SPINAL CORD: DEMYELINATING DISEASE AND MYELITIS

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Non-neoplastic diseases of the spinal cord include a vast series of possible causes, the most frequent being ischemic, traumatic, degenerative, and inflammatory. The corresponding clinical presentation is often non specific, and neurological impairment is essentially related to the transverse extension of the lesion and the consequent involvement of central grey matter, ascending, and descending pathways, and to the height of the compromised level (cervical, dorsal, lumbar).

MRI is the only imaging tool able to offer a satisfactory visualization of the spinal cord and its related pathology.

The term myelopathy generally indicates every generic form of pathological involvement of the spinal cord.

Under this definition, even the category of myelitis is enclosed. Myelitis refers to every case of inflammatory etiology that affects the spinal cord. When inflammation involves a segmental extension (one-three metamers) the definition “transverse myelitis” is used. Under the definition of “radiculo-myelitis” are included forms of inflammatory involvement of spinal cord and nerve roots, that are mostly of infectious origin (HSV2, Tubercular, CMV, Staphylococcal), while the terms “(multi- or poli-)radiculo neuritis” is on a demyelinating base, and essentially from an autoimmune response. In this particular case the autoimmune attack is versus the peripheral myelin, while the central one is spared. Peripheral myel is formed by Schwann cells, and differs antigenically from the central one, produced by oligodendrocytes. Nonetheless, intermediate forms of demyelinating diseases involving both central and peripheral white matter have been described [1].

Three forms of multi-radiculo-neuritis are known: the classic pattern of acute demyelinating disease (Guillain-Barré syndrome), the ascending palsy variant (Landry syndrome) and a variant characterized by ophthalmoplegia, ataxia, and areflexia called Miller Fisher syndrome. In the first two cases, clinical presentation commences with progressive muscular weakness of extremities that may lead to paralysis. It spreads rapidly, ascending to involve the cranial nerves and the diaphragm. Clinical features are pain in the back and the legs and weakness beginning in the feet and legs and progressing upwards. The tendon reflexes are lost. The respiratory muscles are affected in about half the cases, and this puts the patient in vital danger. Recovery after a period varying from several weeks or months is the usual outcome. There is no specific therapy. Usually the spinal fluid contains an increased amount of protein but only rarely the cells are increased. A chronic, milder and rarer form of multi-radiculo-neuritis has been described under the name of CIDP (Chronic Inflammatory Demyelinating Polyneuropathy). In all the mentioned forms, MRI shows very peculiar patterns characterized by diffuse nerve roots enhancement, limited to the cauda equina or extended to the whole spinal cord, or even involving cranial nerves.

Among the pure forms of myelitis, Multiple Sclerosis represents the main cause.

Spinal cord lesions are found in a variable percentage between 50% and 90% of patients affected by MS, representing a frequent inability cause. Involvement is more frequent at cervical than thoracic...
level; conus is also involved. Lesions are asymmetric and regard more frequently posterior and lateral cordonal area and extended to less of the half of the area in the axial sections. Usually, less than two myelomers are involved.

Signal characteristics reflect those of encephalic lesions, as enhancement after Gadolinium injection; usually mass effect is poor, though it is possible that pseudo-tumoral lesions are observed.

In patients presenting spinal symptoms, the first exam being perormed is the spine MRI; if positive, (in 55% of cases), an MRI study of brain is needed to support diagnosis. [2] [3]

In spite of the presence of artifacts, particularly evident at thoracic level, using new techniques (phase arrayed coils, STIR sequences, double echo with cardiac gating) MRI sensitivity is increased with possibility of detecting lesions in more than 90% of subjects with MS. Spinal cord study is requested in those patients in which encephalic imaging is negative or in doubt and diagnosis is still lacking.

A very peculiar form of demyelinating disease specifically involving the spinal cord, is represented by Devic’s Disease, or Optic Neuromyelitis.

In this case usually brain MRI doesn’t show lesions; both symptoms and lesions are connected to spinal cord and optic nerves involvement.

Devic’s disease is a rare disorder which resembles MS in several ways, but requires a different course of treatment for optimal results. NMO has also been suggested to be a variant form of acute disseminated encephalomyelitis (ADEM). The likely target of the autoimmune attack at least in some patients with NMO has been identified; it is a protein of the nervous system called aquaporin-4.

Disease debut, characterized by wide pseudo-tumoral lesions, can represent diagnostic problem both in pediatric population and in the adult one for the risk to confuse lesions with high grade tumors, specially when only one lesion is present; this kind of lesion tends to decrease in its dimensions. [4][5]

Schilder disease (or diffused sclerosis) and the Balò one (or concentric sclerosis) are more rare.

Although (ADEM) is not a real MS varying form, it is strictly connected to its description both because it can mimic MS and also because final evolution to typical MS is registered in 29% of pediatric cases, with higher percentage in adult ones. [6] [7]

It represents a monophasic immuno-mediated demyelinating disease interesting mostly pediatric population, preceded only by a viral or bacteric infection or by vaccination.

Not rare also in adults, ADEM can mimic MS both at its beginning and months / years later.

Clinically, in childs the disease is characterised by not-specific symptoms such as cefalea, vomit, fever and lethargy (rare in MS) or by emi-paresis, ataxia and cranial nerves deficit. [8]

MS and ADEM can also coincide in imaging; ADEM is characterised by wide lesions, with poorly defined margins, asymmetric and distributed in supra and infra-tentorial white matter. Synchronous enhancement is usually considered as a typical aspect. Nevertheless, it has to be considered that some cases of ADEM without barrier damage or with different patterns of the lesion enhancement are described. [9]

Grey matter, thalami, basal ganglia involvement, often asymmetric is much more frequent in ADEM than in MS; relatively less involved the periventricular white matter and corpus callosum.

Spinal lesions tend to be wide and swollen, with similarity to the localization of NMO. It is very important their follow-up, because ADEM lesions tend to resolve partially or completely, leaving only modest signs of illness, usually without the appearance of new lesions.

Finally it is important to keep in mind that symptoms are not specific and the correct etiological diagnosis needs precise correlations of diagnostic imaging with clinical history, signs and symptoms,
laboratory (CSF) and instrumental (EMG, EP) findings in most cases. MRI remains the most sensitive diagnostic tool. In fact, radiological semiotics are always useful and often very specific in addressing the diagnosis: some fundamental concepts include the peculiar pattern of vasogenic edema, which, in opposition to what happens in the brain, tends to involve the central grey more than the peripheral white matter. Distribution pattern of the lesion can also often address to etiological diagnosis. In fact, lesions selectively involving posterior or lateral columns, asymmetrically, and sparing the central grey matter, are more typical expression of demyelinating-inflammatory diseases (Multiple Sclerosis, ADEM, Devic’s disease, LES, Behcet’s disease).

Within this group of diseases, short-segment and asymmetric lesions, are characteristic of multiple sclerosis. In contrast, a longitudinally extensive lesion, especially if it extends rostrally into the brain stem and is located centrally within the cord, is typical of optic neuromyelitis. Lesion enhancement after the administration of gadolinium, suggests acute inflammation of the spinal cord. Selective and symmetrical involvement of both posterior columns, extended for more than 2 myelomers, and possibly associated with involvement of lateral columns, is typical expression of combined sclerosis (B12 vitamin deficit). Selective involvement of the central grey matter is more likely expression of viral infection, and selective signal alteration of the anterior horns is strongly suggestive of poliomyelitis.

Ischemic lesions are also usually symmetrical, but they involve both grey and white matter, extending to the anterior 2/3 of the cord. Sometimes they remain limited to the central grey matter (pencil shaped softening).

Finally “advanced” MRI techniques can be used to obtain additional information regarding the nature and extent of structural damage of nerve tissue. They are usually in use in research or in clinical trial protocols. The use of techniques for post-processing and techniques such diffusion, tractography, functional imaging, magnetization transfer imaging, and future perspectives in the study of MS are extensively covered in a recent review of Bakshy and co-workers [10].

MR spectroscopy is used to measure several metabolites, such as N-Acetyl-Aspartate, Choline, Creatine, Glutamate, Glutamine and GABA; a decrease in NAA local concentration is associated with axonal/neuronal damage or dysfunction, choline is found increased during myelin breakdown, remyelination and inflammation. Creatine increases with cellular density; myoinositol, if found increased is suggestive of glial proliferation and astroglisis.

In active enhancing MS lesions, an increase in creatine, choline, myoinositol and glutamate levels can be observed, while NAA concentration can be found slightly decreased or clearly low. Differently, in chronic non-enhancing lesions, NAA is found strongly reduced, glutamate is normal and myoinositol is increased.

Also in normal-appearing white matter (NAWM) similar values of metabolites can be found, especially for those concerning NAA; using MR spectroscopy, thus, is possible a more precise valuation of disease extension; this kind of evaluation can be perfmormed also in spinal cord MRI, which is a fundamental part of imaging study in MS patients, because often spinal cord lesions are more strictly correlated to clinical disability than brain ones. Specially, reduced NAA in spinal cord spectroscopy has been shown to be associated with axonal metabolic dysfunction and reduced axonal density.

By applying diffusion-weighting magnetic fields in many directions (diffusion tensor MRI, DTI) is made possible to reconstruct and study the pathways of the major white matter bundles.

Tracking fibers is difficult through MS lesions, because of tissue disruption caused by disease, but atlas-based approaches could overcome this problem. Nevertheless, tractography of white matter in
sub-cortical regions is made difficult because of the complex connections, specially if not performed using high-field strenghts. In recent DTI studies, a reduced fractional anistropy suggesting axonal degeneration and myeline breakdown has been reported. Finally, an association between the average cord fractional anistropy and clinical disability seems to be confirmed by first investigations.[11]

REFERENCES


