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The ROYAL COLLEGE of OPHTHALMOLOGISTS

REVIEW ARTICLE

Neuroradiology for ophthalmologists

Bayan Al Othman[1] · Jared Raabe[2] · Ashwini Kini[1] · Andrew G. Lee[1,3,4,5,6,7]

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Abstract

This article will review the best approaches to neuroimaging for specific ophthalmologic conditions and discuss characteristic radiographic findings. A review of the current literature was performed to find recommendations for the best approaches and characteristic radiographic findings for various ophthalmologic conditions. Options for imaging continue to grow with modern advances in technology, and ophthalmologists should stay current on the various radiographic techniques available to them, focusing on their strengths and weaknesses for different clinical scenarios.

Introduction

Modern imaging technology continues to advance the boundaries and increase the options available to physicians with respect to neuroradiology. In ophthalmology, the most common studies employed are computed tomography (CT) and magnetic resonance imaging (MRI). There are several different types of protocols that provide unique advantages and disadvantages depending on the clinical scenario. This article endeavours to review those options and discuss how they are best employed to evaluate a variety of specific ophthalmologic conditions.

✉ Andrew G. Lee
aglee@houstonmethodist.org

1 Department of Ophthalmology, Blanton Eye Institute, Houston Methodist Hospital, 6550 Fannin Street, Houston, TX 77030, USA

2 University of Texas Medical Branch at Galveston, School of Medicine, Galveston, TX, USA

3 Departments of Ophthalmology, Neurology, and Neurosurgery, Weill Cornell Medicine, 1305 York Ave, New York, NY 10021, USA

4 Department of Ophthalmology, University of Texas Medical Branch, 700 University Blvd, Galveston, TX 77555, USA

5 University of Texas MD Anderson Cancer Center, 1515 Holcombe Blvd, Houston, TX 77030, USA

6 Texas A and M College of Medicine, 8447 Bryan Rd, Bryan, TX 77807, USA

7 Department of Ophthalmology, The University of Iowa Hospitals and Clinics, 200 Hawkins Drive, Iowa City, Iowa 52242, USA

Computed tomography (CT)

CT imaging reconstructs a three-dimensional image made of many conventional x-ray images. The conventional x-ray images are ordered in tomographic slices that have been computerized so a viewer can scroll through the images, analyzing sequential cross-sections [1]. Density is the primary characteristic that determines image appearance on a CT scan. As with traditional x-rays, tissues appear on a grey scale ranging from white (i.e., hyperdense tissues such as bone) to black (i.e., hypodense materials such as air). Intermediate densities appear various shades of grey. CT is the imaging modality of choice in a variety of clinical circumstances in ophthalmology, especially when identifying bony defects or haemorrhage is of high priority. This includes acute trauma (e.g., orbital fracture), haemorrhagic lesions (e.g., subarachnoid or intraparenchymal haemorrhage), or calcified lesions (e.g., meningioma, craniopharyngioma, retinoblastoma, and optic disc drusen) [2]. In addition, although MRI is the preferred imaging modality for soft tissue including brain, CT may be indicated when MRI is contraindicated (e.g., ferromagnetic metal implants or foreign body present) or is not tolerated (e.g., claustrophobia, inability to remain still) [2, 3]. The sensitivity and specificity of CT may also be improved through the addition of intravenous contrast material. Contrast may be contraindicated, however, if a patient has renal disease or an allergy to iodine [1].

Magnetic Resonance Imaging (MRI)

MRI is an imaging modality that, unlike CT, does not use radiation. Instead, it relies upon the interactions of protons

within a strong magnetic field and the principles of nuclear magnetic resonance to create a three-dimensional image [1]. Unlike CT where density is the basis of the imaging, MRI depends on intrinsic imaging characteristics of hydrogen protons within individual tissues. MRI can be performed using a variety of different sequences that each highlights a different aspect of the imaged tissue by weighting the study (e.g., T1-weighted, T2-weighted). In T2-weighted images, fluid (e.g., CSF) appears hyperintense. Fluid attenuated inversion recovery (FLAIR) sequencing is most often applied to T2-weighted images and functions to suppress the signal of the CSF, darkening it. This allows better differentiation between pathologic hyperintensity and the CSF. In T1-weighted images, adipose tissue appears hyperintense (bright white), while fluid (e.g., cerebrospinal fluid (CSF), oedema) appears hypointense (dark). Fat saturation sequences are most commonly applied to T1-weighted images. These most commonly suppress the normally hyperintense signal of adipose tissue on T1-weighted images, which allows for better differentiation of pathologic T1 hyperintensity. Other commonly used sequences include diffusion weighted imaging (DWI) and can show restricted diffusion of water (e.g., in hyperacute infarct, in different types of brain oedema, and in hypercellular tumours). Specifically, DWI sequences help to differentiate oedema secondary to acute ischaemia from cytotoxic or vasogenic oedema or other chronic T2 hyperintensities. Orbital MRI is also often utilized, which should include both T1 and T2 weighted sequences with and without fat suppression and gadolinium contrast administration, as well as DWI. It should include both axial and coronal studies [4].

Magnetic resonance angiography (MRA) and magnetic resonance venography (MRV)

MRA and MRV use several techniques to create the images that depend on vascular flow physiology. MRA is considered a non-invasive imaging modality for medium and large size arterial vessels and MRV is used in patients with venous disease (e.g., in cases of papilledema to exclude venous sinus thrombosis). Gadolinium contrast can be given in both MRA and MRV but flow related non-contrast MRA and MRV can also be performed. If a vascular abnormality is suspected, MRA or MRV can be added.

Computed tomography angiography (CTA) and computed tomography venography (CTV)

CTA unlike MRA requires iodinated contrast dye for direct visualization of vessels. CTA is a rapid and sensitive tool for detection of vascular lesions (e.g., aneurysm or stenosis). Like MRV, CTV is another imaging modality used to look at the cerebral venous system (e.g., venous sinus thrombosis).

Catheter angiography

Catheter angiography is considered the gold standard for cerebral vascular abnormalities, it includes iodinated radiodense contrast dye injection and construction of the images by a technique called digital subtraction angiography to reduce the artefacts caused by the bony skull. The injected dye helps to outline the column of blood within the vessels and can show aneurysms, stenosis, vascular malformation, or dissection. Some of the procedural complications include dye reactions related to the contrast dye content which could also occur with contrast MRI/MRV and CT scans, haematoma at the puncture site, vasospasm, or emboli leading to ischaemia.

Specific neuro-ophthalmologic pathologies with common presentations and imaging of choice

Table 1 Summary of various neuro-ophthalmologic pathologies, including their signs and symptoms, imaging of choice, and common imaging findings.

Pathology	Signs/symptoms	Imaging of choice	Common findings
Orbital cellulitis	Blurry vision, chemosis, proptosis, painful ophthalmoplegia	Orbital CT scan	EOM enlargement, orbital fat enhancement, proptosis
Thyroid eye disease	Proptosis, lid lag, lid retraction, diplopia	Non-contrast orbital CT scan	EOM enlargement, orbital fat expansion, proptosis
Orbital inflammatory pseudotumor	Diplopia, pain, proptosis	Orbital CT or MRI	Enlargement or enhancement of EOMs, optic nerve, lacrimal gland, or orbital fat
Horner syndrome	Ptosis, miosis, anhidrosis	MRI/MRA or CT/CTA with and without contrast of the brain and neck (extending to the level of T2)	Any disruption of the sympathetic chain (e.g., Pancoast tumour, spinal cord lesion, ICA dissection)
Nystagmus of unknown origin	Varies with aetiology	MRI of brain and brainstem with and without contrast	Varies with aetiology
Optic neuropathy	Decreased visual acuity, rAPD, visual field defects, dyschromatopsia	MRI of brain and orbit with and without contrast (include T1 and T2 FLAIR sequences)	Demyelinating, inflammatory, infiltrative, or mass lesions, signs of increased intracranial pressure
Multiple sclerosis associated optic neuritis (MS-ON)	Pain, vision loss, rAPD, dyschromatopsia	MRI with and without contrast (include T1 and T2 FLAIR sequences)	Demyelinating white matter lesions

Table 1 (continued)

Pathology	Signs/symptoms	Imaging of choice	Common findings
Neuromyelitis Optica	May mimic MS-ON, but may progress more quickly and more commonly bilateral	MRI with and without contrast (include T1 and T2 FLAIR sequences)	Bilateral or longitudinal ON enhancement, chiasmal enhancement. Longitudinally extensive (>3 vertebral segments) transverse myelitis.
Anti-myelin oligodendrocyte glycoprotein (MOG) IgG antibody associated optic neuritis	recurrent optic neuritis or acute disseminated encephalomyelitis	MRI with and without contrast (include T1 and T2 FLAIR sequences)	Longitudinal enhancement of optic nerve, optic nerve sheath enhancement
Pituitary adenoma	Hyperprolactinemia, visual field defects, hypercortisolism, other endocrine abnormalities	MRI with contrast enhancement	Usually appear hypointense on T1, macroadenomas may appear as heterogeneous "snowman-shaped" on T2
Pituitary apoplexy	Hyperprolactinemia, visual field defects, hypercortisolism, other endocrine abnormalities	Non-contrast CT of the sella with thin slices	Pituitary enlargement or haemorrhage
Craniopharyngioma	Pituitary axis abnormalities	MRI with and without contrast	Cystic and solid components
Meningioma	Varies with location	CT or MRI with and without contrast	Often isodense/isointense with homogenous contrast enhancement, enhancing dural tail, optic nerve sheath enhancement
Optic pathway glioma	Associated with neurofibromatosis type 1	Serial MRIs with contrast to track progression	Fusiform enlargement and enhancement of optic nerve parenchyma with variable cystic change and variable enhancement.
Increased intracranial pressure	Headache, tinnitus, diplopia, blurry vision	CT and MRI of brain and orbit with and without contrast, MRV	Empty or partially empty sella, flattening of the globes, fluid in the optic nerve sheath, narrowing of transverse sinuses

Ocular motor cranial nerve palsies

When a patient presents with a suspected cranial nerve palsy, although a CT scan can be performed in the acute setting, brain and orbital MRI scan with and without contrast to follow the path of the nerve in question is usually the preferred approach to neuroimaging. For a third cranial nerve (CN III) palsy with pupil involvement, a posterior communicating artery aneurysm should be ruled out. To best accomplish this, a non-contrast CT of the head is indicated to look for a sub-arachnoid haemorrhage. Next, a CTA should be ordered to visualize the vasculature. If these are both negative, it is reasonable to proceed to a cranial and orbital MRI with gadolinium, as it offers superior visualization of other non-aneurysmal causes of a third nerve palsy [1]. If the CT, CTA, and MRI are all negative, but the patient is demonstrating signs and symptoms that strongly suggest aneurysm (e.g., severe headache, vomiting), then a standard catheter angiogram may still provide value for aneurysm detection. A sixth cranial nerve palsy with clivus bone involvement (e.g., chordoma, meningioma) is another situation that may require CT imaging in addition to MRI with contrast. Sixth cranial nerve palsy can be associated with idiopathic intracranial hypertension (IIH) which have radiographic signs on MRI like empty or partially empty sella, flattening of the posterior globe and fluid in the optic nerve sheaths. A CT scan can complement the MRI series to better visualize bony involvement [5]. Fourth nerve palsies are generally better imaged with cranial/orbital MRI with contrast. Suspected cavernous sinus lesions (e.g., CN III, IV, or VI) are better imaged with MRI and MRA with and without contrast. This includes combination deficits involving any of the cranial nerves 3–6, a third or sixth nerve palsy combined with an ipsilateral Horner syndrome (e.g., Parkinson sign), or a carotid cavernous fistula (CCF). On MRI, a CCF classically shows enlargement of the superior ophthalmic vein, but may also show EOM enlargement, cavernous sinus enlargement, or proptosis [6]. In addition to MRI/MRA, a CCF usually merits standard catheter angiography to further define the lesion. High resolution 3D MRI could also be used for the evaluation of the cranial nerves anatomy and pathologic conditions [7].

Orbital diseases

Orbital cellulitis

Orbital cellulitis classically presents with blurry vision, chemosis, proptosis, and painful ophthalmoplegia on the affected side. It should be radiographically evaluated urgently to rule out the presence of an abscess and assess the extent of the infection. This is best done with CT scan in the acute setting, because it is faster than MRI and can rule out several other conditions from the differential diagnoses (e.g., sinus disease, thyroid eye disease, subperiosteal abscess, and retrobulbar haemorrhage) [8]. Radiographic findings may include EOM enlargement, orbital fat enhancement, and proptosis. If the diagnosis is still uncertain after CT evaluation, orbital MRI with contrast and fat suppression may be beneficial.

Thyroid eye disease (TED)

TED is an autoimmune condition characterized by EOM enlargement and orbital fat expansion. Patients classically present with proptosis, lid lag, lid retraction, and diplopia. The typical order of involvement of the EOM is inferior rectus, medial rectus, superior rectus then lateral rectus, usually with sparing of the tendons. Radiologic evaluation is important to rule out compressive optic neuropathy that

may lead to permanent vision loss [9]. Non-contrast orbital CT scan is the imaging modality of choice when TED is suspected, because the contrast of CT contains iodine, which may induce thyrotoxicosis (Jod Basedow effect) in hyperthyroidism. In addition, the iodinated contrast is not necessary to appreciate EOM or orbital fat enlargement in TED. CT is also faster and cheaper than MRI, and it allows for better evaluation of bony structures, which is of special importance if surgical intervention has occurred or is under consideration. Though not usually the first option, orbital non-contrast MRI may also be used to evaluate disease progression in patients with TED. If T2-weighted MRI is ordered, muscle inflammation will show as hyperintensity. If T1-weighted MRI is ordered, chronic fatty changes will appear hyperintense [10]. It is important to order fat saturation on these scans, as it facilitates visualization of inflammation and muscle swelling. Other causes of enlarged EOM should be taken in consideration like orbital pseudotumor, sarcoidosis, metastases, lymphoma, and rarely amyloidosis.

Idiopathic orbital inflammatory syndrome (IOIS)/orbital inflammatory pseudotumor

Patients with IOIS or orbital inflammatory pseudotumor classically present with diplopia, pain, and proptosis. Symptom onset may be acute or subacute. CT or MRI scans of the orbits are both adequate studies to assess this condition and may show enlargement or enhancement (indicating inflammation) of EOMs (Fig. 1), the optic nerve, lacrimal gland, or orbital fat. These findings are similar to those of TED; however, in IOIS, structures besides the muscles and orbital fat are often involved (e.g., tendons, lacrimal gland, and cavernous sinus).

Horner syndrome

Patients with Horner syndrome present with the classic triad of ptosis, miosis, and anhidrosis. These signs are due to interruption of the sympathetic chain, which is a three neuron system. Starting in the hypothalamus, the first order neuron descends posterolaterally through the brainstem to the ciliospinal centre of Budge at the C8-T2 level of the spinal cord where it synapses. The second order neuron then travels over the apex of the lung and synapses again in the superior cervical ganglion of the cervical sympathetic chain. Finally, the third order neuron travels with the internal carotid artery to the cavernous sinus, where it briefly courses with cranial nerve six, then passes to cranial nerve five and goes on to innervate the lid and pupillary dilator through the superior orbital fissure [11]. The presence or absence of anhidrosis or pharmacological localization of Horner is no longer used to rely on before imaging of the patient. MRI/MRA or CT/CTA of the brain and neck with and without contrast are the imaging studies of choice for this condition. CT/CTA is usually obtained in the acute setting, while MRI/MRA is used more often with non-acute presentation. In either case, the studies should be ordered to cover the entire sympathetic chain, as the Horner syndrome may be secondary to different causes (e.g., apical lung Pancoast tumour, spinal cord lesion, internal carotid artery (ICA) dissection, and cavernous sinus lesion). If the Horner syndrome presents acutely and is accompanied by pain, an ICA dissection should be suspected. T1-weighted MRI of the neck with fat suppression may show a diagnostic "crescent" sign of hyperintensity that will occlude some of the normal hypointensity that would normally occupy the ICA lumen.

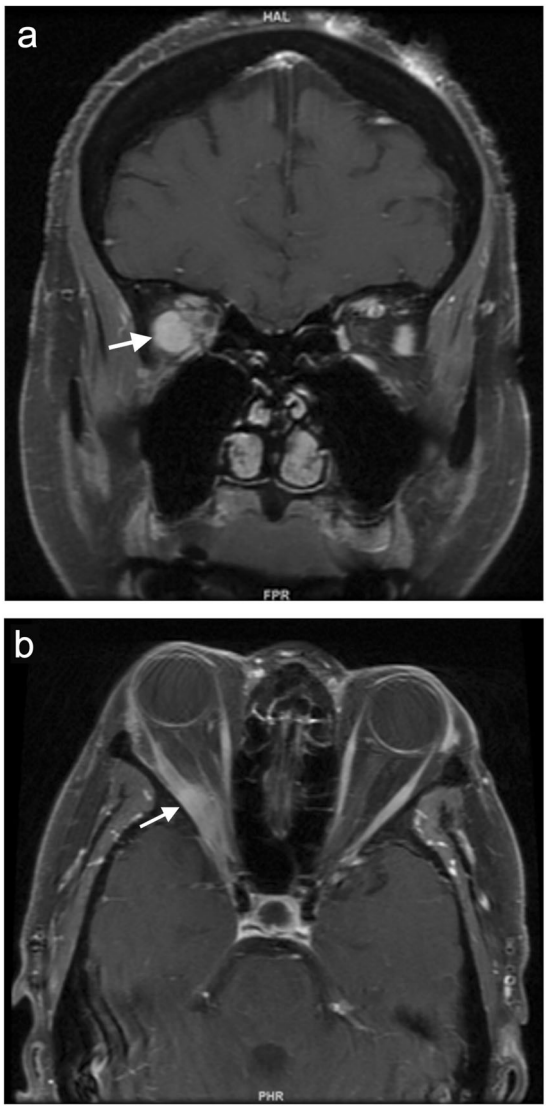

Fig. 1 Patient with idiopathic orbital inflammatory disease. a, b Coronal and axial magnetic resonance images (MRI) of the brain show enhancing lesion involving the posterior aspect of the right lateral rectus muscle with extension into the right orbital apex in patient with idiopathic orbital inflammatory disease.

Nystagmus

Nystagmus is a rhythmic eye movement that can be a sign of a variety of different pathologies. Some specific types of nystagmus seen clinically localize to specific areas of the brain. For example, "see-saw" nystagmus is often caused by midbrain or parasellar lesions, and downbeat and periodic alternating nystagmus can be caused by Chiari malformations or other lesions at the cervicomedullary junction [12–14]. If nystagmus is present without a known cause, MRI with and without contrast of both the brain and the brainstem is indicated.

Optic neuropathies

Patients with suspected optic neuropathies require neuroimaging to assess disease progression, because as the optic nerve becomes more and more affected, vision may be lost progressively. Signs of optic neuropathy can include decreased visual acuity, a relative afferent pupillary defect, visual field defects, or dyschromatopsia. Imaging may identify causes that include demyelination, inflammation, infiltration (e.g., sarcoidosis), mass lesions, or increased intracranial pressure (ICP). MRI of the head and orbit with and without gadolinium is the imaging modality of choice to evaluate an optic neuropathy. The MRI should include T1-weighted scans with and without contrast of the head and orbit (preferably with dedicated orbital protocol) along with T2-weighted scans with FLAIR [15, 16]. CT scan can also be used if haemorrhage or bony abnormalities are suspected. It may also be employed if MRI is contraindicated for any reason.

Multiple sclerosis associated and sporadic optic neuritis

When young adult patients present with painful, unilateral vision loss, optic neuritis (ON) should be considered. Though imaging is not required to make the diagnosis of ON, orbital and cranial MRI with and without gadolinium contrast are often used to look for signs of multiple sclerosis (demyelinating white matter lesions) or optic nerve enhancement. When ordering MRI to assess for multiple sclerosis associated ON, T2-weighted imaging with FLAIR should be ordered (suppressing CSF hyperintensity and highlighting the demyelinating white matter lesions) [17, 18]. T1-weighted post contrast images with fat suppression were reported to identify abnormal enhancement of the optic nerve in about 95% of cases of ON [19]. Due to this, if abnormal enhancement is not present on an initial T1-weighted study with fat suppression, the diagnosis of ON may be doubted.

Neuromyelitis optica associated optic neuritis

Patients with neuromyelitis optica spectrum disorders (NMOSD) often suffer from immune-mediated (aquaporin-4 immunoglobulin G antibodies) optic neuritis and transverse myelitis [17]. This disorder is aggressive and may lead to visual impairment more quickly than MS. It is also more often bilateral and will more frequently reveal papillitis on fundus examination than MS-associated ON and warrants neuroimaging to assess disease progression. Imaging may show bilateral, long segments of optic nerve or chiasmal enhancement or extension into the optic tract [20]. This is in distinction to MS-associated ON, in which the disease is more often unilateral and the enhancement typically appears in shorter segments. Involvement of the aquaporin-4 rich areas like area postrema and hypothalamus have also been associated with NMOSD, so these findings should prompt clinical exploration of NMOSD as a possible diagnosis. NMOSD should be considered any time a patient presents with ON without demyelinating white matter lesions (typical of MS).

Anti-myelin oligodendrocyte glycoprotein (MOG) IgG antibody associated optic neuritis

MOG is another immune-mediated CNS disease, similar to NMOSD. MOG differs from NMOSD in the target of the implicated antibody (myelin oligodendrocyte glycoprotein instead of aquaporin-4). Patients with this condition may initially present with recurrent ON or acute disseminated encephalomyelitis with negative testing for aquaporin-4 antibodies. MRI can help to distinguish ON secondary to MS, NMOSD, or MOG, but oftentimes does not yield a conclusive diagnosis due to significant overlap in imaging findings. Like NMOSD, optic nerve enhancement tends to be present in longer segments in MOG (Fig. 2), as opposed to shorter segments in MS-associated ON [21, 22]. Chiasmal involvement and spinal cord lesions are more common in NMOSD [23–25], and it has been reported that lesion resolution with and without treatment is more common in MOG [26, 27]. In addition, in MOG, post contrast, fat suppressed T1-weighted MRI may show optic nerve or nerve sheath enhancement that can mimic optic perineuritis [28].

Other autoimmune optic neuropathies

Systemic lupus erythematosus, sarcoidosis, and many other autoimmune diseases can affect the optic nerve. When evaluated with T2-weighted MRI, they typically show abnormal hyperintensity [15, 29]. Involved structures may include but are not limited to the cavernous sinus, leptomeninges, or pituitary gland.

Ischemic optic neuropathies

Ischemic optic neuropathies may be divided into arteritic anterior ischemic optic neuropathy (AAION), non-arteritic

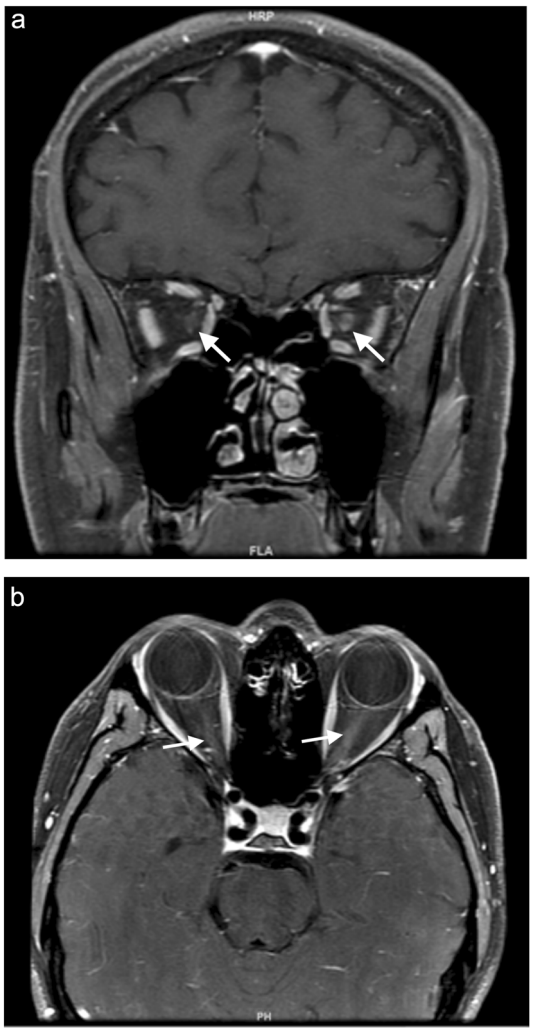

Fig. 2 Patient with anti-myelin oligodendrocyte glycoprotein (MOG) IgG antibody associated optic neuritis. a, **b** Coronal and axial magnetic resonance imaging (MRI) of the brain show diffuse abnormal enhancement of the left optic nerve and sheath in patient with anti-myelin oligodendrocyte glycoprotein (MOG) IgG antibody associated optic neuritis.

ischemic optic neuropathy (NAION), and posterior ischemic optic neuropathy (PION). All three have slightly different findings on diagnostic imaging although it is usually the history and physical exam that help distinguish between them, not the imaging. In AAION, patients present with jaw claudication, headache, scalp tenderness, and possible progression to vision loss, making this a visual emergency. Enhancement of the optic nerve and temporal arteries are often seen in the context of temporal cell arteritis [30]. In NAION, imaging studies are generally not indicated, because the diagnosis is made clinically. However, if the patient experiences unusual or worrisome symptoms (e.g., vision loss, significant pain), imaging may be used to look for compressive, demyelinating, or compressive lesions affecting the optic nerve [31]. On T2-weighted MRI, findings may include non-specific hyperintensity of the optic nerve. This is a rare finding, so when enhancement of the optic nerve or optic sheath is found radiographically in the context of NAION, other diagnoses should be ruled out. Posterior ischemic optic neuropathy usually occurs after cardiothoracic or spinal surgeries. Findings on T2-weighted MRI include hyperintensity and diffusion restriction on DWI within the optic nerve [32].

Traumatic optic neuropathy

In the context of an acute trauma, imaging is often not necessary to make the diagnosis, but CT scan with thin slices is often employed to check for fractures of the optic canal or orbit. The CT scan can also assess for intracranial haemorrhage secondary to head trauma [33].

Sometimes a haematoma of the optic nerve sheath may not be seen on CT scan, so if the clinical suspicion is high, an MRI may also be ordered as an adjunct study to assess for this possibility. If a nerve sheath haematoma is seen, it may be treatable surgically. Hyperintensity in the optic nerve would indicate traumatic optic neuropathy.

Intracranial and orbital tumours

Pituitary adenoma

Adenomas of the pituitary gland may present with hyperprolactinemia, visual field defects, hypercortisolism, other endocrine abnormalities, or it may be an incidental finding on imaging. Microadenomas can be distinguished from macroadenomas by the mass effect that the latter can exert on the optic pathway or invasion of the cavernous sinus. Usually evaluated with MRI, pituitary adenomas typically appear hypointense on T1-weighted images with gadolinium contrast enhancement (Fig. 3). On T2-weighted imaging, the macroadenoma may be "snowman-shaped." [34] It can be indistinguishable from normal pituitary gland on late contrast MRI images and small adenomas could be hard to see without dedicated high-resolution imaging through the sella.

One possible complication of pituitary lesions is pituitary apoplexy (acute pituitary haemorrhage). It warrants special mention, because it is often not visible on a standard CT scan. Due to this, when evaluating for possible pituitary apoplexy, MRI or CT with specially ordered thin slices should be employed [35, 36]. The imaging of choice is usually non-contrast CT to visualize acute bleeding, as patients often present with subarachnoid haemorrhage.

Craniopharyngioma

Craniopharyngiomas are typically evaluated with MRI because they delineate the classically cystic and solid

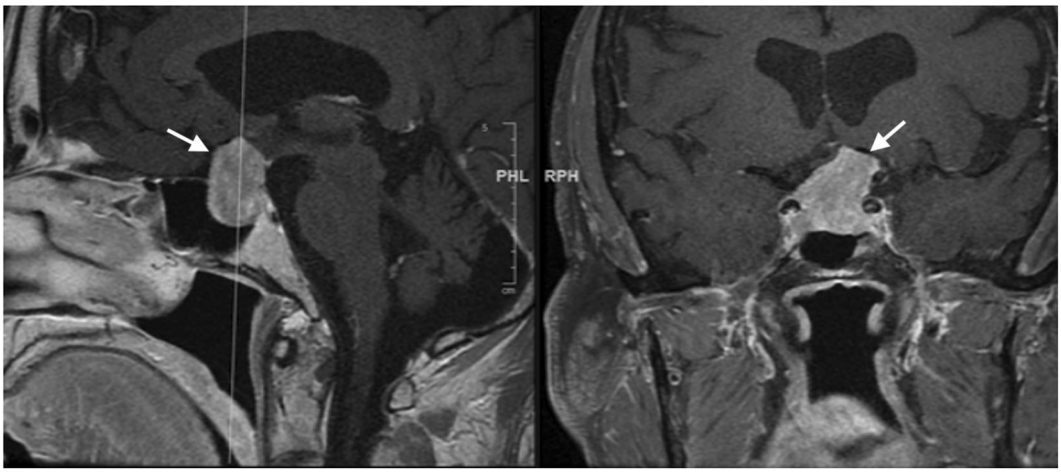

Fig. 3 Coronal and sagittal magnetic resonance images (MRI) of the brain show sellar mass with suprasellar extension and mass effect on the overlying chiasm and optic pathways compatible with pituitary macroadenoma.

components of the tumour well [34]. On T2-weighted imaging, the cystic components of the lesion appear hyperintense. On T1-wighted imaging, the cystic components may appear either hyper- or hypo-intense, and the solid components may enhance with contrast (Fig. 4). If there is uncertainty as to the type of mass in question, a CT scan may be useful as an adjunct study to identify calcifications in a craniopharyngioma that may not be present in other cystic sellar/suprasellar lesions.

Meningiomas

Meningiomas may cause an optic neuropathy via compression of the optic nerve, optic chiasm, or optic tract. They can originate from the tuberculum sella, diaphragm sellae, or the anterior clinoid process [37]. They are visible on both CT and MRI. Upon CT evaluation, meningiomas usually appear isodense or slightly hypodense with homogenous contrast enhancement (Fig. 5). They may also show calcifications or hyperostosis of bone. On MRI, they tend to appear isodense with T1-weighted imaging and may show either isodense or hyperdense on T2-weighted imaging, homogenously enhancing with contrast administration [38]. A dural tail of contrast enhancement may also be seen. Meningiomas may also originate from the optic nerve sheath (Fig. 6). Patients with this condition usually present with painless visual loss of insidious onset associated with optic disc atrophy or oedema, and may be associated with the formation of retinochoroidal venous collaterals (i.e., optociliary shunt vessels) [15]. They are usually unilateral, but may rarely be bilateral (e.g., neurofibromatosis type II). On CT scan, calcifications, bony erosions, or hyperostosis of the optic canal may be present [39]. On MRI, the optic sheath may enhance with contrast administration, revealing the characteristic "tram track" appearance. When ordering imaging to investigate a patient presenting with insidious unilateral visual loss, it is important to recognize that intracanalicular optic nerve sheath meningiomas are easily missed on initial imaging. If the index of suspicion is high,

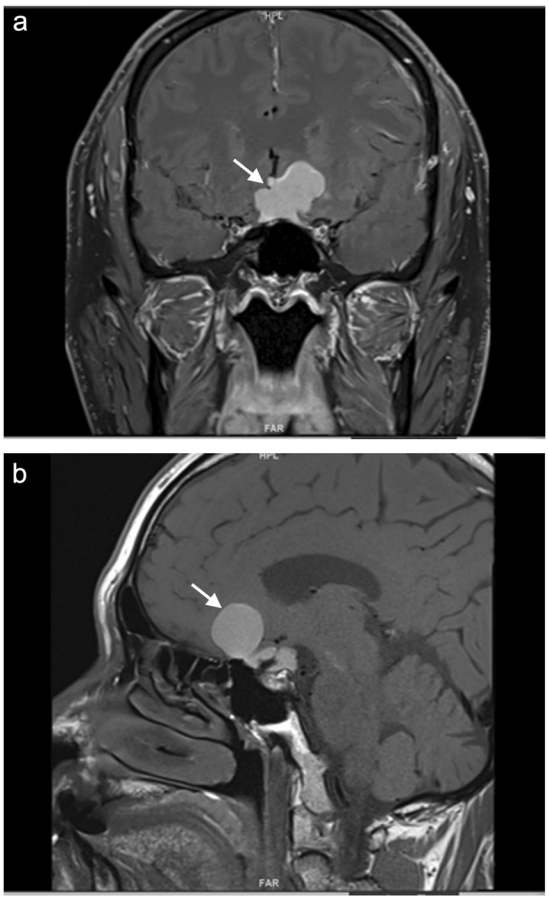

Fig. 4 Patient with craniopharyngioma. a, **b** Coronal and sagittal magnetic resonance images (MRI) of the brain show a mass in the suprasellar region and adjacent anterior cranial fossa with mass effect on the optic nerves and chiasm consistent with craniopharyngioma (pathology proven).

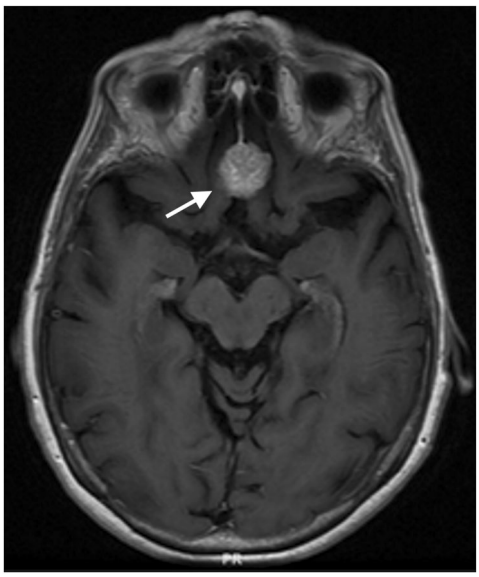

Fig. 5 Axial T1 weighted magnetic resonance images (MRI) of the brain shows the midline subfrontal enhancing extra-axial mass consistent with planum sphenoidale meningioma.

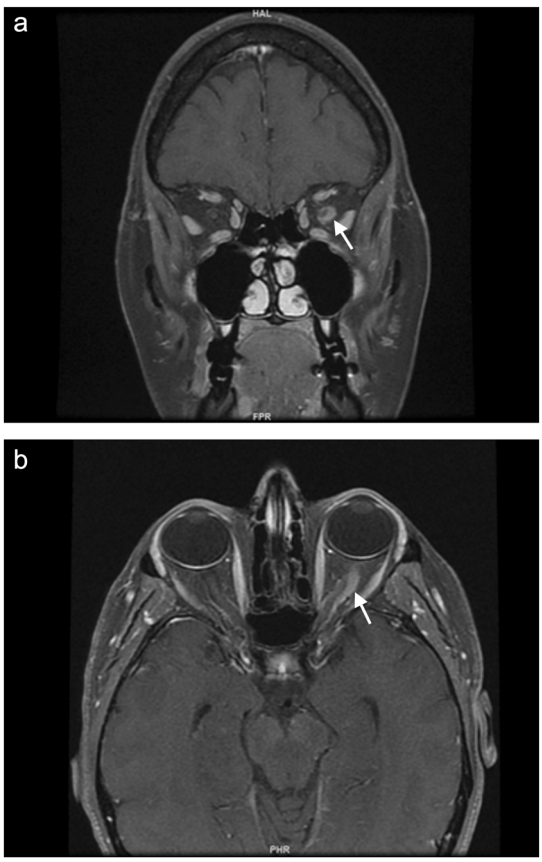

Fig. 6 Mass consistent with optic nerve sheath meningioma. a, **b** Coronal and axial magnetic resonance images (MRI) of the brain show left enhancing intraorbital extra axial mass around the left optic nerve consistent with optic nerve sheath meningioma.

an MRI could be ordered with thin slices and contrast directed at the optic canal. Another possible pathologic process that can be caused by a meningioma is Foster Kennedy syndrome (ipsilateral compressive optic neuropathy causing optic atrophy with contralateral papilledema resulting from increased ICP). This may occur when a meningioma grows slowly and gets large enough to increase ICP and cause papilledema. MRI of the brain and orbit with contrast is recommended for the Foster Kennedy syndrome.

Gliomas

Optic pathway gliomas (OPG) are associated with neurofibromatosis type I and primarily occur in children. OPG are primary tumours of the optic nerve [40]. MRI usually shows fusiform enlargement and enhancement of the optic nerve parenchyma (in contrast to the optic sheath enhancement seen in meningiomas). Extension into the optic chiasm, optic tracts, and optic radiations may also occur [40]. Though more common in children, OPG may also occur in the adult population where they tend to be more aggressive [15] and do not respond as well to treatment. The clinical approach for OPG in adults and children differs due to this difference in aggression. In children, OPG may be observed for progression on serial imaging or for symptomatic progression. Rapid change or growth would indicate the possibility of malignancy or the presence of a glioblastoma (more common in adults). In adults, serial imaging is always indicated, and they could be considered for surgical biopsy.

Papilledema

Papilledema is caused by increased ICP and results in swelling of the optic disc. Patients may present with headache, tinnitus, diplopia, or blurry vision. This can occur secondary to a variety of different pathologies, so imaging is important to narrow the list of possibilities. CT and MRI of the brain and orbits with and without contrast are recommended to rule out easily visible aetiologies (e.g., intracranial masses, haemorrhage). In addition to conventional scans, the venous system of the brain should be evaluated with MRV or CTV to rule out venous sinus thrombosis or stenosis (Fig. 7). If a cause for the high ICP is not identified, IIH could be considered as a diagnosis. Findings on imaging suggestive of IIH include posterior flattening of the globes, empty or partially empty sella (Fig. 8), prominent fluid within the optic nerve sheath (Fig. 9), and narrowing of the transverse sinuses [41], narrowing of the junction between the transverse and sigmoid sinuses, pinched lateral ventricle frontal horns and vertical tortuosity of the optic nerves.

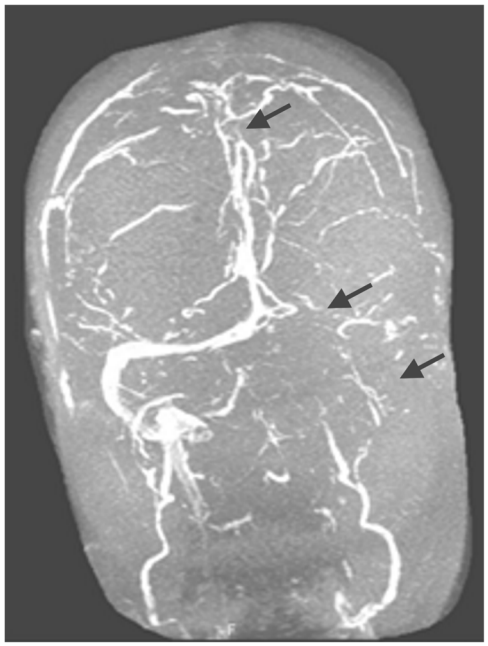

Fig. 7 Magnetic resonance venogram (MRV) shows occlusion/severe stenosis with poor venous flow in the superior sagittal sinus, left transverse sinus and left sigmoid sinus in patient with increased intracranial pressure.

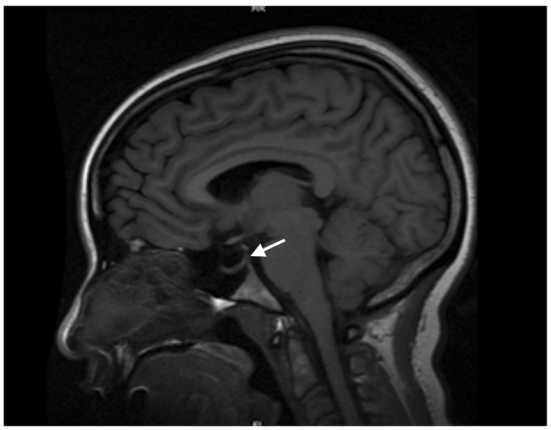

Fig. 8 Sagittal magnetic resonance imaging (MRI) of the brain shows the partially empty sella as a radiographic sign of increased intracranial pressure. Other differential diagnosis for the empty sella include pituitary cystic lesions like arachnoid cyst, Rathke cleft cyst, craniopharyngioma, epidermoid, and cystic pituitary adenoma, although in most cases this finding is essentially an incidental finding.

Retrochiasmal disorders

Retrochiasmal lesions may affect the optic tract, lateral geniculate nucleus, optic radiations, or the occipital cortex. Depending on the lesion's location, it will produce a variation of homonymous hemianopsia. Because of this, when a patient presents with homonymous hemianopsia, imaging

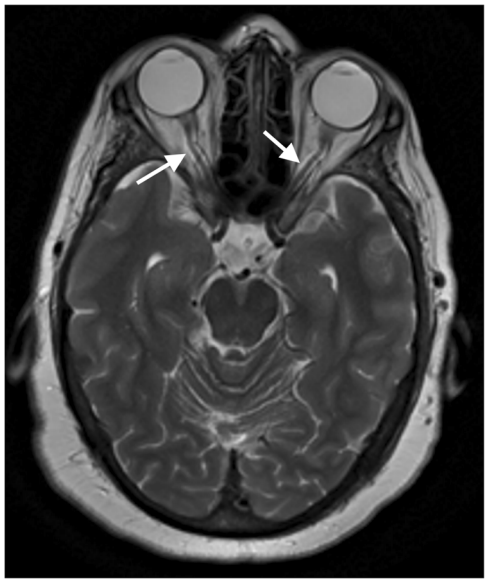

Fig. 9 Axial magnetic resonance imaging (MRI) of the brain shows fluid in the optic nerve sheath of both eyes and flattening of the optic disc particularly on the right in patient with increased intracranial pressure.

is indicated. Which study to order depends on the acuity of presentation. If the onset is acute, non-contrast CT scan is indicated to rule out intracranial haemorrhage. If the CT is negative, enhanced cranial MRI with DWI is recommended to search for lesions suggestive of acute ischaemia. DWI and ADC techniques will make this evident by showing restricted diffusion [1]. These techniques are also important, because they can differentiate between vasogenic oedema (typical of posterior reversible encephalopathy syndrome) and cytotoxic oedema (typical of acute stroke). Both vasogenic and cytotoxic oedema will show hyperintensity on T2-weighted imaging with FLAIR and DWI, but only cytotoxic oedema will show dark signal on ADC compatible with restricted diffusion. Cytotoxic oedema is less reversible than vasogenic oedema and, therefore, impacts prognosis [1, 42, 43]. Clinicians should also keep in mind that both types of oedema may be present, which could result in mixed findings on imaging.

Positron emission tomography (PET)

PET scan is a non-invasive nuclear medicine study used in patients with neuro-ophthalmological diseases to quantify the brain metabolism and alterations in regional blood flow [44] using three-dimensional images of the functional processes of the brain, a positron (a subatomic positive particle) emitting tracer is injected into the blood stream on a molecule emits gamma rays that are detected by the system to create the three-dimensional images of the parts of the body that the tracer accumulate in using a computer analysis

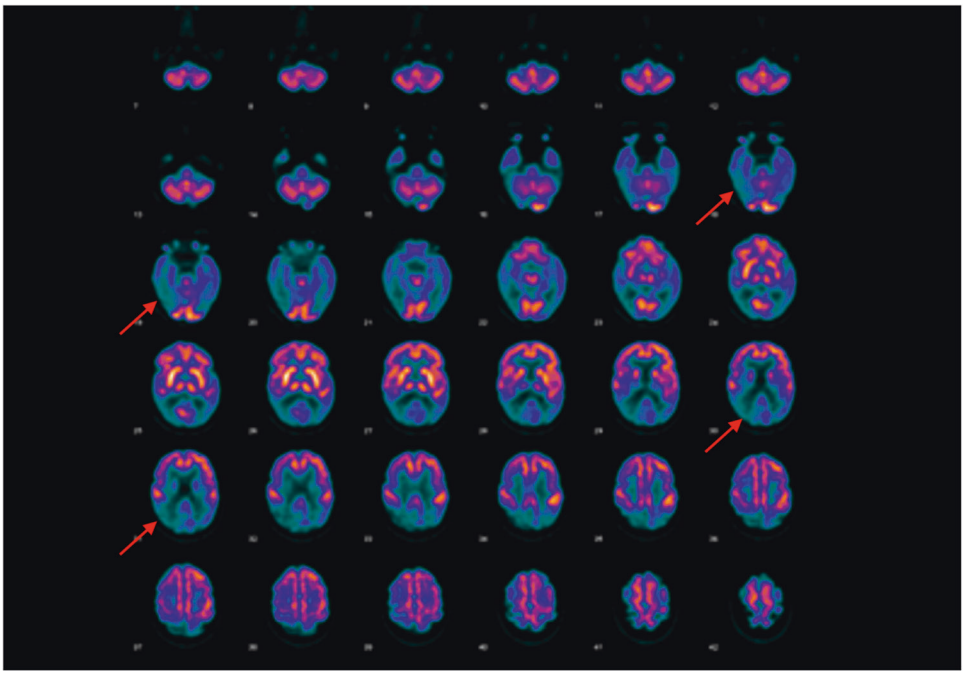

Fig. 10 Positron imaging tomography (FDG-PET) scan showing asymmetric hypometabolism in the occipital and parietal lobes indicative of a diagnosis of posterior cortical atrophy (PCA).

system. {18F}-fluoro-2-deoxyglucose (18F-FDG) is a radiotracer used in PET scans which is an analogue of glucose that helps to delineate the metabolic activities of the parts of the body that is accumulating in [45]. It has many implications, one of the most common uses is in patients with cancer to detect the possibility of metastases. In neurology, it can be used to evaluate for memory loss and can help to differentiate the various types of dementia in some patients. Ophthalmologists can be the initial physicians to see patients with posterior cortical atrophy (PCA) due to visual complaints early in the disease process, those patients can present with visuospatial and visuo-perceptual deficits; dysgraphia, optic ataxia, oculomotor apraxia, and simultanagnosia, they may have homonymous visual field defects but the eye examination usually is normal, other differential diagnosis of PCA syndrome are Alzheimer's disease, dementia with Lewy bodies and Creutzfeldt–Jakob disease. Most cases are due to Alzheimer's disease but unlike typical Alzheimer's disease, patients with PCA usually present at younger age (mid 50s) and develop memory problems later. It needs a high index of suspicion to diagnose it and PET scan can help establishing the diagnosis. PCA has been associated with hypometabolism in the occipital lobe evident on 18F-FDG PET scan (Fig. 10) and with atrophy of the primary visual cortex and thalamus that may explain the visual hallucinations in PCA [46]. Other differential diagnosis of visual hallucinations is Charles Bonnet syndrome which is characterized by complex visual hallucinations in patients with decreased vision and without mental disorders, it can cause disturbances that could be visualized by FDG-PET exam as a hypermetabolism in the visual cortex [47].

In summary, ophthalmologists should be aware of the major structural imaging studies (CT and MRI) and in most cases MRI with contrast is superior to CT for brain lesions. In cases where a more rapid or emergent scan is necessary (e.g., stroke, trauma) or in patients in whom a hyperdensity is suspected (e.g., calcification, haemorrhage, hyperostosis, and fracture), or in patients who cannot undergo an MRI, CT may be a reasonable adjunctive or alternative imaging study.

Special sequences including CTA/CTV and MRA/MRV may be necessary for vascular lesions in addition to traditional CT and MR scans. Special sequences in MRI include FLAIR and fat suppression to attenuate normal high signal intensity and to better allow differentiation of pathologic hyperintensity in specific ophthalmic disorders. The topographic anatomy of the structures being imaged (e.g., Horner syndrome, optic neuropathy, and ocular motor cranial neuropathy) is critical in determining the type and extent of the neuroimaging. Clinicians should be aware of the basic indications and contraindications for CT, MRI, and PET in ophthalmology.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

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