Neurodegenerative diseases: advances in functional neuroimaging

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Learning objectives

Degenerative brain diseases occasionally show some MR imaging findings that can lead to a definitive diagnosis.

Advanced functional imaging (MR-spectroscopy, diffusion tensor imaging, functional MR and PET-FDG) provide useful information for an early diagnosis of these disorders and specially for the evaluation of the disease evolution.

Our work presents a pictorial review of the morphological changes of some of the most common neurodegenerative diseases as Alzheimer dementia (AD), multisystem atrophy (MSA), hypertrophic olivary degeneration (HOD), frontotemporal dementia (Pick disease) (FTD), amyotrophic lateral sclerosis (ALS), Parkinson disease (PD) and Creutzfeld-Jacob disease (CJD). We specially focus in the utility of diffusion tensor image for the diagnosis of some of these diseases, as AD ALS or FTD, due to the early involvement of the uncinate fasciculus (UF)

Background

Neurodegenerative disease (Fig. 1 on page ), entail a progressive cerebral desestructuration that only have symptomatic temporally treatment. These brain pathologies are a continous challenge for the departments of radiology and neurology.

Neuroradiologist attempt to classify these patients to include them into clinical trials to validate new therapies. These new therapies try to stabilize the disease and to improve their quality of life. Often there is an overlap of the clinical findings of these entities and in these cases the information from the neuroimaging can be crucial.

Neuroimaging can corroborate the clinical diagnosis with the dates from the structural MR findings or in the last years with those obtain from the functional imaging, specially diffusion tensor imaging, MR-spectroscopy, functional-MR or FDG-PET, aiding to the probable final diagnosis.

We review the morphological MR findings obtain from the conventional sequences and the possible findings from the advanced functional imaging in the most common degenerative brain diseases in the daily practice.

Images for this section:
Neurodegenerative diseases

Fig. 1: FIGURE 1
Imaging findings OR Procedure details

ALZHEIMER DEMENTIA (AD)

Progressive degenerative disease of brain due to abnormal accumulation of tau protein, which plays a key role in neuronal dysfunction and cell death (Fig. 2 on page ).

The degenerative process begins in the medial temporal lobe, in the hippocampus (Fig. 3 on page ) followed by the parahippocampal gyrus and the frontotemporal regions. Visual and motor areas are spared in the early process.

Current role of imaging in AD is to exclude treatable dementias and identify early-onset cases for possible innovative therapy.

Conventional MR sequences can show morphological findings like (Fig. 4 on page ): a) temporal cortical atrophy with disproportionate hippocampal volume loss; b) loss of the lobulated hippocampal surface and some degree of hyperintensity on FLAIR images; c) enlargement of the parahippocampal fissure; d) enlargement of the Sylvian fissures and of the lateral ventricles; e) parietal and temporal cortical atrophy (Fig. 5 on page ).

MR volumetric analysis of the hippocampus and the enthorinal cortex are useful to the evaluation of the disease evolution. (Fig. 6 on page , Fig. 7 on page ). This method may help in distinguishing patients with mild cognitive impairment from normal elderly subjects. The principal disadvantage is the requirement of sophisticated software for segmentation and volumetric acquisition.

The role of advanced functional imaging, is to provide more information from normal MR-appearing brain areas.

MR-spectroscopy may contribute to the diagnosis in AD. It is a fact that AD patients evidence an increase in the glial metabolite myoinositol (mI) (3.56 ppm) in the posterior cingulum, with a marked reduction in the ratio NAA/mI (Fig. 8 on page ). This low ratio may help in distinguishing AD and the other dementias, but is not specific. Another ratio, NAA/Cr <1.61 in the posterior cingulum or in the enthorinal cortex may predict in three years the evolution of mild cognitive impairment to AD.

Wallerian degeneration of the white matter due to the neuroaxonal damage in AD lead to the decrease of the density of the association white matter fibers of the temporofrontoparietal regions. One of the consequence of this mechanism is the volume loss of the temporal stem, a crucial area for intrahemispheric connections through the uncinate fasciculus, the inferior frontooccipital tract and the loop of Meyer. In early AD the most affected tracts are the uncinate, involved in the memory circuits and the inferior frontooccipital fascicle, involved in the language network. (Fig. 9 on page ), while...
the tracts involved in primary functions (visual and motor functions) as the loop of Meyer and the corticospinal tract, are affected in late AD. These changes are difficult to verify in the studies of diffusion tensor imaging because of the lack of a cut off of normal value of the fractional anisotropy, but are very useful to support the clinical diagnosis when an evident left-right asymmetry in the fractional anisotropy or in the diffusion values can be documented. Sometimes the 3D tractography reconstruction shows a global volume loss of the white matter fascicles, specially of the left uncinate fascicle.

In the literature, activation functional MR imaging (fMRI) have revealed decreased areas of activation in the medial temporal cortex in AD in functional studies of memory. In our experience memory paradigms are difficult to reproduce even in healthy patients, so its routine application in daily practice is very unlikely. (Fig. 10 on page ).

Classically used, SPECT studies evidence extensive areas of cortical hypoperfusion, specially centered in parietal and temporal regions (Fig. 11 on page ), but this technique lacks of anatomical correlation.

Actually, FDG-PET studies can measure the amount of glucose deposition in cerebral regions and its distribution, deducing the cortical activity in the different areas. The primary site of affection is the hippocampus, the entorhinal cortex and the posterior cingulum, but the classic pattern of AD is the reduced metabolism in the association parietal and temporal areas (Fig. 12 on page ) (Silverman y cols. 2002). In mild cognitive impairment, the risk to develop AD increases 5 times if the classic hypometabolism is documented in PET studies. Indeed the regional hypometabolic areas (decrease utilization of the glucose) in temporal/parietal lobes correlate with the degree of cognitive impairment.

FDG-PET imaging also helps to differentiate AD from other types of dementia like vascular dementia (global hypometabolism or patched hypometabolic areas), diffuse Lewy body disease (hypometabolism of the visual cortex and cerebellum). These nuclear medicine findings can be seen in asymptomatic patients (one or two years before the clinical onset) (Fig. 13 on page ).

In the immediate future #-amyloid-PET imaging may contribute to the accurate and early diagnosis of AD.

**FRONTOTEMPORAL DEMENTIA (FTD)**

Frontotemporal dementia is the new name for clinical Pick disease. This term is restricted to the pathologic variant with Pick bodies into an atrophyc cerebral lobe Anterior frontotemporal atrophy is the major morphological finding. (Fig. 14 on page ).

The marked atrophy is restricted to the anterior frontotemporal lobes, sparing the posterior aspect of the superior temporal gyrus and the pre- and postcentral gyri. Parietal and occipital lobes are also spared. Usually the dominant hemisphere is the most affected.
Structural-MR imaging evidence the secondary changes to the cortical atrophy (Fig. 15 on page ) a) dilated subarachnoid space over the frontal and temporal lobes b) hyperintensity in frontotemporal white matter secondary to gliosis (Fig. 16 on page ); c) enlargement of the frontal ventricular horn.

MR-Volumetric analysis document not only the progressive cortical atrophy, but the volumen loss in the anterior commissure, the anterior cingulum and the caudate nucleus related to healthy patients and patients with AD (Fig. 17 on page , Fig. 18 on page ). FTD is characterized by changes in social behavior and cognitive disfunction, in which are implicated in frontostriatal networks. The reduction in the "inputs" to the caudate is related to the degree of the cognitive disfunction.

In the studies of advanced functional imaging, spectroscopy-MR may evidence reductions in the NAA peak and increases in the ml peak in the frontal lobe or in the anterior cingulum (Fig. 19 on page ), with normal spectral curves in the posterior cingulum that may aid in the differential diagnosis with AD patients..

Diffusion tensor imaging can show interesting MR findings. First, the uncinate fasciculus, implicated in the memory network, (Fig. 20 on page ) is clearly reduced. The progressive decline in the fractional anisotropy values, may be an indicator of the evolution of the disease. Second the volume loss of the anterior cingulum, involved in the initiation and stimulation process, and the decrease in the fractional anisotropy values of the inferior longitudinal fascicle, involved in the semantic visual aphasia (Fig. 20 on page ), are correlated with the clinical findings in patients with FTD

FDG-PET imaging reveals a marked hypomethabolic areas in the frontotemporal cortices and the anterior cingulum with a normal metabolism in the parietal and occipital regions (Fig. 21 on page )

AMYOTROPHIC LATERAL SCLEROSIS (ELA)

Lou Gehring disease, motor neuron disease or Charcot disease.

Selective degeneration of the somatic motor neurons of the brainstem, of the lower motor neurons of the spinal cord, of the large pyramidal neurons of the motor cortex and eventual loss of the corticospinal fibers (Fig. 22 on page )

The clinical presentation may be with symptoms from affectation of the upper motor neuron (UMN) (Babinsky sign, spasticity, hyperreflexia) or lower motor neuron (LMN) (asymmetric muscle weakness, atrophy, fasciculations, hyporreflexia) as well with symptoms from bulbar disfunction (slurred speech, dysphagia). The later are patients with more rapid deterioration and death.

Structural classic MR-imaging has been solely made to exclude other entities, specially with patients with clinical bulbar symptoms, but the actual high field MR scans provide
more information from the conventional sequences and may aid to the radiologist to support the clinical diagnosis.

The imaging interpretation pearl in the ALS with LMN signs, is the high signal intensity on T2-weighted images in the anterior and lateral tracts of the spinal cord. This finding can be accompanied with a change in the normal convexity of the cord. (Fig. 23 on page ). This change can appear early in the evolution of the disease and is commonly seen in young patients with a more rapid disease progression. Later the atrophic cord can be visualize with a "heart shape" secondary to the marked affectation of the lateral and anterior fascicles of the corticospinal tract.

The radiological findings of the ALS with UMN signs are less specific but commonly may aid to support the clinical diagnosis. These findings are (Fig. 24 on page ) a) hyperintensities on T2 weighted images in the perirolandic cortex, b) hypointensity on T2 weighted sequences in the perirolandic cortex, related to iron deposits c) hypo or hyperintensity on T1 weighted images in the internal capsule; d) hyperintensities on T2 weighted images along the intracranial corticospinal tracts (unspecific finding found in other diseases as adrenomyeloneuropathy, Krabbe disease, Charcot Marie Tooth disease and even in health patients when is restricted to the posterior limb of the internal capsule). This isolated imaging finding can be consider pathologic when extended beyond the internal capsule and when progressed overtime (Fig. 25 on page ).

**Volumetric series** can reveal a mild progressive cerebral volumen loss in patients with ALS. In cases of ALS associated to dementia with frontal and temporal atrophy, these techniques are useful because this subtype has the same imaging features that FTD. There are some studies in the literature reporting the progressive volumen loss of the spinal cord, but the programs used in the volume measurements are sophisticated and expensive to the daily clinical practice.

MR-spectroscopy and diffusion tensor imaging are the most useful **advanced functional techniques** in the diagnosis of ALS

**MR-spectroscopy** reflects the neuronal loss and the viability, with reduced ratios of NAA/Col and NAA/Crt and ratio NAA/ ml in perirolandic cortex, the later due to the increase of ml secondary to gliosis (Fig. 26 on page ). These abnormal spectral curves can also be seen in normal appearing cerebral tissue, making the spectroscopy an important method for monitoring the disease because the changes in the metabolites, although not specific for ALS, correlate with the clinical severity.

**Diffusion tensor images** evidence differences in the fractional anisotropy values in the corticospinal tract in ALS from normal subjects. There is a reduction of these values from the posterior limb of the internal capsule to the pyramids. Volume loss of the corticospinal tract can be seen in the 3D-reconstructions in ALS (Fig. 27 on page , Fig. 28 on page ). These radiological findings, although very useful in a particular patient, are
not yet included in the general protocols of the monitoring of the disease, because it is difficult to assess an standard cut-off value of fractional anisotropy.

**Activation functional MR images** in patients in ALS reveal abnormal patterns of motor activation related to controls (normal subjects) with a greater activation outside the primary motor areas (supplementary motor area, sensorial cortex, inferior parietal cortex, cerebellum) with simple paradigms as the classic "finger-tapping". Sometimes, ALS-patients have some difficulties to perform the motor task and the MR technician must aid to the patient to do the task, obtaining the similar activations (Fig. 29 on page ). There is not a typical pattern of motor activation in ALS-patients and the findings depend on the major or minor involvement of the UMN or LMN.

**MULTIPLE SYSTEM ATROPHY (MSA)**

Also known as Olivopontocerebellar atrophy (OPCA) (Fig. 30 on page ) when the cerebellum, cerebellar peduncles, inferior olives and pons are affected, or striatonigral degeneration (SND) when the putamen, pontine base are involved and Shy-Drager syndrome when the autonomic disorders are the most prominent

Sporadic disorder with progressive neurodegeneration with a combination of cerebellar, pyramidal, extrapyramidal and autonomic disorders.

**Structural-MR** images reflect some characteristic findings, but non-specific (Fig. 31 on page ):
- a) marked atrophy of the cerebellar peduncles with > or < hyperintensities on T2-weighted sequences
- b) mild to marked atrophy of the cerebellar white matter with > or < hyperintensities on T2-weighted sequences
- c) enlargement of the fourth ventricle and the cerebellar pontine angle
- d) flat ventral pons without any signal changes
- e) "hot cross bun" sign (Fig. 32 on page ) or cruciform pontine on T2-weighted images due to the degeneration of transverse fibers
- f) hyperintensity on T1-weighted images or hypointensities on T2-weighted sequences in the lateral e inferior margin of the putamen due to ferromagnetic deposits (more commonly seen in the SND)
- g) "slit-like void" sign, linear hypointensity on T1-weighted or hyperintensity on T2-weighted images in the lateral rim of the putamen, secondary to the atrophy and to the enlargement between external capsule and the putamen (Fig. 32 on page ).

Volumetric studies are not commonly used in this entity, but may show the progressive cortical atrophy of the frontal and parietal areas.

**Advances functional imaging:** spectroscopy-MR and diffusion tensor imaging tensor may aid to the diagnosis of this entity when the morphological findings are not seen yet.

Spectroscopy-MR show in early stages marked increased of the Choline and myo-inositol peaks in pons and cerebellum, and in late stages a marked decline in NAA peak (Fig. 33 on page ). Decreased ratio NAA/Crt has been correlated with disability.
Diffusion tensor imaging in MSA helps to differentiate between MSA and other similar diseases as SND or Parkinson disease (PD). The fractional anisotropy (FA) in the medium cerebellar peduncles or in the cerebellar white matter are clearly reduced (Fig. 34 on page ). Transverse pontine fibers degeneration can be easily seen in 3D or 2D reconstructions (Fig. 34 on page ).

**IDIOPATHIC PARKINSON DISEASE (PD)**

Classic parkinsonism is a progressive neurodegenerative disorder caused by a dysfunction of the pars compacta of the substantia nigra. The progressive loss of the dopaminergic neurons of the nigrostriatal system (Fig. 35 on page ) results in the development of motor symptoms as resting tremor, bradykinesia, rigidity and gait difficulty. The clinical onset is typically between 50-60 years.

DaTSCAN imaging is considered the gold standard technique with a specificity of 94% in later stage, but about a 70% in early stage of the disease, when clinically is considered as "probable" PD.

*Structural-MR images* do not evidence specific findings: a) low signal intensity on T2-weighted images in substantia nigra, globus pallidus; b) dilation of the lateral ventricles and third ventricle due to atrophy; c) enlargement of the interpeduncular cystern (Fig. 36 on page ).

*Advanced functional images*, specially spectroscopy-MR, diffusion tensor and f-MRI may confirm the suspected clinical diagnosis when the DaTSCAN is not conclusive.

Diffusion tensor studies may show a reduced FA in the substantia nigra, as in another locations as the superior longitudinal fascicle and the genu of the corpus callosum secondary to the damage of the nigro cortical and the consequent cortical hypoactivity to the reduced excitatory thalamic outflow (Fig. 37 on page , Fig. 38 on page ). The values in the FA and the mean diffusivity can aid to the differential diagnosis of the PD from other similar clinical entities as MSA and progressive supranuclear palsy (PSP). In the MSA, mean diffusion values in the posterior putamen, midbrain, pons, middle cerebellar peduncle, and cerebellar white matter were significantly higher than in PD. In the PSP group, the values in the globus pallidus, head of the caudate nucleus, midbrain and superior cerebellar peduncles are significantly increased relate to the values of PD.

Patients with clinically "probable" or "possible" PD, can evidence a neuronal recruitment in *functional activation (f-MR) motor studies*, with a marked decreased of the activation of the primary motor areas and a shifting of the activation to the parietal cortex. In line with the "functional deafferentation" hypothesis, de novo PD patients show hypoactivation of the contralateral SM1 and initially no areas of hyperactivation during execution of a motor task. Increased in the activation in motor, premotor and parietal cortex parallels disease severity. (Fig. 39 on page )
MR-spectroscopy in PD in line with the deafferentation hypothesis reveals a reduced NAA peak in frontal white matter; the normal spectral curve in the cerebellum may contribute to the differential diagnosis with MSA or essential tremor. (Fig. 40 on page ).

PET-FDG is very useful in the differential clinical diagnosis of the patients with PD, basically with PSP and corticobasal degeneration. (CBD). While the metabolic pattern in PD is unspecific, with only bilateral hypometabolism in temporal and parietal lobes when a clinical dementia is evident, the metabolic pattern of the FDG-PET maps in PSP is clearly distinct from PD, with a characteristic bilateral thalamic hypometabolism. Another radiological features of this entity are: atrophy of the midbrain, high intensity on T2 weighted images in the periaqueductal gray matter and calcifications in both putamina. The metabolic pattern of the FDG-PET in the CBD, which appears with a mild to marked atrophy in the pre-postrolandic gyri, can show a hypometabolism in bilateral parietal cortex (Fig. 41 on page ).

HYPERTROPHIC OLIVARY DEGENERATION

Secondary degeneration of the inferior olivary nucleus usually caused by primary lesions in the dento-rubro-olivary pathway (Guillain-Mollaret triangle) (Fig. 42 on page ). This pathway connects the dentate nucleus with the red nucleus of the mesencephalon and with the inferior olivary nucleus of the medulla. The fibers interconnect the dentate nucleus with the contralateral red nucleus through the decussation of the superior cerebellar peduncles, the red nucleus with the inferior olivary nucleus through the tegmen central fibers and the inferior olivary nucleus with the contralateral dentate nucleus through the inferior cerebellar peduncle. (Fig. 43 on page ). Lesions in the cerebellum, superior cerebellar peduncle or pons may affect the inferior olivary nucleus with enlargement of the nucleus secondary to vacuolar degeneration and marked astrocytosis.

Ipsilateral HOD occurs when the primary insult is located in the tegmen, contralateral HOD if the lesion is in the dentate nucleus or superior cerebellar peduncle and bilateral HOD if both tegmen and cerebellar superior peduncle are involved. HOD does not occur when the primary lesion is located in the inferior olivary nucleus.

Structural- MR shows characteristic findings with enlargement of the medullary olive and high signal intensity on T2-weighted images (Fig. 44 on page ). The hypertrophy of the olivary nucleus appears between six months to three-four years after the clinical insult. The high signal intensity can persist indefinitely.

Advanced functional imaging as spectroscopy-MR or f-MRI are limited for the diagnosis; spectroscopy curves are suboptimal because of the location of the lesions and activations maps are commonly contaminated with the patients movements due to the palatal tremor.
Diffusion tensor images and fiber tractography can demonstrate the disruption, the change in the morphology or the volumen loss of the fiber tracts involved in the triangle of the Guillain Mollaire (Fig. 45 on page , Fig. 46 on page ).

CREUTZFELDT-JACOB DISEASE (CJD)

Rapidly progressing fatal dementia, potentially transmissible disorder caused by a prion protein. Also called transmissible spongiform encephalopathy.

Structural- MR on conventional sequences are characteristic (Fig. 47 on page ) and advanced imaging is not necessary. a) high signal intensity on T2 weighted images in the caudate nuclei, putamina and thalami; b) high signal intensity on T2 images in the peripheral cerebral cortex (most commonly frontal and temporal lobes and slight atrophy; c) hyperintensity in the adjacent cerebral white matter; d) restricted diffusion in the striatum and the cerebral cortex.

Images for this section:

Fig. 2: FIGURE 2
Fig. 3: FIGURE 3
Fig. 4: FIGURE 4
Fig. 5: FIGURE 5
The degenerative process begins in the hippocampus, followed by the para-hippocampal gyrus and the parietal and temporal regions. Visual and motor areas are spared in the early process. Structural 3D images evidence temporal and parietal cortical atrophy.

**Fig. 6: FIGURE 6**
Fig. 7: FIGURE 7
Fig. 8: FIGURE 8
Volume loss of the temporal stem in patients with AD, with reduced volume fibers of the uncinate fasciculus (U), more evident in the left side. 3D tractography reflects the volume loss and the clustering of the frontooccipital (IFOF) fibers. SFOF: superior frontooccipital fascicle.

**Fig. 9:** FIGURE 9
SIMPLE FUNCTIONAL STUDY OF VERBAL-SPATIAL MEMORY in patients with cognitive impairment. Indeterminate results

**Patient 1**: global cognitive impairment: *posterior and bilateral temporal activity*

**Patient 2**: mild cognitive impairment of the verbal memory: *more prominent medial temporal activity on the right side*

**Patient 3**: Left mesial sclerosis, mild cognitive impairment of the verbal memory: *medial temporal activity on the right side*

Patients with unilateral damage can show a marked contralateral activation on f-MR. Patients with a more global damage may evidence an early stage of "compensation" with bilateral hiperactivity, followed of a clearly reduced activation in the medial temporal regions.

**Fig. 10: FIGURE 10**
**Normal findings in an aging brain**

**CEREBRAL SPECT**

**Late stage Alzheimer disease**

**Fig. 11: FIGURE 11**
Diagnostic performance of FDG-PET in early stage of cognitive impairment

**Mild cognitive impairment** → **Alzheimer disease**

- The regions that predict the evolution (ED: 80-97% in the literature):
  - Entorhinal cortex
  - Inferior parietal cortex
  - Superior temporal cortex
  - Posterior cingular cortex

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The hippocampus is the first location involved in AD, but the posterior associative cortex and the posterior cingular are the hypometabolic areas seen on studies of FDG-PET.


**Fig. 12:** FIGURE12
Fig. 13: FIGURE 13

Mild cognitive impairment age-related
Focal unilateral hypometabolism of the parietal cortex, with preservation of the posterior cingulum, temporal, frontal and occipital areas

Alzheimer disease moderate stage
Bilateral parietal cortices and posterior cingulum hypometabolism
Temporal and frontal regions preserved

Advanced Alzheimer disease
Bilateral temporal and frontal hypometabolism
Occipital regions preserved
Fig. 15: FIGURE 15

Low signal intensity in the subcortical white matter due to fronto-temporal asymmetric gliosis. In late stage, only the atrophic lobe is seen.
Fig. 16: FIGURE 16
Fig. 17: FIGURE 17
Fig. 18: FIGURE 18

Frontotemporal atrophy, more prominent in frontal lobe. Perirolandic cortices are spared.
Fig. 19: FIGURE 19
Fig. 20: FIGURE 20
**Fig. 21:** FIGURE 21

*FDG-PET*

**Frontotemporal dementia**
Low glucose hypometabolism in frontal and anterior temporal cortices
Normal metabolism in the occipital and parietal cortex

**Frontotemporal dementia**

**Moderate Alzheimer disease**
Fig. 22: FIGURE 22
Fig. 23: FIGURE 23
Fig. 24: FIGURE 24

- **AMYOTROPHIC LATERAL SCLEROSIS**
- Progression in time of the involvement along the craneocaudal axis of the CST

The high signal intensity of the CST, extends beyond the posterior internal capsule (ICp)
Fig. 25: FIGURE 25
AMYTROPHIC LATERAL SCLEROSIS

Bulbar-ALS may show low concentration of NAA and high of myo-inositol metabolites in the periolandic cortices

Spectroscopy-MR can detect in LMN disease reduced concentrations of the NAA peak in the internal capsule. Spectroscopy-MR can monitor the evolution of the disease.

Fig. 26: FIGURE 26
Fig. 27: FIGURE 27

Clear asymmetry in the fractional anisotropy map, along the CST. The color maps reflect the asymmetry in the left motor cortex, the posterior internal capsule, and the right pyramid after the bulbar decussation.
Fig. 28: FIGURE 28
AMYOTROPHIC LATERAL SCLEROSIS

ALS-patient with right hand motor difficulties. Motor paradigms are performed with the aid of the MR-technician.

Mild-moderate increased in motor activation areas. Greater activation in left parietal cortex.

ALS-patient, without right hand motor difficulties.

Greater ipsi and contralateral motor activity and in the basal ganglia (recruitment).

Fig. 29: FIGURE 29
**Fig. 30:** FIGURE 30
High intensity signal on T2-images and atrophy in medium cerebellar peduncles. Progression in time to the superior cerebellar peduncles and to the cerebellar white matter. Dilation of the 4th ventricle.

Fig. 31: FIGURE 31
**Flat ventral pons, without signal changes on MR images**

"hot cross bun" sign: cruciform pontine high signal intensity due to the degeneration of the transverse pontine fibers

PONS
Transversas pontine fibers
Putamen

"slit-like void" sign or high signal linear band in the lateral margin of the putamen
Fig. 33: FIGURE 33
**Fig. 34:** FIGURE 34

Red areas evidence the major fractional anisotropy values (FA).

**MULTIPLE SYSTEM ATROPHY**

Clearly reduction of the red areas, specially in the medium cerebellar peduncles.

“Hot cross bun”, (in red in the FA map)
Fig. 35: FIGURE 35
Structural—MRI changes are minimal, except for the dilation of the interpeduncular cistern and the presence of low signal intensities on T2 images in substantia nigra and globus pallidum.
Fig. 37: FIGURE 37

SN: substantia nigra; CP: cerebral peduncle; Cbp: cerebellar peduncle
FA values are reduced in the pars compacta of the substantia nigra, increasing the ratio CP/SN related to health patients.

FA be reduced in white matter fascicles of frontal lobes secondary to the hypothesis of deafferentation.

Putamen FA values are similar to health patients and patients with essential tremor.

FA is similar to healthy patients in cerebellum.

Fig. 38: FIGURE 38
“Finger tapping” paradigm reflects a decreased of the activation in primary motor areas with posterior shifting to the parietal regions due to the damage in the nigro-corticostriatal pathway.

Reduction in the activation of the supplementary motor area in early stage, with hyperactivation or return to normal activation in late stage is a common finding, but also SMA activation can be unchanged.
Fig. 40: FIGURE 40
Fig. 41: FIGURE 41

The metabolic pattern in PSP: NORMAL PARIETAL METABOLISM (typical finding in CBD) and TALAMIC BILATERAL HYPOMETABOLISM (absent in MSA and CBD)
Fig. 42: FIGURE 42
Fig. 44: FIGURE 44
Hipertrophic olivary degeneration: prominence of the bulbar pyramids, leaning forward (→) widening of the pontine transverse fibers secondary to the flattening of the tegmen (-----) and to the horizontalization of the sup cerebel peduncles (→).

The broad of the decusation of the sup cereb peduncles (→) is reduced.
Fig. 46: FIGURE 46
Fig. 47: FIGURE 47
Conclusion

Degenerative brain diseases may often demonstrate characteristic imaging findings on conventional MR images aiding to confirm the clinical diagnosis, but usually are "late findings".

Advanced functional imaging (spectroscopy, diffusion tensor imaging, f-MRI and PET-FDG) provide suggestive information that may aid to support the suspected diagnosis, to classify the patients (allowing their inclusion into clinical trials to validate new therapies), and to monitor their evolution.

References


Personal Information