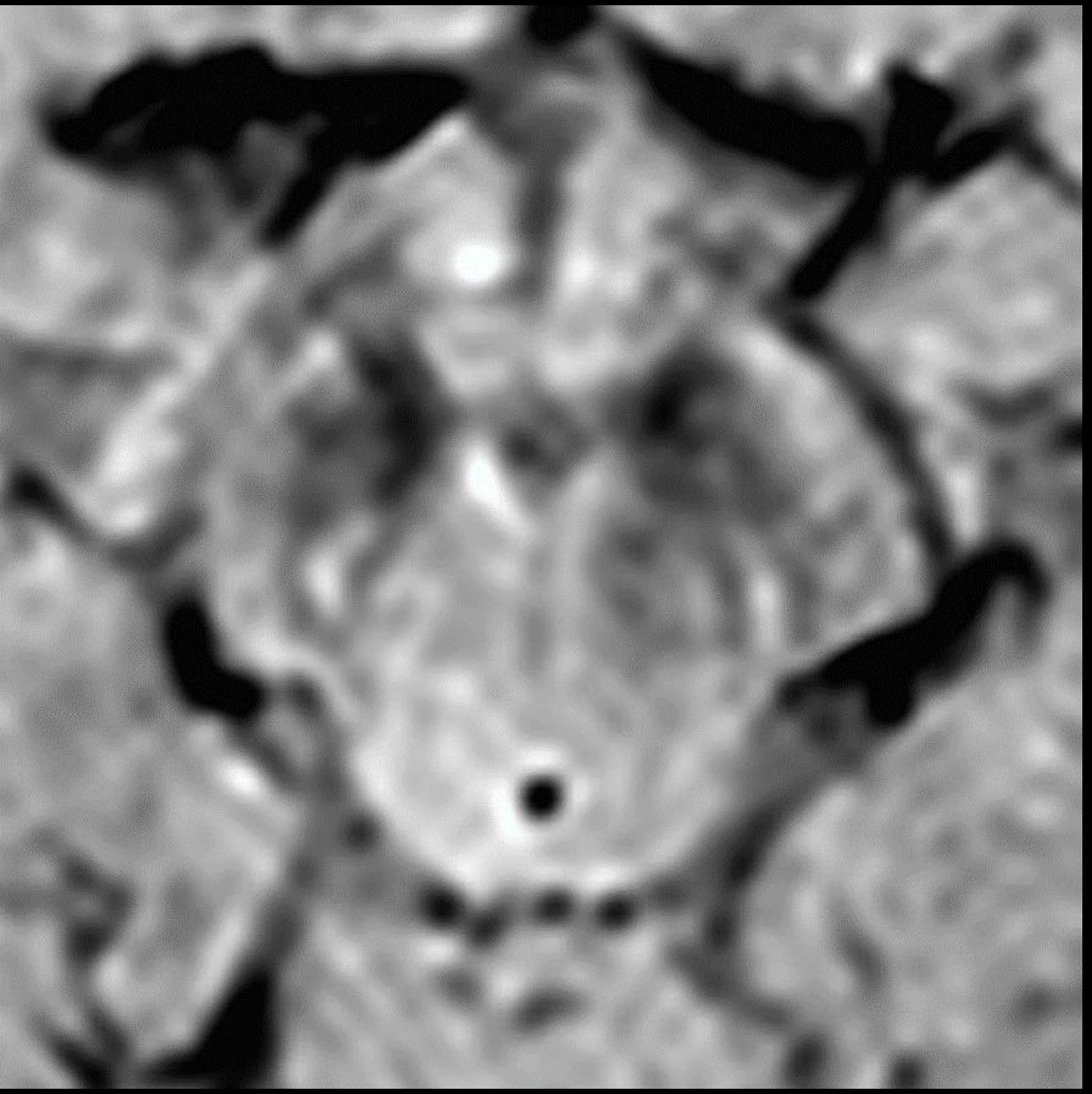
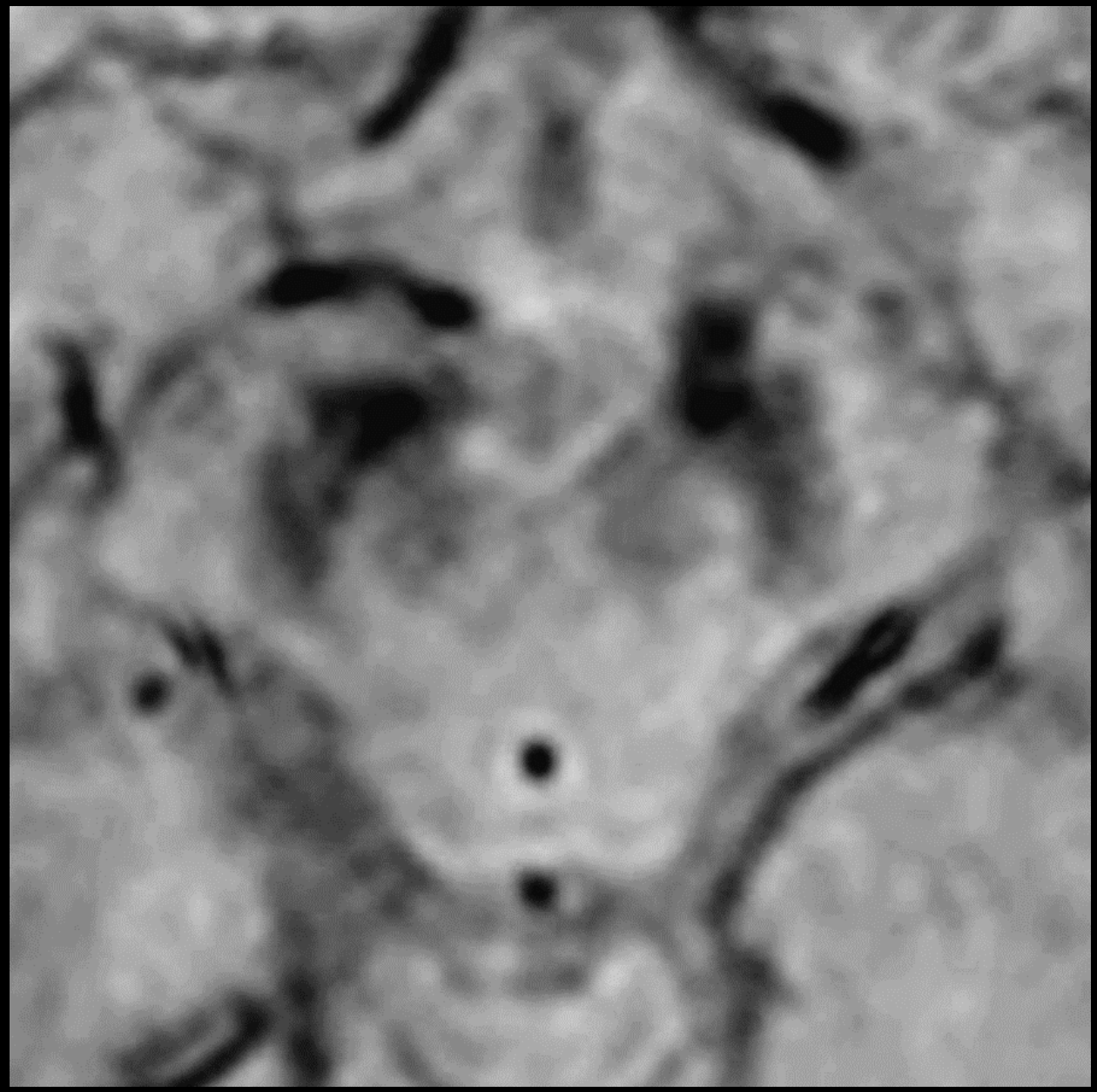






Parkinson-plus syndromes

- **Progressive supranuclear palsy (PSP):**
- more common forms of secondary parkinsonism.
- As with idiopathic Parkinson's disease***, progressive supranuclear palsy has a late age of onset, but the symptoms tend to progress far more rapidly once they appear.***
- However, dementia tends to have a later onset as the disease progresses.
- **Multiple system atrophy (MSA)**
- results in symptoms that are similar to idiopathic Parkinson's disease, but with a much faster progression.
- **Corticobasal degeneration (CBD)**
- This is the least common of the atypical parkinsonisms.
- caused by a build-up of proteins called tau, which damage parts of the brain
- ***The condition tends to start on one side of the body and slowly spread to other areas over time***

I.Progressive supranuclear palsy

- **becomes clinically apparent in the *6th decade of life*, and *progresses to death usually within a decade***

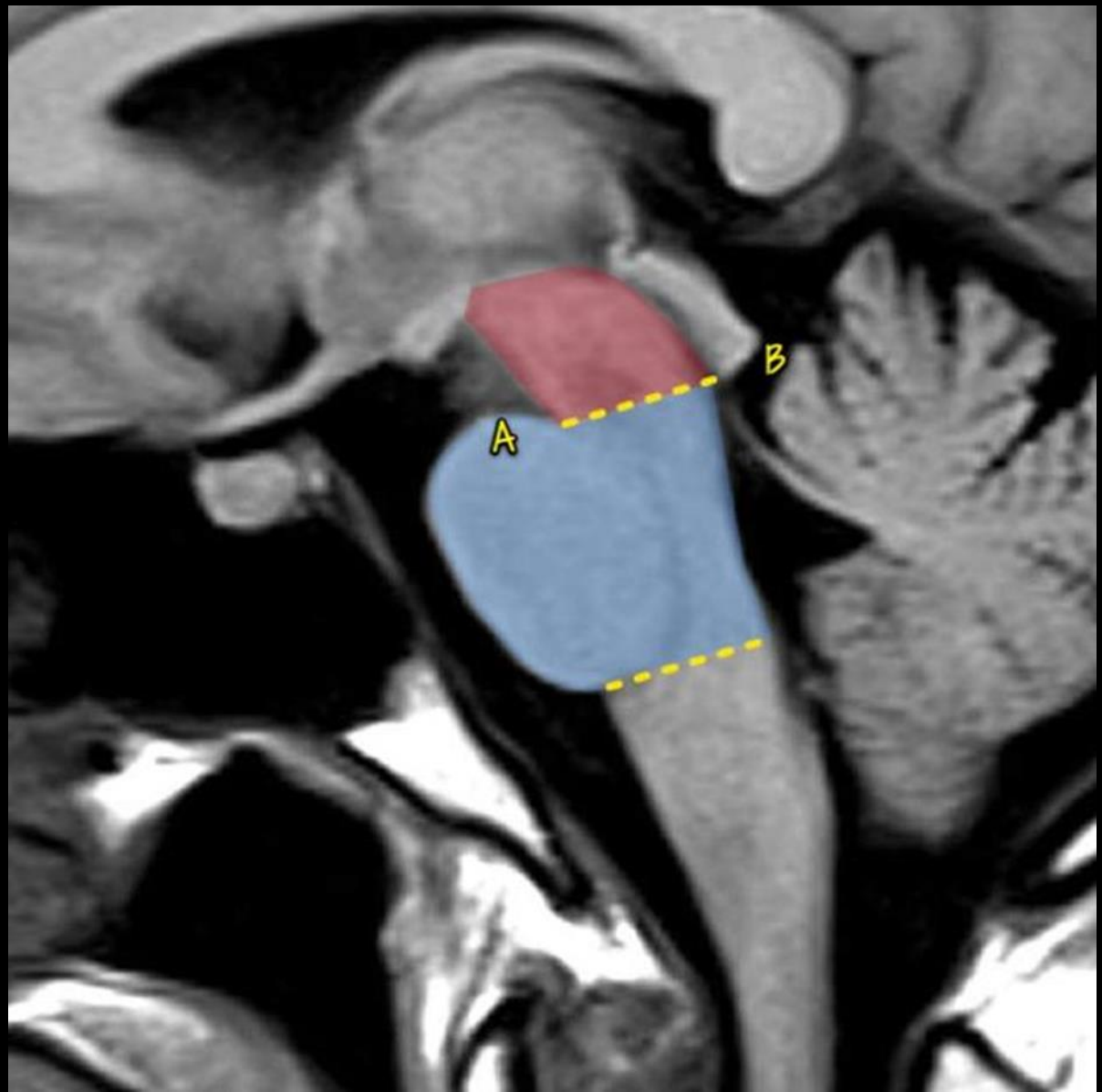
- **Sign and symptoms:**

1. decreased cognition
2. abnormal eye movements (supranuclear vertical gaze palsy) *there is pronounced **atrophy of the midbrain** (mesencephalon), which accounts for the typical upward gaze paralysis*
3. postural instability and falls, as well as parkinsonian features
4. speech disturbances

Main radiologic feature: midbrain atrophy

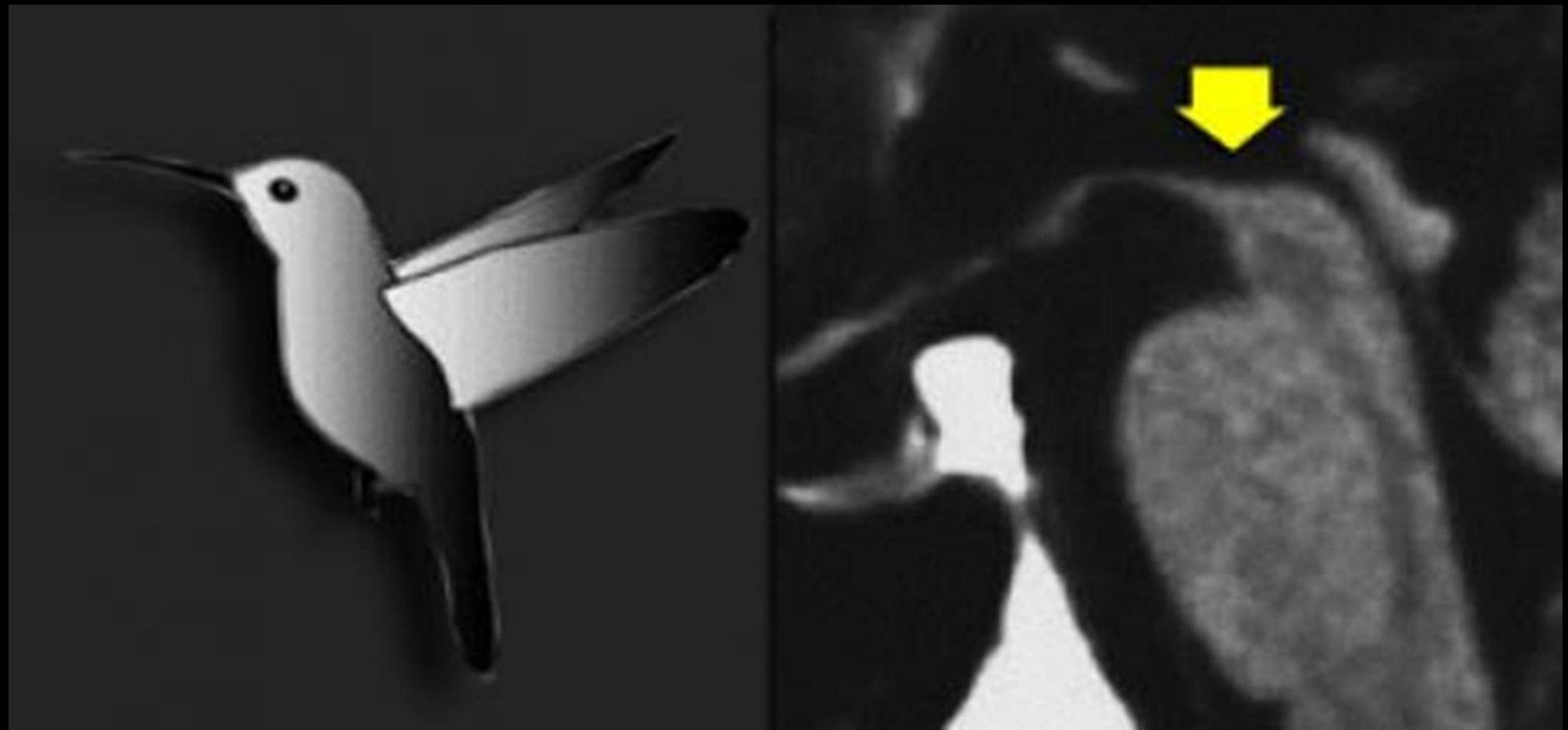
1. Midbrain to pons area ratio: A normal value is approximately 0.24 whereas it is significantly reduced in PSP to 0.12

- **Measurement is done at midsagittal images**



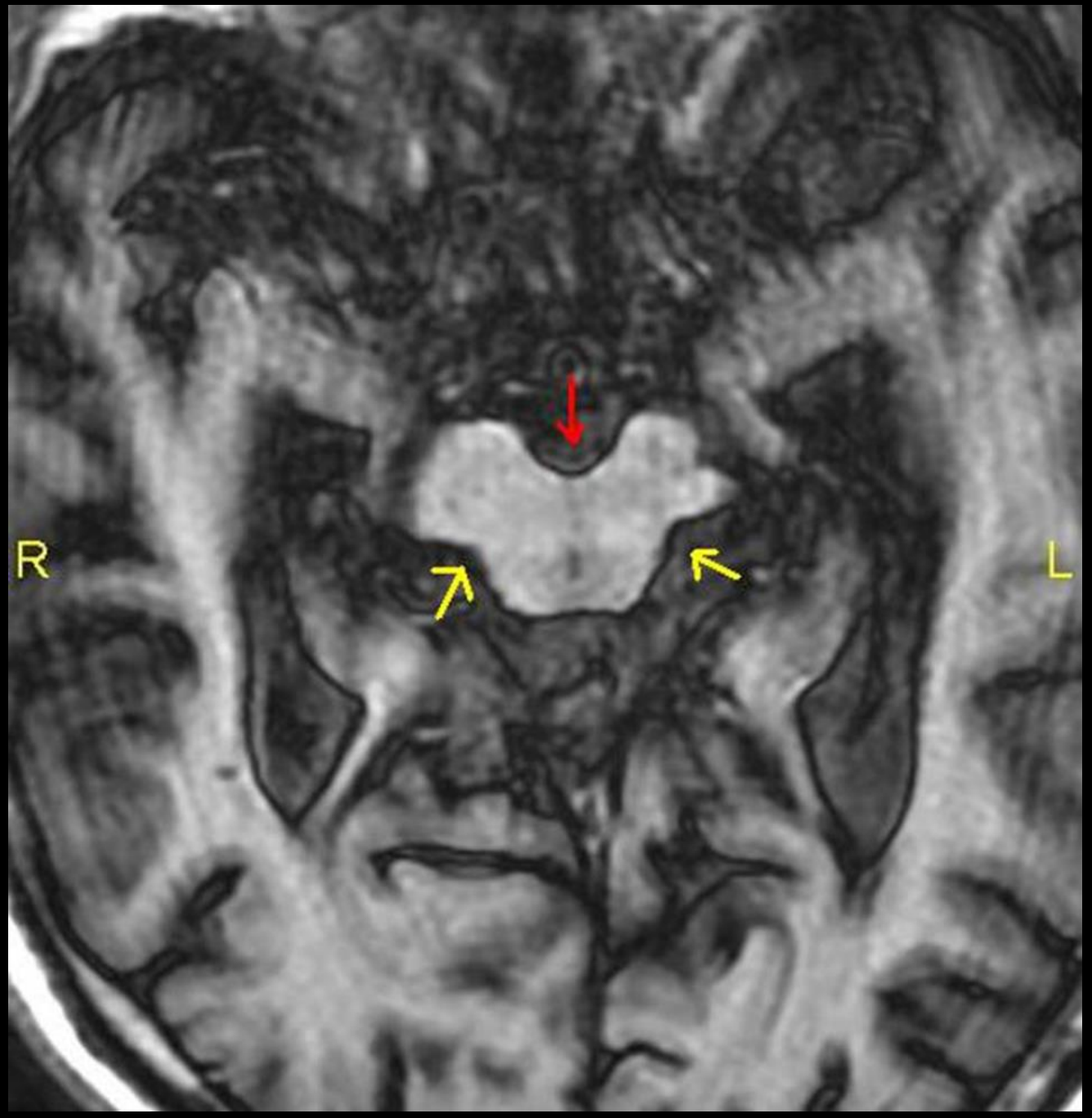
2.hummingbird sign (penguin sign): Normally the upper border of the midbrain is convex.

- ***The atrophy of the midbrain in PSP results in a concave upper border of the midbrain***



3.mickey mouse appearance reduction of anteroposterior midline midbrain diameter,

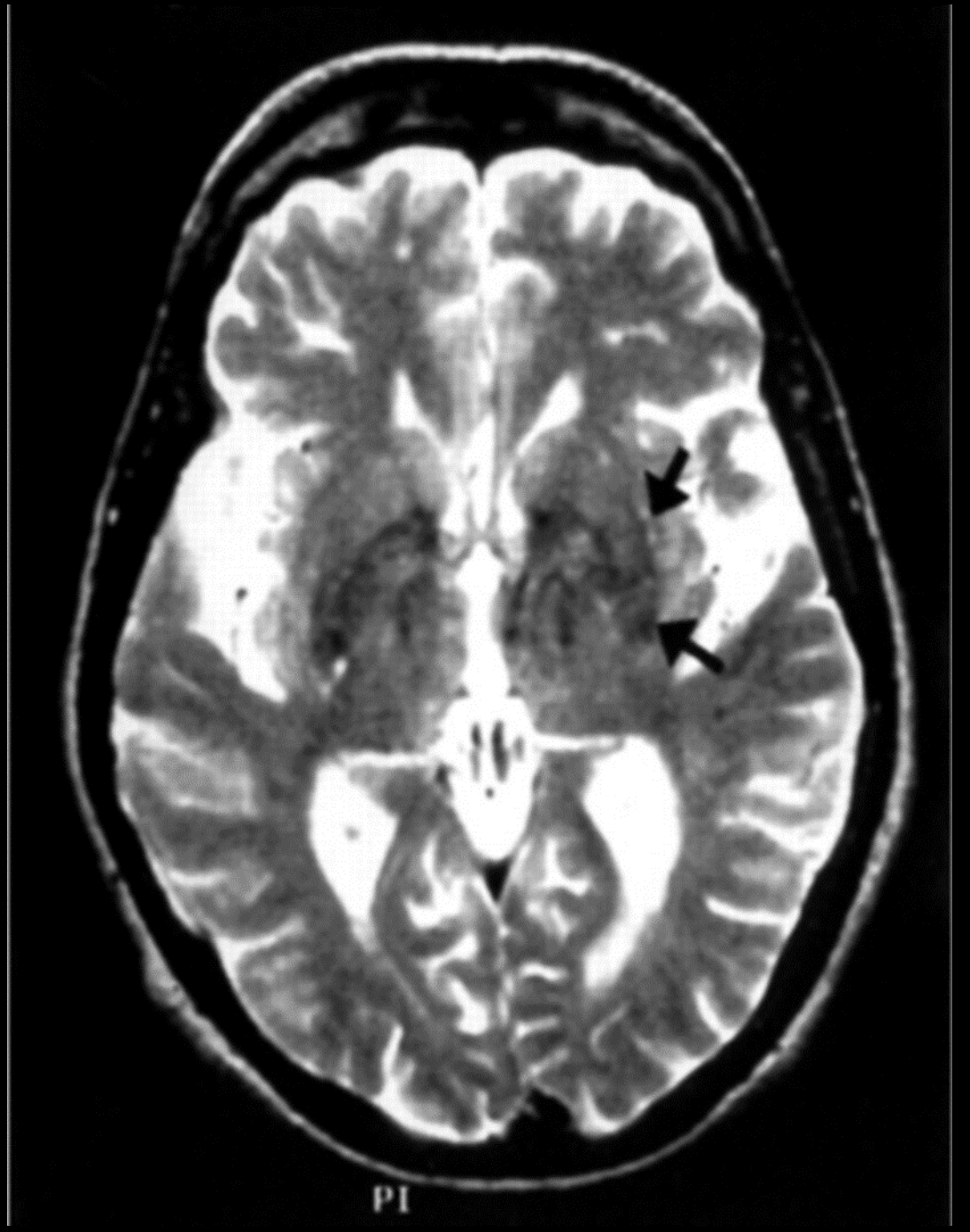
at the level of the superior colliculi on axial imaging ,from interpeduncular fossa to the intercollicular groove: <12 mm



Other imaging findings:

1. Increased iron may be found in the putamen so that it appears more hypointense than the globus pallidus on T2WI (the opposite of normal patients).

2. third ventricle may be enlarged



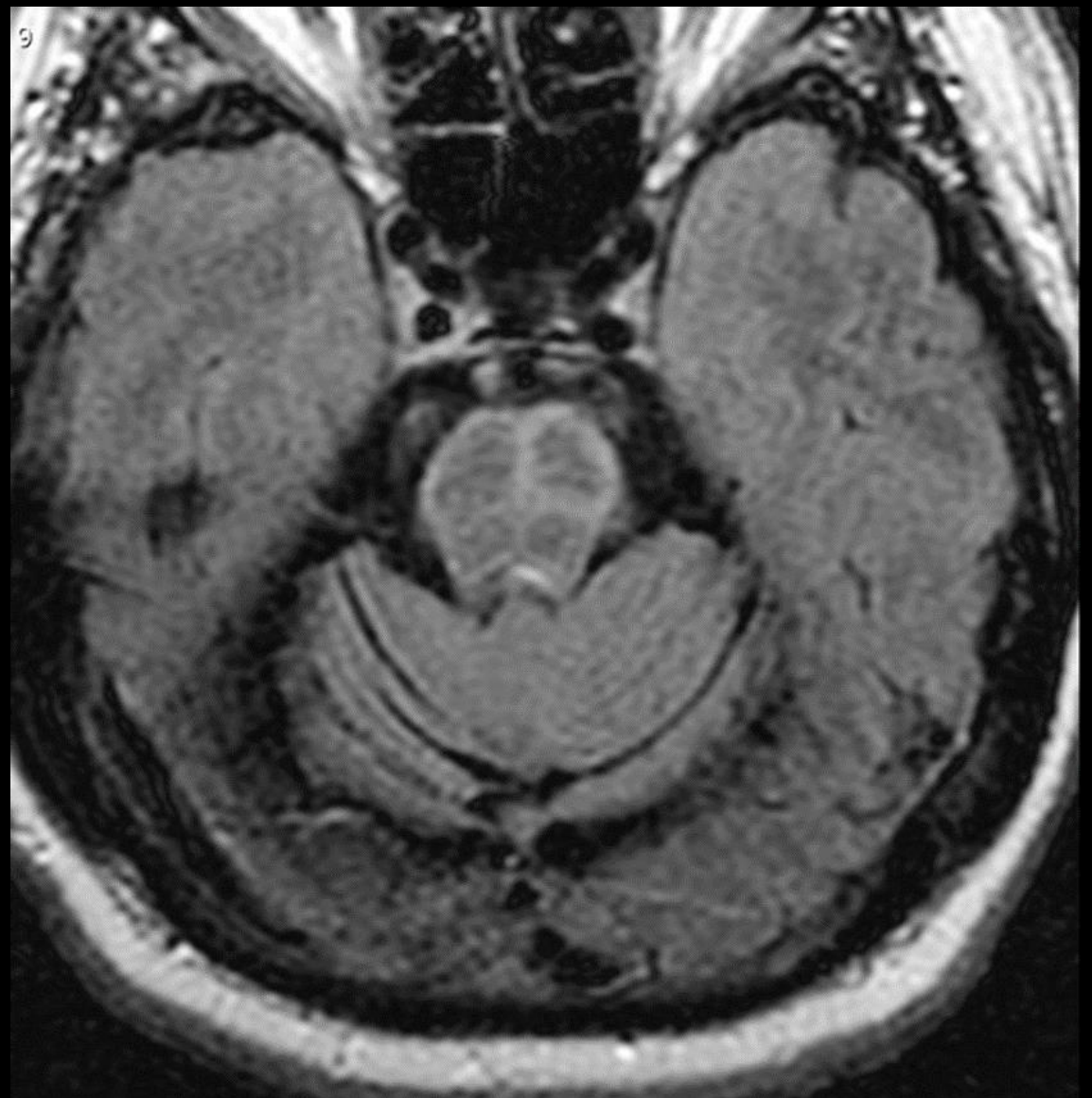
II.Multi System Atrophy (MSA)

- rare neurological disorder characterized by a combination of **1.**parkinsonism, **2.**cerebellar and pyramidal signs, and **3.**autonomic dysfunction ***(such as autonomic abnormalities of temperature regulation, sweat gland function, and maintenance of the blood pressure (orthostatic hypotension).***
- can be classified as MSA-C, MSA-P or MSA-A.

- **MSA-A** is the form in which autonomic dysfunction predominates and is the new term for what was formerly known as **Shy-Drager syndrome.**

MSA-C: predominance of cerebellar symptoms (olivopontocerebellar atrophy)

- **hot cross bun sign** refers to the MRI appearance of the **pons** *when T2 hyperintensity forms a cross on axial images,* ***representing selective degeneration of pontocerebellar tracts.***



III.Corticobasal degeneration

- **present with cognitive dysfunction, usually in combination with Parkinson-like symptoms.**

- **MR will demonstrate symmetric or asymmetric thinning of precentral and postcentral gyri with central sulcus dilatation.** ***The superior parietal lobule and superior frontal gyrus seem to be at particular risk for volume loss(knife-blade atrophy)***

- **Parasagittal involvement is prominent**
- **typical feature is the asymmetry in the parasagittal and paracentral atrophy between the two hemispheres.**