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Contribution of imaging to the diagnosis of optic neuropathies

Place de l'imagerie dans le diagnostic des neuropathies optiques

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ABSTRACT

Magnetic resonance imaging is the modality of choice to investigate patients with central nervous system diseases. In patients with optic neuritis, early MRI allows positive and differential diagnosis, and gives arguments for prognosis. The technique should be adapted for the exploration of visual pathways. Inflammatory optic neuritis is characterized by an early signal abnormality within the optic nerve, whereas optic nerve MRI is normal in the early phase of ischemic optic neuropathy. MRI also detects compressive and infiltrative lesions. Meningiomas are characterized by abnormalities within the peri-optical spaces, whereas a global increase in the size of the optic nerve is in favor of a glioma.

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RÉSUMÉ

L'imagerie par résonance magnétique (IRM) est l'examen de choix pour l'exploration des affections du système nerveux central. Devant une suspicion de neuropathie optique, l'IRM, pratiquée tôt, permet d'orienter le diagnostic positif et différentiel, et fournit des éléments pronostiques. La technique doit être adaptée à l'exploration des voies visuelles. La mise en évidence d'un hypersignal du nerf optique oriente vers une neuropathie optique inflammatoire, alors que l'IRM précoce est habituellement normale si le mécanisme est ischémique. L'IRM peut révéler par ailleurs des lésions compressives ou infiltrantes. Les méningiomes sont caractérisés par des anomalies des espaces péri-optiques, alors qu'une augmentation globale de la taille du nerf optique et des espaces péri-optiques oriente vers un gliome.

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1. Introduction

Magnetic resonance imaging (MRI) of the optic nerves is a complementary tool to the clinical assessment that helps establishing the diagnosis, and often contributes to defining the prognosis of patients with optic nerve dysfunction. Besides, MRI of the brain checks for asymptomatic lesions that can help in determining the cause of optic neuropathy. In inflammatory optic neuritis, MRI is very sensitive (over 90%) at

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detecting symptomatic lesions (Deschamps et al., 2002). Causes of optic neuropathies include inflammatory (CIS, multiple sclerosis, vasculitis and systemic diseases, orbitopathies, uveitis, post-vaccinal, infectious, traumatic, post-radical), ischemic, compressive and infiltrative, glaucoma-related, toxic and carential, metabolic and hereditary, paraneoplastic. The aim of this review is to describe different abnormalities that are detected on MRI, and how MRI contributes to the diagnosis. Different situations are classified according to the detection of abnormalities in the optic nerve or brain on MRI.

2. Anatomy and MRI imaging of the optic nerve

The optic nerve is composed of axons that are projections from the retinal ganglion cells located in the sensory retina. It is an extension of the diencephalon, and thus centrally myelinated and covered with meninges. It is divided into four segments:

- intraocular, the shortest (1 mm);
- intraorbital the longest, extending from the posterior sclera (lamina cribrosa) to the optic canal and surrounded by the optic sheath and cerebrospinal fluid;
- intracanalicular, with a relatively fixed position and susceptibility to injury;
- intracisternal extending to the optic chiasm (Fig. 1).

The best axial plane to visualize optic nerve is the neuro-optical plane. Coronal plane is always necessary. Other planes are often used such as sagittal or neuro-optical transhemispheric, particularly when exploring tumors. In some circumstances dynamic MRI may help. Image thickness should not exceed 4 mm (usually 3 mm).

Sequences always include fat saturation to remove orbital fat such individualizing different components of the optic nerve (Figs. 2a and b). This is easily obtained with the T2 weighted (T2w) short tau inversion recovery (STIR) sequences (Fig. 2a). In other cases, fat saturation is applied on spin echo T2 weighted images (T2wi). Whenever it is necessary fat saturated spin echo T1 weighted (T1w) sequences with and without gadolinium infusion should be performed. Normal optic nerve appears, like white matter with low signal on T2wi, isosignal on T1 weighted images (T1wi). It is surrounded by peri-optic spaces containing CSF that appears with high signal on T2wi and low signal on T1wi. The axial thickness of the optico-perioptical package is about 5 mm (3 mm for the diameter of the optic nerve and 2 mm for the peri-optical spaces) (Mahima et al., 1996).

3. Clinical situations

In a patient presenting with visual loss, different situations may be described according to the presence or absence of abnormalities in the optic nerve(s), or in the brain.

If a clinical picture is suggestive of optic neuritis, an early MRI may or not show abnormalities in the optic nerve. In the absence of hypersignal on T2wi, T1 pre- and post-gadolinium infusion should be performed (Youl et al., 1991). If a hypersignal is observed on T2wi with or without enlargement of the optic nerve, or gadolinium enhancement, the diagnosis of inflammatory optic neuritis is certain (Fig. 2c). In the absence of optic nerve abnormalities at this early phase (less than 2–3 weeks), the visual loss may not be related to an optic nerve dysfunction (sero-central chorioretinitis for example). In the other cases, this corresponds to an ischemic optic neuritis, or to another less common etiology. If the MRI is performed several weeks after clinical onset, any abnormality observed inside the optic nerve (hypersignal on T2wi, or atrophy) is not specific (Fig. 2g), as in ischemic ON abnormalities of the optic nerves are delayed. In case of abnormalities in the ON, brain abnormalities suggestive of MS have a prognosis value, as the risk over 15 years increase from 20% to over 60% (ONTT, 2008).

In some cases, the abnormalities of the optic nerve(s) are so important with diffuse involvement of the peri-optical spaces that the picture is called perineuritis. This is an unusual form of orbital pseudotumor that has a different prognosis with dramatic response to steroid therapy. The optic nerve itself is rarely involved and brain imaging is usually normal. Some inflammatory and infiltrative disorders need to be considered such as sarcoidosis leukemia and lymphoma. The prognosis for optic perineuritis is excellent with no risk of progression to MS (Purvin et al., 2001).

Optic neuropathy may be observed in auto-immune and systemic diseases. In sarcoidosis, optic neuropathy may be acute, sub-acute or chronic corresponding to neuro-retinitis, inflammatory or granulomatous, or even ischemic ON. A suggestive aspect is the opto-chiasmatic arachnoiditis (Hosseini and Tourbah, 1999). Devic Neuromyelitis Optica may start with optic neuritis. Normal brain MRI or suggestive abnormalities (peri-ventricular symmetrical, enhanced, aera postrema or hypothalamus involvement...) and positive NMO-IgG may help establishing the diagnosis or classifying the patient in a group at risk. In Lupus erythematosus, ON is rarely the presenting symptom. On MRI increased volume of

Fig. 1 – Normal anatomy of the visual pathways on axial neuro-ocular plane. High resolution T2 weighted sequence with contrast inversion showing the optic nerves and the peri-optical spaces. Three portions of the optic nerve may be individualized, the intraorbital (arrow) and intracanalicular (arrowhead) in a plane, and intracisternal (double arrow) in the next plane.
the optic nerve and chiasma with gadolinium enhancement may be present. Other systemic diseases include Wegener disease (granulomas), Sicca syndrome, Rheumatoid Polyarthritis and Behcet disease.

In some cases ON dysfunction appears in patients with toxic metabolic or hereditary diseases. Metabolic and hereditary diseases with optic nerves involvement include mitochondrial cytopathies (Leigh, MELAS, MERRF, NOHL), biotinidase and pyruvate dehydrogenase deficiencies, and leucodystrophies (adrenoleucodystrophy, metachromatic leucodystrophy, Krabbe disease). In the chronic phase bilateral atrophy with diffuse hypersignal on optic nerves and absence of peri-optical spaces is usually observed (Fig. 2f), with a transversal T2 hypersignal in the chiasma on coronal planes (Fig. 3). At the acute phase, especially in Leber hereditary optic neuropathy NOHL, slight and diffuse hypersignal appears on T2wi extending from the orbital segment of the optic nerve to the chiasma.

Compressive and infiltrative lesions of the optic nerves are easily recognized with an appropriate technique. During the acute state compressive lesions can be confused with optic neuritis. It is important to evaluate if the lesion involves the optic nerve itself or the peri-optical spaces. In the first case the aspect is suggestive of glioma (Dutton, 1994). It appears like an expansile mass of the optic nerve (Fig. 2k) that can occur anywhere from the globe to the optic tracts with iso- to hypointense to brain on T1wi and hyperintense on T2wi, and variable contrast enhancement. Widening and kinking of the optic nerve are often seen. The MRI helps in determining the full extent and rate of change of the tumor, which have a direct impact on its the management. It has been reported that some optic gliomas undergo spontaneous regression, often with improvement in visual function (Parsa et al., 2001).

The primary involvement of the peri-optical spaces is observed in meningiomas (Dutton, 1992) (Figs. 2h–j) or other infiltrative processes such as leukemia and lymphoma. Optic

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Fig. 2 – MRI of the optic nerve on T2 weighted images (a–h, k) and T1 weighted images after gadolinium infusion (i, l). Coronal planes (a, c, d, f–h, j, k), axial planes (b, e, i), sagittal plane (l). Normal optic nerves on T2 weighted images (a and b); hypersignal of the left optic nerve in a patient with inflammatory optic neuritis (c); papilledema with bilateral enlargement of peri-optical spaces (d and e); bilateral (f) and left (g) atrophy; optic nerve meningioma (h–j) with a left sided tram-track sign on axial plane (j); glioma of the optic nerve (k and l).
sheath meningiomas are benign tumors that are typically relatively isointense to the brain on T1- and T2wi with striking enhancement (Fig. 2i). The tumor often appears around the optic nerve in a tram-track configuration (Atlas and Galetta, 1996) (Fig. 2i) that differentiates it from an optic glioma. The tram-track has been described as a sign of optic perineuritis (Atlas and Galetta, 1996; Purvin et al., 2001), optic nerve sarcoidosis (Carmody et al., 1994), chronic relapsing inflammatory optic neuropathy (Kidd et al., 2003). Some meningiomas have a more globular appearance or cystic components (Lindblom et al., 1992).

4. Other MRI appearance of ON dysfunction

In papilledema due to raised intracranial pressure, a bilateral enlargement of peri-optical spaces is observed (Gass et al., 1996; Imamura et al., 1996) (Figs. 2d and e). It is associated to indirect signs of intracranial hyperextension: small ventricles, empty sellae, poorly visible cortical sulci. ON may be observed in Basedow disease. It is usually due to compression of the optic nerve by hypertrophic intraorbital muscles. Vascular compressions are also possible. The presentation may be acute in aneurysms (internal carotid), or chronic with progressive visual field defect and atrophy.

5. Conclusion

MRI is the indispensible complementary investigation to clinical assessment of patients with optic neuropathy, and should be performed at the acute phase whenever this is possible. The technique should be adapted to the exploration of the visual tracts. It is an important step in defining the diagnosis and determining the prognosis. After excluding a compressive or infiltrative cause, the presence of an early abnormality of the optic nerve is in favour of inflammatory optic neuritis.

Disclosure of interest

The author declares that he has no conflicts of interest concerning this article.

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