#### **REVIEW ARTICLE**



# Extraocular muscle enlargement

Khizar Rana<sup>1,2</sup> • Valerie Juniat<sup>1,2</sup> • Sandy Patel<sup>3</sup> • Dinesh Selva<sup>1,2</sup>

Received: 1 March 2022 / Revised: 28 March 2022 / Accepted: 2 April 2022 / Published online: 17 June 2022 © The Author(s) 2022

#### **Abstract**

Extraocular muscle enlargement can occur secondary to a range of orbital and systemic diseases. Although the most common cause of extraocular muscle enlargement is thyroid eye disease, a range of other inflammatory, infective, neoplastic, and vascular conditions can alter the size and shape of the extraocular muscles. Imaging with computed tomography and magnetic resonance imaging plays an essential role in the workup of these conditions. This article provides an image-rich review of the wide range of pathology that can cause enlargement of the extraocular muscles.

Keywords Extraocular muscle · Computed tomography · Magnetic resonance imaging · Orbit

# **Key messages:**

- Extraocular muscle enlargement has a broad differential diagnosis.
- The shape, enhancement and pattern of extraocular muscle enlargement, and involvement of surrounding orbital fat, nerves and veins can help to narrow the differential.
- In some cases, a diagnosis can be confidently made through recognition of characteristic imaging features.

# Introduction

Extraocular muscle enlargement (EOME) can occur secondary to an array of inflammatory, neoplastic, infective or vascular conditions. Orbital imaging with computed tomography (CT) or magnetic resonance imaging (MRI) plays an essential role in the workup of these conditions. High-resolution orbital MRI has allowed for a more detailed characterisation of orbital disease. Recognising the pattern of muscle involvement along with accompanying orbital signs can help to narrow the differential diagnosis, or even enable

a single diagnosis. This review focuses on the radiological findings associated with pathology that cause extraocular muscle enlargement.

#### Thyroid eye disease

Thyroid eye disease (TED) is the most common inflammatory myopathy affecting the extraocular muscles. Radiologically, it presents with bilateral enlargement of the muscle belly with relative sparing of the anterior tendon. Traditionally, muscles are said to be involved in the following order: inferior rectus, medial rectus, superior rectus and lateral rectus (Fig. 1A) [1]. However, it is now being recognised that superior muscle complex involvement is also very frequent, possibly the most common [2, 3]. Furthermore, isolated levator muscle enlargement can also occur (Fig. 1B) and is associated with upper eyelid retraction [2–4]. Both superior and inferior oblique involvement has also been recognised and should be assessed on imaging (Fig. 1C, D) [5, 6].

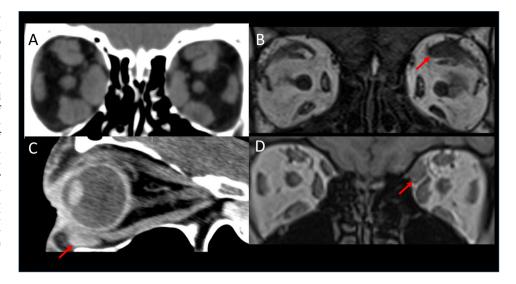
Most patients with TED have muscle enlargement. However, up to one-fourth of patients will have no fat or muscle

- Department of Ophthalmology & Visual Sciences, University of Adelaide, North Terrace, Adelaide, SA 5000, Australia
- South Australian Institute of Ophthalmology, Royal Adelaide Hospital, Port Road, Adelaide, SA 5000, Australia
- Department of Medical Imaging, Royal Adelaide Hospital, Port Road, Adelaide, SA 5000, Australia



 <sup>⊠</sup> Khizar Rana khizar.rana@adelaide.edu.au

Fig. 1 Thyroid eye disease. Coronal CT (A) of the orbit demonstrating a 'typical' case of TED with bilateral extraocular muscle enlargement of the inferior rectus, medial rectus and superior muscle group. Coronal T1-weighted MRI (B) showing enlargement of the left levator palpebrae superioris (arrow) without involvement of the other muscles. Quasi-sagittal CT (C) of the right orbit showing enlargement of the inferior oblique muscle (arrow) with relative sparing of the inferior rectus. T1-weighted coronal MRI (D) showing predominant enlargement of the left superior oblique muscle (arrow)



enlargement, and a smaller proportion will only have fat expansion without muscle enlargement [7]. TED can also be accompanied by bilateral, unilateral or asymmetric lacrimal gland enlargement [8, 9].

TED is a clinical diagnosis. Imaging can assist in differentiating TED from alternative pathologies in unclear cases, identify active orbital inflammation, predict response to therapy and assist in diagnosing dysthyroid optic neuropathy.

# Dysthyroid optic neuropathy

Dysthyroid optic neuropathy (DON) is a sight-threatening complication of TED. A number of radiological features can assist in diagnosing DON: orbital apex crowding [10], higher extraocular muscle volumes [11], superior ophthalmic vein enlargement [12], effacement of the fat plane surrounding the optic nerve [13], anterior displacement of the lacrimal gland [13], enlarged tendons [14] and superior orbital fissure fat prolapse [15]. It should be noted that DON is essentially a clinical diagnosis of exclusion and absence of the aforementioned radiological signs does not preclude the diagnosis.

A muscle index of greater than 50%, defined as the percentage of orbital height occupied by the superior and inferior recti or percentage of width occupied by the horizontal recti at the midpoint between the posterior globe and orbital apex, has a high sensitivity for DON (Fig. 2) [15, 16]. The anatomy of the bony orbit can also predispose patients to the development of DON. Patients with DON have shown to have narrower bony orbital angles than patients without DON for an identical muscle diameter index [10].

Enlargement of the superior muscle complex plays an important role in the development of DON [17, 18]. This correlates clinically with the inferior visual field deficits seen in TED. The ratio of the superior rectus-levator complex volume to the summated soft tissue volume is

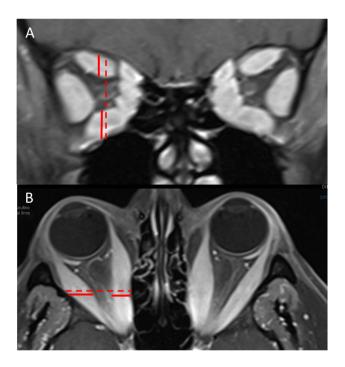


Fig. 2 Right dysthyroid optic neuropathy. Mid-coronal fat-suppressed contrast-enhanced T1-weighted MRI (A) showing the vertical muscle index, which is a ratio of the sum of the vertical height of the superior and inferior recti (solid line) to the vertical orbital height (dashed line). Axial fat-suppressed contrast-enhanced T1-weighted MRI (B) showing the horizontal muscle index, which is a ratio of the sum of the diameters of the medial and lateral recti (solid line) to the horizontal orbital distance (dashed line)

significantly higher in patients with DON, whereas no significant differences have been observed for the other extraocular muscles [18]. Anatomically, this may be explained by the fact that the superior rectus narrows vertically less than the inferior rectus as it approaches the annulus of Zinn.

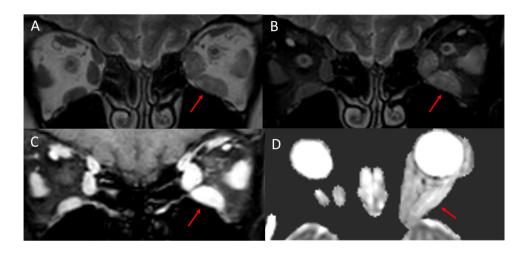


# Active thyroid eye disease

Various imaging techniques have been used to assess orbital inflammation to help diagnose active disease and predict response to therapy. Active TED involves inflammatory oedema of the extraocular muscles which corresponds to T2-hyperintensity and increased contrast enhancement relative to unaffected extraocular muscles (Fig. 3) [19, 20]. This is in comparison to the findings seen in a normal patient (Fig. 4) with no T2-hyperintensity of the extraocular muscles, no significant or asymmetric extraocular muscle enhancement and no hyperintense signals on ADC map.

#### Signal intensity ratio

A higher signal intensity ratio (SIR), the ratio of signal intensity from the extraocular muscles to the signal intensity of normal tissue (e.g. brain white matter), is a marker of active TED and has shown correlation with the clinical activity score [21–23]. The SIR can be measured on STIR and T2-weighted sequences (Fig. 5) [19, 21–24]. The SIR usually decreases following immunosuppressive therapy or radiotherapy [25, 26]. Patients with persistently high SIRs despite medical therapy may be at an increased risk of further clinical deterioration [25].



**Fig. 3** Active thyroid eye disease. Coronal T2-weighted MRI (**A**) showing enlargement and hyperintensity within the left inferior rectus, medial rectus and lateral rectus muscles (arrow). Coronal fat-suppressed T2-weighted MRI (**B**) demonstrating enlargement and hyperintensity of the left inferior rectus, medial rectus and lateral

rectus (arrow). Coronal fat-suppressed T1-weighted MRI ( $\mathbf{C}$ ) showing relatively increased enhancement of the affected left extraocular muscles (arrow). ADC map ( $\mathbf{D}$ ) demonstrating a hyperintense signal within the left orbit (arrow)

Fig. 4 Normal extraocular muscles. Coronal T2-weighted MRI (A) demonstrating no muscle enlargement and no hyperintensity within the muscles. Coronal fat-suppressed T2-weighted MRI (B) showing symmetrical signal intensity of the muscles. Fat-suppressed contrast-enhanced T1-weighted MRI (C) showing symmetrical enhancement of the extraocular muscles. ADC map (D) showing no hyperintense signals within either orbit

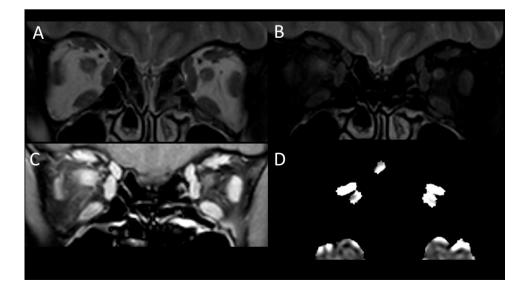
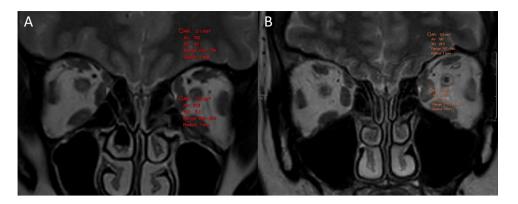




Fig. 5 Signal Intensity ratio.
Normal coronal T2-weighted
MRI (A) showing the signal
intensity ratio of the extraocular
muscle and brain white mater.
Coronal T2-weighted MRI (B)
in a patient with active TED
involving the left inferior
and medial rectus muscles
showing the signal intensity
ratio between the inflamed left
medial rectus and brain white
mater



#### Apparent diffusion coefficient

Diffusion-weighted imaging and the corresponding apparent diffusion coefficient (ADC) values can quantitatively and qualitatively assess the diffusion of water at a cellular level. Higher ADC values are seen in the acute inflammatory phase of TED and can help differentiate the active phase of TED from the inactive fibrotic phase (Fig. 3D) [27]. Hyperintense ADC signal within the extraocular muscles may be seen prior to any abnormality being seen on conventional MRI [28]. This may allow for earlier initiation of medical therapies in these patients. Additionally, ADC values can also help to monitor treatment response as they have shown correlation with the CAS and decrease following steroid therapy [27].

#### T2 relaxation time

T2 relaxation time (T2RT) is a tissue-specific time constant describing the decay of transverse magnetisation of tissues. Accordingly, changes within the cellular architecture of extraocular muscles can be quantitatively assessed with T2RTs. Patients with active TED have higher T2RT than inactive TED or healthy controls, and the T2RT has been correlated with the CAS and muscle areas [29–31]. A higher pre-intervention T2RT may be used to predict patients who are less likely to response to steroid therapy [32]. T2RT decreases following steroid therapy and can also be used to monitor the patient's response to steroid therapy [30, 33].

# Inactive thyroid eye disease

Inactive TED is characterised by interstitial fibrosis, collagen deposition and fatty infiltration. Fatty infiltration can be seen as areas of hypodensity within the extraocular muscles on computed tomography and is significantly more common in patients with TED as compared to healthy controls [34]. Heterogenous areas of low signal intensity within the

extraocular muscles on T2-weighted imaging may represent areas of fibrotic change and is more common in patients with irreversible diplopia following medical therapy [24].

#### Other inflammatory conditions

Myositis of the extraocular muscles can be secondary to idiopathic orbital myositis or a range of specific autoimmune conditions. McNab [35] has proposed a comprehensive classification system for orbital myositis focused on defining the underlying aetiology of the myositis.

# **Idiopathic orbital myositis**

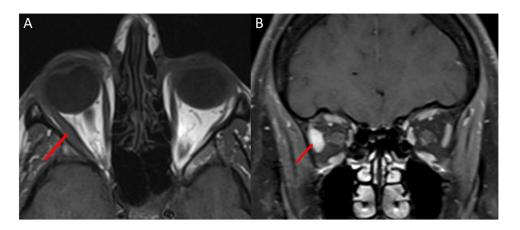
Idiopathic orbital myositis is a subset of idiopathic orbital inflammatory syndrome or orbital pseudotumor. Idiopathic orbital myositis commonly presents unilaterally with single muscle involvement (Fig. 6) [36–38]. The medial or lateral recti are most commonly involved [39, 40]. Enlargement of the anterior muscle tendon can help differentiate it from TED; however, this is only seen in approximately half of all cases and in approximately 6% of TED patients [37, 38, 41, 42]. Orbital fat infiltration and lacrimal gland involvement can also be observed [43–45]. Multiple and bilateral muscle involvement and muscle tendon sparing are risk factors for recurrent disease [38, 46]. On MRI, the affected muscles are isointense on T1 and hyperintense on T2-weighted imaging [47–49].

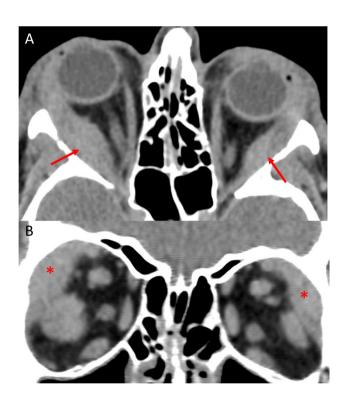
# Specific myositis

**IgG4-related disease** IgG4-related disease (IgG4-RD) is a multisystemic inflammatory disease of unknown aetiology. IgG4-related ophthalmic disease (IgG4-ROD) is being increasingly recognised as a cause of previously labelled idiopathic orbital inflammation [50]. Bilateral enlargement of multiple muscles, particular the lateral rectus, is common (Fig. 7). This is usually accompanied by a combination of lacrimal gland enlargement, infraorbital or frontal nerve enlargement, sinus mucosal thickening, and pterygopalatine



Fig. 6 Idiopathic orbital myositis. Axial T1-weighted MRI (A) showing isointense enlargement of the right lateral rectus muscle including the anterior tendon (arrow). Coronal fatsuppressed contrast-enhanced T1-weighted MRI (B) showing enlargement and enhancement of the right lateral rectus muscle





**Fig. 7** IgG4-related ophthalmic disease. Axial CT ( $\bf A$ ) showing bilateral enlargement of the lateral recti ( $\bf R > L$ ) including the anterior tendon (arrows). Coronal CT ( $\bf B$ ) demonstrating bilateral enlargement of the lacrimal glands (\*)

fossa involvement [51–54]. Trigeminal pathway involvement is a useful diagnostic sign for IgG4-ROD with one study finding that 50% of patients with infraorbital nerve enlargement had IgG4-ROD [55]. On CT, the muscles are isodense compared to unaffected muscles and on MRI muscles are isointense on T1 and hypoisointense lesions on T2 imaging are seen [53, 54, 56].

# Inflammatory bowel disease

Inflammatory bowel disease (IBD) consisting of ulcerative colitis and Crohn's disease has been associated with orbital myositis. Presentation can be unilateral or bilateral with the medial and inferior recti most commonly involved [57–59]. Compared to idiopathic orbital myositis, bilateral muscle involvement is more common in the IBD group [35]. Fusiform enlargement of muscles with tendon sparing can mimic the presentation of TED [57, 59, 60]. The past medical history, thyroid hormone, and antibody studies can help make the distinction in such cases. The muscles can be large enough to cause optic nerve compression [57]. On MRI, the muscles are isointense on T1 and hyperintense on T2-weighted imaging [58, 60, 61].

#### **Sarcoidosis**

The most common ocular presentation of sarcoidosis is with anterior uveitis with orbital myositis being relatively infrequent [62]. When affecting the muscles, bilateral disease with multiple muscle involvement and anterior tendon involvement is commonly seen [36, 63–66]. Involvement of surrounding structures can include the lacrimal gland, lateral wall of the cavernous sinus, and thickening and encasement of optic nerve producing a 'tram-track' sign [64, 67, 68]. The affected muscles are isointense on T1 and hypo-isointense on T2 with potential areas of higher intensity nodules [66–68].

#### **Granulomatosis with polyangiitis**

Granulomatosis with polyangiitis (GPA) is a multisystemic autoimmune disorder characterised by necrotising granulomatous inflammation. Orbital involvement occurs in up to 50% of GPA patients usually presenting with an orbital mass, or dacryoadenitis with isolated extraocular myositis being uncommon [69]. Concomitant sinus and nasal



disease is common. Pachymeningeal enhancement may occur [70]. Extraocular myositis usually presents unilaterally with fusiform muscle enlargement and tendon sparing (Fig. 8) [36, 71–73]. Unilateral or bilateral involvement is possible [74–76]. The affected muscles are isointense on T1 [72]. Areas of hypoenhancement following contrast administration representing zonal necrosis may be suggestive of GPA [73]. On T2, a high signal intensity with a rim of hypointensity may also represent intralesional necrosis suggestive of GPA [71].

A range of other autoimmune conditions can be associated with orbital myositis including systemic/discoid lupus erythematosus [77, 78], giant-cell myositis [79–82], rheumatoid arthritis [83] and psoriasis [84]. Orbital myositis can occur secondary to drug reactions from the newer monoclonal antibodies, bisphosphonates or statins [35]. The pattern of enlargement is similar to idiopathic orbital

Fig. 8 Granulomatosis with polyangiitis (Wegener's). Axial T1-weighted MRI (A) and fatsuppressed contrast-enhanced T1-weighted MRI (B) showing fusiform enlargement of the right medial rectus muscle with anterior tendon sparing. Coronal T1-weighted MRI (C) demonstrates isointense enlargement of the right medial rectus. Coronal fat-suppressed contrast-enhanced T1-weighted MRI (D) shows heterogenous enhancement and enlargement of the right medial rectus

myositis, with the muscles showing T1 isointensity and T2 hyperintensity.

#### **Neoplastic conditions**

A number of tumours can metastasise to the orbit causing extraocular muscle enlargement. Uncommonly, EOME may be the initial manifestation of a previously undiagnosed tumour [85].

Lymphoma Orbital lymphoma is the most common malignant tumour of the orbit in adults [86]. Intramuscular involvement is uncommon [87]. Extraocular muscle lymphoma presents unilaterally with either fusiform muscle enlargement and tendon sparing or diffuse muscle enlargement with tendon involvement (Fig. 9) [88–93]. Lymphoma is hypoisointense on T1 and iso to hyperintense on T2

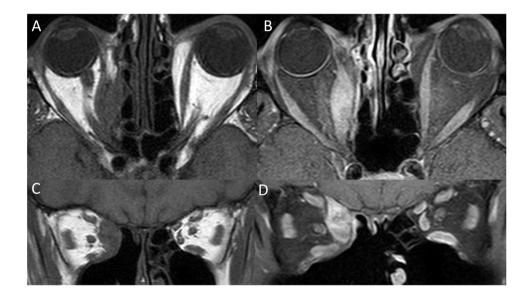
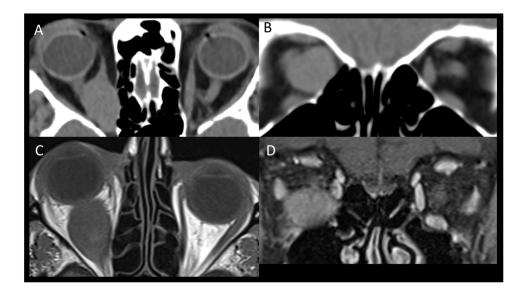


Fig. 9 Intramuscular lymphoma. Case 1—axial (A) and coronal (B) CT scan showing significant enlargement of the right medial rectus muscle without involving the anterior tendon and causing optic nerve displacement. Case 2—axial T1-weighted MRI (C) shows isointense enlargement of the right inferior rectus muscle and right proptosis. Coronal fatsuppressed contrast-enhanced T1-weighted MRI (D) demonstrates heterogenous enlargement of the right inferior rectus muscle





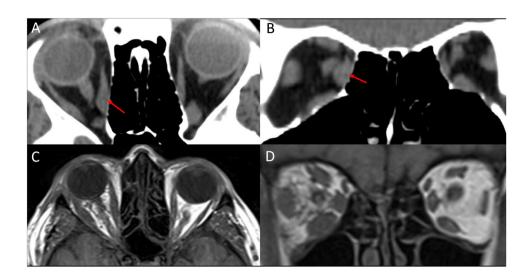
imaging [75, 89, 94–96]. The presence of an orbital mass or lacrimal gland involvement that moulds to the globe or bone can help aid the diagnosis of lymphoma [89, 97–102].

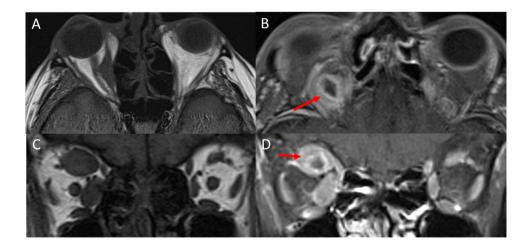
**Breast cancer** Breast cancer is one of the most common primary tumours to metastasise to the orbit [75, 103]. Breast cancer may present with unilateral [95, 104, 105] or bilateral extraocular muscle involvement (Fig. 10A, B) [106–108]. The horizontal recti are most commonly involved; however, involvement of the obliques has also been reported [85, 105, 108]. It commonly presents with fusiform muscle enlargement of affected muscles with tendon sparing [85, 108–112]. On MRI, the lesion is iso-hyperintense on T1- and

T2-weighted imaging (Fig. 10C, D) [95, 104, 110, 113] with homogenous contrast enhancement [105, 112, 114].

Melanoma Melanoma most commonly involves the medial rectus muscle [95, 115–117]. Well-defined focal nodular enlargement of muscles without involvement of the anterior tendon is most common [115, 116, 118]. On CT, the affected muscles are isodense compared to unaffected muscles. On MRI, the lesion is iso-hyperintense on T1 and hypo-isointense on T2 imaging. Heterogenous contrast enhancement with areas of central hypointensity reflecting haemorrhage, and increased peripheral rim enhancement can be seen (Fig. 11) [111, 115, 119]. Atypical presentations showing

Fig. 10 Breast cancer metastasis. Case 1-axial CT image of the orbits (A) demonstrating focal enlargement of the right medial medial rectus (arrow). Coronal CT image (B) demonstrates enlargement of the right medial rectus which is approaching the optic nerve. Case 2-axial T1-weighted MRI (C) demonstrating isointense enlargement of the medial and lateral recti muscles and intraconal fat infiltration. Coronal T1-weighted MRI (D) shows diffuse enlargement of the right extraocular muscles with intraconal fat infiltration





**Fig. 11** Melanoma metastasis. An axial T1-weighted MRI (**A**) shows isointense nodular enlargement of the right medial rectus muscle without involvement of the anterior tendon. An axial, contrastenhanced, fat-supressed T1-weighted image (**B**) through the superior orbit shows an area of central hypointensity (arrow) with increased peripheral rim enhancement. Coronal T1-weighted MRI (**C**) shows

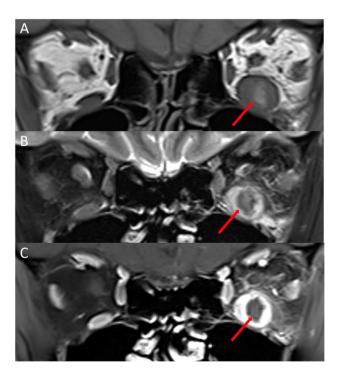
isointense enlargement of the right medial rectus and superior rectus muscles. A coronal, contrast-enhanced, fat-supressed T1-weighted image (**D**) demonstrates enlargement and enhancement of the right medial and superior rectus muscles, with a central area of hypoenhancement in the right superior rectus (arrow)



cystic masses with fluid–fluid levels have also been reported [118].

Neuroendocrine tumour Neuroendocrine tumours originate from the enterochromaffin cells of the gastrointestinal tract or bronchial lining. Unilateral presentation is most common with involvement of any of the extraocular muscles including the obliques [120–122]. Well-defined focal intramuscular masses are seen on imaging [95, 120, 122–125]. The masses may be of a significant size causing compression of adjacent structures including the optic nerve [95, 124–126]. They are isointense on T1 and hypo-isointense on T2-weighted imaging [95, 123, 127]. Heterogenous contrast enhancement can be seen [122, 124, 128]. Intralesional haemorrhage may be seen as ovoid areas of low signal intensity following contrast administration (Fig. 12) [129, 130].

*Gastric adenocarcinoma* Gastric adenocarcinoma presents with diffuse muscle enlargement [117, 131, 132]. Nodular and fusiform enlargement of muscles has also been reported [133, 134]. The diffuse enlargement may involve the anterior muscle tendon [132]. Gastric adenocarcinoma appears isointense on T1 and iso-hyperintense on T2-weighted imaging [131, 134].



**Fig. 12** Neuroendocrine tumour metastasis. Coronal T1-weighted MRI (**A**), fat-suppressed T2-weighted MRI (**B**), and fat-suppressed contrast-enhanced T1-weighted MRI (**C**) showing enlargement of the left inferior rectus muscle with intralesional haemorrhage (arrows) suggested by the hyperintensity on T1-weighted scan (**A**) and hypointensity on fat-suppressed T2-weighted (**B**) and fat-suppressed T1-weighted contrast-enhanced scans (**C**)



# **Amyloidosis**

Amyloidosis is a multisystemic disease with deposition of amyloid protein occurring in various parts of the body. Amyloidosis can uncommonly infiltrate the extraocular muscles presenting with unilateral or bilateral involvement of the extraocular muscles [135–137]. The horizontal recti are most commonly involved [138]. It usually causes tendon sparing fusiform enlargement of the affected muscles [138–142] and can mimic TED particularly when it involves the inferior or medial recti [142, 143]. Calcified lesions infiltrating the extraocular muscles can however be seen on CT helping to differentiate it from TED [139, 142]. Adjacent hyperostosis and bony irregularity can support a diagnosis of amyloidosis [144]. On T1-weighted imaging, the enlarged muscles are isointense to the extraocular muscles [137, 139, 141]. On T2-weighted imaging, hypointense lesions can be seen reflecting the amyloid involvement (Fig. 13) [135, 139]. Contrast-enhanced scans may reveal heterogenous enhancement with patchy areas of reduced enhancement [135, 140, 141].

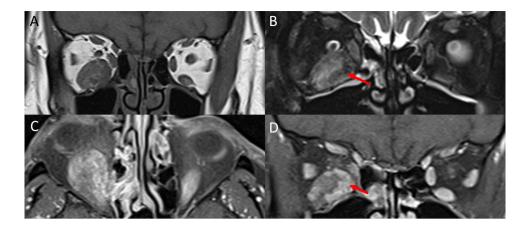
#### **Vascular conditions**

Vascular malformations can cause extraocular muscle enlargement. Arteriovenous malformations cause an increase in pressure within the cavernous sinus resulting in decreased venous outflow from the superior ophthalmic vein. The resultant elevated capillary pressure causes enlargement of the extraocular muscles.

Primary vascular malformations of the orbit can also lead to extraocular muscle enlargement by direct infiltration of the muscles, arteriovenous shunting or haemorrhage within the muscles. This is a separate section later anyway. Intraorbital arteriovenous malformations or AV malformations secondary to retinoencephalofacial angiomatosis or facial angiomatosis have also been reported to cause EOME [145–147].

# Carotid-cavernous fistula

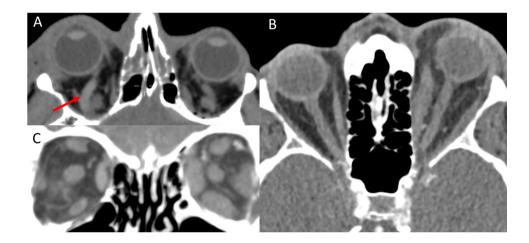
The description of the pattern of muscle enlargement in patients with carotid-cavernous fistulas (CCF) is limited in the literature. Accompanying radiological signs can often help aid the diagnosis. These signs include a dilatation of the superior ophthalmic vein which is seen in more than 80% of CCF cases (Fig. 14A) [46, 147, 148]. Other supportive findings include internal signal void within the cavernous sinus, and prominent venous drainage in the anterior, posterior or lateral venous sinuses [149]. Unilateral enlargement of multiple extraocular muscles can occur (Fig. 14B, C). Enlargement of the extraocular muscles will generally involve all the muscles on the affected side and may be accompanied



**Fig. 13** Intramuscular amyloidosis. Coronal T1-weighted MRI (**A**) demonstrating enlargement of the right inferior rectus muscle with areas of relative hyperintensity. Coronal T2-fat-suppressed MRI (**B**) shows enlargement and hypointense lesions of the right inferior rectus muscle (arrow). An axial fat-suppressed contrast-enhanced

T1-weighted MRI (**C**) shows heterogenous pattern of enhancement of the right inferior rectus. Coronal fat-suppressed contrast-enhanced T1-weighted MRI (**D**) demonstrated a heterogenous pattern of enhancement, with areas of increased enhancement corresponding to T2-hypointense areas (arrow)

Fig. 14 Carotid-cavernous fistula. Case 1—axial CT (A) of the orbits demonstrates enlargement of the right superior ophthalmic vein (arrow) and proptosis in a patient with a right direct carotid-cavernous fistula. Case 2—axial CT (B) showing left proptosis, enlargement of the left medial rectus muscle along with left globe tenting. Case 2—coronal CT (C) demonstrating enlargement of the left extraocular muscles



by intraconal fat stranding [149]. Expansion and bulging of the lateral wall of the cavernous sinus is more prominent on the fistula side [148, 150]. If the intercommunicating vessels within the cavernous sinus are large enough, there may be bilateral expansion of the cavernous sinuses and involvement of both eyes [148].

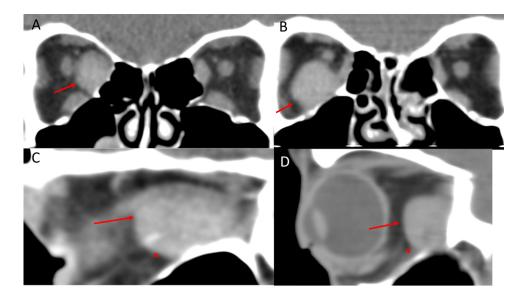
# Spontaneous haemorrhage related to the extraocular muscles

Nontraumatic orbital haemorrhage within the extraocular muscle belly or related to the muscle sheath can occur spontaneously in the absence of underlying vascular malformations. It usually presents in older females often with a history of hypertension and hyperlipidaemia [151]. Intra/juxtamuscular haemorrhage has a distinct radiological appearance which can help make the diagnosis and prevent unnecessary further investigations.

Intramuscular haemorrhage most commonly presents with unilateral involvement of the inferior rectus muscle [151, 152]. A large, often well-defined mass with an anterior rounded border and posterior tapering edge towards the orbital apex ("tear drop" appearance on axial scans and "beached whale" appearance on sagittal when involving the inferior rectus) is characteristic (Fig. 15) [151]. On CT, the haematoma is hyperdense if acute and hypo-isodense if subacute [153]. A fluid level may be observed in the clot, further supporting a diagnosis of a haematoma [152, 154]. Haemorrhage on MRI has a variable appearance depending on the age of the blood, stages of the clot and MRI sequence used. A fresh clot with deoxyhaemoglobin (< 2 days) is isointense on T1 and hypointense on T2W-MRI [153]. As the clot changes to intracellular methemoglobin, the signal on T1W-MRI becomes hyperintense and hypointense on T2W-MRI [153]. Extracellular methemoglobin is hyperintense on T2 and is seen between 2 weeks and 2 months after



Fig. 15 Two cases of Spontaneous haemorrhage related to extraocular muscles. Case 1—coronal (A) and sagittal (C) CT scans showing a large, well-defined mass in the right medial rectus with an anterior rounded border (arrow) and posterior tapering edge (\*) towards the orbital apex. The sagittal image has a 'bleached whale' appearance suggestive of intramuscular haemorrhage. Case 2—coronal (B) and sagittal (D) CT scans showing a large, well-defined mass in the right inferior rectus with an anterior rounded border (arrow) and posterior tapering edge (\*) towards the orbital apex



the initial haemorrhage. Serial MRIs can help to identify these changes and support the diagnosis of an intramuscular haemorrhage. Although gradient sequences are distorted at the level of the orbit, some sequences such as the  $B_0$  of the diffusion sequence may demonstrate low signal in the acute phase (susceptibility artefact) suggesting haemorrhage.

#### Infective conditions

# Cysticercosis

Cysticercosis is a parasitic infection caused by Cysticercus cellulosae, which is endemic in regions with poor sanitation. Orbital or extraocular muscle cysticercosis most commonly presents in teenagers [155, 156]. Extraocular muscle cysticercosis presents unilaterally with single muscle involvement of any of the extraocular muscles including the obliques [155]. Rath, Honavar, Naik, Anand, Agarwal, Krishnaiah and Sekhar [156] reviewed 138 cases of extraocular muscle cysticercosis and reported the superior rectus muscle to be most commonly involved, followed by the inferior, medial and lateral recti in descending order. On CT, imaging reveals an intramuscular hypodense, ring enhancing cystic lesion (Fig. 16), often in association with a scolex in approximately 50% of cases [156-158]. On T2-weighted imaging, a round hyperintense nodule can be seen on a hypointense background [159, 160] and on T1-fatsuppressed post contrast, a ring enhancing lesion with a hypointense central nodule is typical [161, 162].

## **Hydatid** cyst

Hydatid cysts result from infection with the parasite *Echinococus granulosus* and can result in cyst formation anywhere in the body. Extraocular muscle hydatid cysts are



**Fig. 16** Cysticercosis. Contrast-enhanced CT of the orbits demonstrates a hypodense cystic mass within the left superior rectus with peripheral rim enhancement (arrow)

rare. On MRI, it presents as a multiloculated cystic lesion which is hypointense on T1 and demonstrates peripheral rim enhancement following contrast administration [163].

# **Pyomyositis**

Pyomyositis is an acute bacterial infection of skeletal muscle, usually caused by *Staphylococcus aureus*. It has a characteristic appearance on imaging. It presents unilaterally with involvement of any of the extraocular muscles. On CT, a round-oval hypodense lesion with rim enhancement is characteristically observed [164–166]. On T1-weighted imaging, a hypointense central lesion with rim enhancement can be seen, alongside oedema and enlargement of the affected muscle [166–168]. An abscess within the muscle usually results in restricted diffusion.



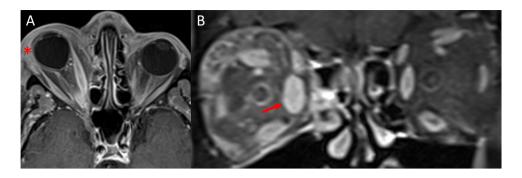


Fig. 17 Orbital cellulitis. Axial fat-suppressed contrast-enhanced T1-weighted MRI (A) demonstrating preseptal soft tissue swelling (asterisks, \*), enlargement and enhancement of the right medial rectus muscle and right axial proptosis. Coronal fat-suppressed con-

trast-enhanced T1-weighted MRI (**B**) demonstrates enlargement of the right medial rectus muscle (arrow) with associated intraconal fat stranding

#### **Orbital cellulitis**

Orbital cellulitis most commonly occurs from direct extension of adjacent sinus infection or preseptal cellulitis and imaging may reveal inflammation in these adjacent areas. Any of the extraocular muscles can be enlarged in orbital cellulitis (Fig. 17) [169]. Associated features include intraconal fat stranding, dacryoadenitis and optic perineuritis [170]. Potential complications include orbital abscess formation, intracranial extension and cavernous sinus thrombosis [170, 171].

# **Conclusion**

Extraocular muscle enlargement is most commonly due to thyroid eye disease; however, it can occur secondary to a number of other inflammatory, vascular or neoplastic conditions. Imaging is playing an increasingly important role in the characterisation of orbital diseases and can aid the diagnosis and management of orbital conditions. Ultimately, the diagnosis will be made in conjunction with an understanding of the patient's demographics, past medical history and presentation. In cases where there is diagnostic uncertainty, an orbital biopsy may be required for definitive diagnosis.

Funding Open Access funding enabled and organized by CAUL and its Member Institutions

#### **Declarations**

**Ethical approval** This article does not contain any studies with human participants or animals performed by any of the authors.

**Conflict of interest** The authors declare no competing interests.

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