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Imaging of the pre-chiasmatic optic nerve

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Abstract Damage to the optic nerve (ON) is characterised by a reduction in visual acuity. Pre-chiasmatic lesions to the optic nerve may be of traumatic, congenital, tumoral (meningioma, glioma), inflammatory or vascular origins. In all cases, MRI is the choice means of exploration, carried out with axial and coronal sections with a thickness of 2.5–3 mm and T1 and T2-weighted spin echo sequences. The coronal sections may be carried out with fat signal saturation for an elective study of the size of the retrobulbar portion of the ON.

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The optic nerve (ON) is part of the central nervous system. The axons of the retinal ganglion cells converge at the optic disk and cross the sclera to form the optic nerve. The orbital segment goes from the posterior part of the eyeball to the orbital apex. In this segment, the nerve is clearly distinguished by the MRI and is surrounded by cerebrospinal fluid and a peripheral dural sheath.

The ON is fully visible in this section in T2-weighted coronal sections with fat signal saturation or in the axial plane on sections carried out in a neuro-ocular plane.

Morphological data derived from CT-scan imaging [1] or MRI [2] in the adult or child [3] is rare. In the adult, the mean diameter of the ON—ON sheath complex in the axial plane, with T1-weighting measured half-way between the posterior part of the eyeball and the orbital apex, is 4.4 mm (3.4–5.5 mm) without a significant difference in the size according to the side or sex [2].

The intracanal segment, in the optic canal, over a length of 5 mm and a diameter of 3 to 4 mm, leads the ON accompanied by meningeal sheaths from the orbit to the suprasellar region. The intracranial segment goes from the optic foramen to the chiasm over about 10 mm, directed upwards, behind and inwards, surrounded by pia mater. Upon exiting from the optic canal, the ON crosses over the internal carotid artery and passes below segment...

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A1 of the anterior cerebral artery. The optic nerves converge on the median line to form the optic chiasm where the fibres from the nasal hemiretina cross the median line, whereas those coming from the temporal hemiretina continue their homolateral pathway.

**Clinical semiology**

Damage to the ON is characterised by a reduction in visual acuity in the form of a central scotoma and/or a quadratic sector amputation of the visual field. Installation of the visual disorders varies: when slowly progressive, they suggest a tumoral disease; when the installation is sudden, they call to mind an inflammatory process.

**Means of exploration**

Study of the ON calls on sonography (useful to explore lesions of the eyeball and their extensions to the ON head). By backscatter X-ray, carried out in helicoidal acquisition with multi-planar reconstructions, 0.5 mm sections, with study in bone and tissue window. The angioscan may be of value in the study of tumoral lesions and vascular malformations of the orbit.

MRI is the choice means of exploration and should be carried out according to the guidelines provided by the SFR, carried out with axial and coronal sections with a thickness of 2.5—3 mm and T1 and T2-weighted spin echo sequences. The sections may be carried out with fat signal saturation for an elective study of the size of the retrobulbar portion of the ON.

The T1 sequences after intravenous injection of gadolinium should be carried out in the axial and coronal planes with fat signal saturation for an elective view of the enhancement of the ON, meningeal sheaths, retrobulbar content of the orbit, musculo-aponeurotic cone and lacrimal glands.

3D TOF MR or T1-weighted dynamic angiography after intravenous injection of gadolinium is proposed in case of suspicion of fast-flow vascular malformation.

An MRI encephalographic (at least flair sequence and 3D T1 gadolinium) is systematically carried out in case of inflammatory, infectious or tumoral lesions of the anterior visual pathways.

**Traumatic lesions of the optic nerve**

Traumatic lesions of the optic nerve result from penetrating wounds of the orbit or traumatic lesions of the base of the skull with the entry of bone splinters in the ON. The most vulnerable zone is the passage of the ON in the optic canal. The CT-scan is the choice way to detect bone lesions of the orbit, the walls of the optic canal, (aliform process of the sphenoid, sphenoidal yolk), any intra-orbital foreign bodies, an intracranic haematoma or subperiosteal haematoma responsible for the indirect compression of the ON. MRI should not be proposed before eliminating the possibility of a metal foreign body by CT-scan [4].

**Congenital malformations—degenerative diseases**

**Colobomas**

This consists of a defect in the closing of the embryonic fissure or coloboma notch. This anomaly may be isolated or associated with anomalies of the iris, the lens and the three-membrane tunica of the eyeball. Colobomas account for 2% of all congenital ocular malformations whether isolated or associated with polymalformation syndromes.

In MRI, the retinal coloboma appears as a cystic dilation of the optic nerve head extending along the intra-orbital ON over a variable distance, with formation of a diverticulate liquid pocket that may compress the ON.

The detection of a coloboma should lead to the search for a CHARGE syndrome (Fig. 1) (choanal atresia, agenesis of the olfactory bulbs [5], malformation of the inner or outer ear [6], etc.).

**Dominant optic atrophy**

The hereditary optic neuropathies are degenerative diseases of the ganglion cells of the retina leading to a loss of axons of the ON. Dominant optic atrophy with an insidious beginning and slow evolution, and Leber’s hereditary optic neuropathy, with a sudden onset and rapid evolution are the two most common non syndromal forms [7]. The diagnosis is molecular (genes OPA1 and OPA3 and mutation on the mitochondrial DNA). In 10% of the cases, there is a multi-systemic impairment that may associate deafness, ataxia, myopathy, peripheral neuropathy, spastic paraparesis, SEP-Like syndrome and progressive bilateral external ophthalmoplegia.

The imaging tries to detect severe atrophy of the optic nerves (Fig. 2), anomalies in the white matter, the cerebellum and the presence of a lactate peak in spectroscopy.

**Leber’s hereditary optic neuropathy**

The origin is also mitochondrial by a deficiency in complex 1 of the respiratory chain by mutation of mitochondrial genes ND1, ND4 and ND6. The transmission is non-mendelian with variable penetrance. The loss of vision begins between the age of 18 and 35 years in men and towards menopause in women. A possible extra-ocular manifestation has been described: ataxia, deafness, dystonia, parkinsonian syndrome, cardiac conduction disorder (Wolff-Parkinson-White), myopathy, SEP-Like syndrome with impairment of the white matter in MRI [8].

**Tumours**

The most common tumours of the ON are glioma and meningioma. Other tumours are more rare and consist of haemangiblastomas, haemangiopericytomas and gangliogliomas.
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Figure 1.  Load syndrome. The axial sections passing through the orbits and petrous part of the temporal bone in T2-weighted 3D CISS (a and b) identify bilateral colobomas of the iris (a, tip of white arrow) and retina (a, tip of black arrow) as well as cochlear hypoplasia (a, white arrow) and agenesis of the semi-circular canals (b, tip of white arrow). The coronal sections on the olfactory bulbs (c and d) detect hypoplasia of the olfactory bulbs (white arrows).

Glioma of the optic nerve

This is a tumour in the child with a peak incidence between the age of 2 and 8 years. Gliomas of the ON are slowly evolving pilocytic astrocytomas. The sporadic forms are distinguished from the syndromal forms that occur within a type 1 neurofibromatosis (NF1).

The gliomas of the ON may develop on all intraorbital, intracanal or intracranial parts of the ON. They are characterised by a fusiform enlargement and a
Figure 2. Dominant optic atrophy. MRI in T1-weighted spin echo coronal sequences on the optic nerves (a) and optic chiasm (b), and T2-weighted spin echo coronal sequences with fat signal saturation on the optic nerves (c) and optic chiasm (d). The examination reveals severe atrophy of the optic nerves (tips of black arrows) and optic chiasm (tips of white arrows).

In MRI (Figs. 3 and 4), on the T1-weightings, the lesion is iso-hypointense when compared with the white matter or without mucinous or necrotic cystic component of very hypodense signal.

In the sporadic forms, a constant extension of the chiasm and the presence of cysts is noted in 54% of the cases [10]. In the syndromal forms associated with NF1, the impairment of the ON is isolated in 71% of the cases and a cystic component is not found. The tumoral proliferation is the cause of an elongation of the nerve with tubular enlargement and sinuosities. The enhancement is
variable and homogenous or heterogeneous. Within NF1, the bilateral nature of the glioma is characteristic as is the association with ‘‘unidentified white objects’’ (basal ganglia, arch, cerebellum), plexiform neurofibromas, sphenoid dysplasia and other brain tumours. The evolution is slow and regression possible. However, the evolution of glioma of the ON that occur outside of NF1 are prejudicial and propagate to the visual pathways and outside the brain parenchyma [4,11].

Meningioma of the optic nerve

The meningiomas of the optic nerve occur in the woman between the age of 30 and 50 years. The bilateral form is characteristic of neurofibromatosis type 2. It is possible to distinguish the primary forms from the arachnoid cells of the ON sheath from the secondary forms by extension of the ON sheath of parasellar meningiomas of the medial level.

They occur as a progressive reduction in visual acuity, exophthalmia, optical atrophy and retinal folds at the fundus of the eye. In imaging (Fig. 5), the appearance is either that of a fusiform enlargement or an excentric nodular formation next to the ON with or without microcalcifications (CT-scan). The T1 sequence after intravenous injection of gadolinium and fat signal saturation allows for a better evaluation of the extension of the tumour.

The ‘‘railway’’ appearance in MRI and CT-scan is suggestive: central hypointensity or hypodensity of the ON emphasised by the reinforcement of signal in parallel bands of the dura mater sheath of the ON after injection of contrast product. The effacement of the peri-optic liquid spaces, the thickening of the ON sheath and the hypointensity of the optic nerve are searched for in T2 coronal sections. The extension of the optic canal, at the optic groove and the contralateral extension are looked for in the coronal and axial planes in T1-weighting after injection of gadolinium and fat signal saturation.

Extension of the tumours of the eyeball at the optic nerve

Retinoblastoma in the child and malignant melanoma in the adult are the malignant tumours most commonly found. Sonography and MRI allow for tumoral staging and post-treatment monitoring [12].
Inflammatory disorders

Retrobulbar optic neuritis

From a clinical point of view, it is characterised by a sudden onset with a reduction in visual acuity (or rapidly progressive in 48 hours to 2 weeks) associated with retrobulbar pain. In 10 to 15% of all cases, visual disorders of variable severity persist.

In the acute phase, MRI in coronal T2-weighting with fat signal saturation detects the oedema of the ON in the form of a T2 hypersignal and segmental or total contrast
Figure 5. Meningioma of the optic nerve. Orbit MRI. The T2-weighted spin echo coronal sequences with fat signal saturation identify a tumoral formation enclosing and compressing the optic nerve (a, tip of black arrow), presenting an intrafascicular hypersignal in its intracranial portion (b, tip of white arrow). The sequences after injection of contrast product and fat signal saturation identify a homogenous enhancement of the mass (c, white arrow) with railway reinforcement of the sheaths of the optic nerve (d, black arrow).
enhancement of the ON may reach the optic chiasm. The associated signs consist of enhancement to the sheaths with sequestration of cerebrospinal fluid upstream from the constricted zones in 45% of the cases [8].

Study of the optic nerve (Fig. 6) should be combined with an encephalic and medullar MRI to detect the hypersignals of the white matter in 77% of the cases, a lesion activity with rupture of barrier in 26% of the cases and medullar T2 hypersignals in 26% of the cases.

The risk of developing multiple sclerosis (MS) is 40% in 10 years and 50% in 20 years [13].

The diagnosis of MS by MRI no longer requires a reference examination on D30 with an increase in the lesion load on D180 [14]. The diagnosis is based on the initial MRI examination if the criteria of spatial and temporal dispersion are united:

- spatial dispersion: two lesions or more in asymptomatic T2 hypersignal distributed on at least two of the four target locations (periventricular, juxta-cortical, infra-tentorial or medullar);
- temporal dispersion: asymptomatic enhancement [15].

However, retrobulbar optic neuritis is not specific of MS and may be seen in auto-immune vasculatititis, neurosarcoïdosis or be associated with an infectious disease (cat scratch disease).

Devic disease or neuromyelitis optica

It associates a picture of retrobulbar optic neuritis and myelitis. The detection of an autoantibody (NMO-IgG) directed against aquaporine 4 demonstrated that Devic disease was an autonomous auto-immune disease with mainly humeral mediation. The diagnostic criteria of neuromyelitis optica [16] associate acute transverse myelitis and optic neuritis and at least two of the following criteria:

- normal brain MRI (or not evocative of MS);
- medullar MRI with a lesion of three or more vertebral segments;
- positive NMO-IgG serology.

Inflammatory disease of the orbit and the anterior visual pathways

Inflammatory disease of the orbit and the anterior visual pathways [17]: are either idiopathic ("pseudotumour" of the orbit) or secondary to a systemic disease (vasculitis, granulomatosis, lymphoproliferative disorder).

Impairment of the ON characterised by lymphoplasmyocytary infiltration of the meningeal sheaths is an integral part of a more extensive picture in which all of the constituents of the orbit may be involved: muscular impairment (myositis), impairment of the posterior pole of the eyeball (episcleritis), infiltration of the retrobulbar fat, impairment of the lacrimal glands (dacryoadenitis).

The inflammatory lesions of the orbit may be diffuse or monofocal (myositis, dacryoadenitis or perineural inflammation of the ON). The radiological picture is non-specific. However, a great many differential diagnoses must be eliminated before making the diagnosis of idiopathic inflammation of the orbit (orbital pseudotumour): sarcoidosis, Wegner’s granulomatous, Crohn’s disease, autoimmune thyroiditis, disseminated lupus erythemateous, Erdheim-Chester disease, histiocytosis X, myofibroblastic tumour.

The inflammation may involve the cavernous sinus and create the Tolosa-Hunt syndrome (Fig. 7) characterised by painful ophthalmoplegia with paralysis of the ocular motor nerves, impairment of V1 and the pericarotid plexus. Impairment of the ON, by extension of the inflammatory process to the orbit apex, is rare. The forms benefiting from a histopathological diagnosis demonstrate the presence of an inflammatory granuloma, which has to be detected by MRI in order to establish a formal diagnosis [18].

Nevertheless, it is necessary to keep in mind the limits of MRI in the aetiological diagnosis of lymphoproliferative lesions of the orbit based on MRI data. Therefore, the lymphoproliferative diseases of the orbit may not be differentiated by MRI (lymphoma, reactive lymphoid hyperplasia, idiopathic or secondary inflammatory disease of the orbit and IgG 4 related orbital disease). The final diagnosis is based on the use of immunohistochimical techniques and the techniques of molecular biology on biopsy samples.

Vascular disease

Ischemic optic neuropathy

This disease occurs either in a non-arteritic context (non contributive imaging) or in a context of vascularitis (Horton’s temporal arteritis). 3-tesla imaging of the vascular wall after injection of gadolinium is indicated before starting treatment with corticoids. The profile of the enhancement of the vascular lining differentiates an unstable atheromaous plaque (focal, excentric enhancement) from inflammatory enhancement (diffuse, concentric enhancement) and an intracranial dissection [19].

Carotid cavernous fistula

Direct arteriovenous shunts (traumatic) or indirect arteriovenous shunts (dural fistula); of multipedicular feeding by

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**Figure 6.** Right retrobulbar optical neuritis. Cerebro-orbital MRI. The T2-weighted spin echo sequences, in axial sections of 3 mm reveal a very slight increase in the size of the right intracranial optic nerve (a, tip of white arrow). The T1-weighted spin echo axial sequences after injection of gadolinium and fat signal saturation identify a homogenous enhancement of the intracranial optic nerve without extension to the optic chiasm or the orbital optic nerve (b, tip of black arrow). The T2-weighted spin echo coronal sections detect the increase in the size of the intracranial optic nerve in hypersignal (c, white arrow) and the T1-weighted coronal sections after intravenous injection of contrast product identify an enhancement of the intracranial portion of the nerve (d, black arrow). The encephalic acquisition in T2-weighted FLAIR axial sections discerns the lesions in hypersignal of the white paraventricular matter (e) and the oval centre (f).
Figure 7. Tolosa-Hunt syndrome. MRI of the cavernous sinus. On the T2-weighted spin echo sequences, in the axial plane, the right cavernous sinus appears thickened, in T2-weighted hyposignal (a, tip of black arrow). This aspect of the cavernous sinus is associated with an obliteration of the intracavernous venous spaces on the right side, detected in the T2-weighted spin echo coronal sequences after fat signal saturation (b, tip of black arrow). The sheath of the optic nerve is dilated (c, tip of white arrow). The T1-weighted axial sections after injection of contrast product and elimination of the fat signal allow for the identification of a homogenous enhancement of the right cavernous sinus (d, tip of black arrow).
the meningeal branches of the carotid siphon and external carotid territory and the cavernous sinus. The fistula is responsible for a reduction in visual acuity by venous blocking of the orbit with an increase in eye tension. The reduction in visual acuity is associated with exophthalmos, chemosis, conjunctival oedema and intracranial pressure. The MRI (or CT) diagnosis is based on the dilation of the superior ophthalmic vein, the expansion of the cavernous sinus, the visualisation of the branching of the arterial pedicles feeding in the cavernous sinus on the native images in TOF 3D MRA, and finally on the arteriovenous shunt confirmed on the dynamic T1 angioRM after intravenous injection of gadolinium.

Intracranial aneurysm

Two aneurismal locations with direct repercussions on the ON are possible: para-ophthalmic aneurysms are located on the carotid end, developing upwards and within at the contact with the ON and aneurysms of the anterior communicating artery when the aneurismal sac is directed downwards in the pre-chiasmatic cistern compressing the median side of the ON.

The diagnosis is suggested by the data from the MRI imaging in sections (vascular structure with empty rapid flow of signal in T1 and T2). Modifications in the intra-sac signal related to spontaneous thrombosis is searched for: hypersignal if recent thrombus, lamellar structuration and hypo-isointensity in onion bulb if old thrombosis. The diagnosis of aneurysm is confirmed by MRA or angioscan.

Idiopathic intracranial hypertension

Characterised by disabling headache and visual disorders, it is most often found in obese women of childbearing age. The evolution may be prejudicial and lead to optical atrophy. The diagnosis is based on the signs (papillary oedema) and symptoms reported by intracranial hypertension (ICH) (pulsatile tinnitus, diplopia, cognitive deficiency), the increase in the pressure of the opening of the CSF in lateral position greater than 20 cm of water, a cytochemical analysis of the normal CSF and exclusion by MRI of any other cause of intracranial hypertension (in particular thrombosis of the intracranial venous sinuses). The risks suggesting idiopathic ICR are: a flattening of the posterior pole of the eyeball, a protrusion and contrast enhancement of the ON head, an enlargement...

![Figure 8](image_url). Benign intracranial hypertension. Cerebro-orbital MRI. On the axial (a) and coronal sections (b) T2-weighted turbo spin echo, the sheath of the optic nerves is dilated (tips of black arrows). In sagittal section, the sella turcica assumes the appearance of “empty sella turcica” (c, black arrow). The angioMR venous sequence allows for identification after intravenous injection of contrast product, by MIP in axial (d) and fronto-sub-occipital view (e), bilateral stenosis of the lateral sinuses (tips of white arrows).
of the ON sheaths, an empty sell syndrome [20] (Fig. 8). The recommended MRI-venous technique is an angioMR sequence after injection of gadolinium and cancellation of the signal from the cerebral parenchyma that allows for a 3D study of the intracranial venous network. The idiopathic ICH by steno-occlusive lesion of the lateral sinus is debated. However, stenosis of an intracranial venous sinus with pressure gradient greater or equal to 10 mmHg may benefit from the insertion of a stent [21].

Conclusion

The causes of impairment of the pre-chiasmatic portion of the optic nerve are numerous, traumatic, inflammatory, tumoral, malformative or vascular. The study of the nerve relies on MRI comprising dedicated, millimetric sequences in the axial and coronal plane. On the basis of the diagnostic hypotheses, this examination is completed by an encephalic, medullar or intracranial vascular study involving the arterial or venous sectors.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

References