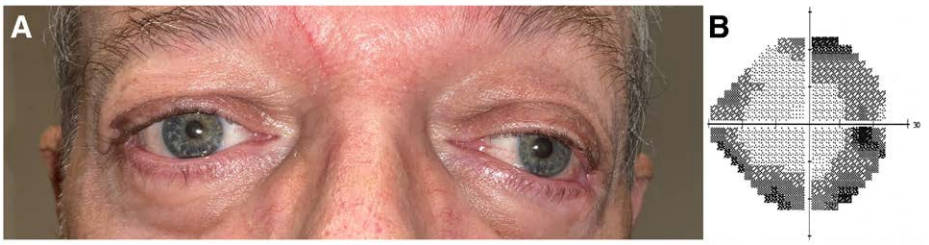

FIG. 4. At 2-month follow-up, the patient's swelling had almost completely resolved, but he had a new left exotropia that was not present before the incident (**A**). His vision had improved to 20/20 OD, but remained NLP OS. A 24-2 humphrey visual field (HVF) test of the OD (**B**) showed significant field constriction with many false negatives.

prothrombin time (value >100 seconds, reference range 12–14.5) and partial thromboplastin time (132 seconds, reference range 25–35 seconds). He received 4,000 units of 4-factor prothrombin complex concentrate and 10 mg of Vitamin K with an improvement in his INR to 1.72 18 hours after arrival. Unfortunately, the patient continued to bleed as his INR was down-trending and his vision deteriorated to NLP OU with IOP in the 40s OU despite bilateral canthotomy and upper and lower cantholysis (Fig. 3). Given his extensive ongoing hemorrhage, orbital decompression was not pursued due to the risks of profuse bleeding. A regimen of combined dorzolamide-timolol, brimonidine, and acetazolamide 250 mg was started twice daily and cool compresses were applied to his wounds to attempt to slow bleeding. He was closely monitored throughout his hospitalization and at the time of discharge, his vision improved to 20/70 OD, but remained NLP OS with normal IOP OU.

At 2-week follow-up, the swelling had largely resolved and his vision had improved to 20/25 OD, but remained NLP OS with IOP of 9 OD and 9 OS. At the subsequent 2-month follow-up, the patient's appearance was nearly normal and visual acuity was 20/20 OD and NLP OS with IOP of 17 OD and 14 OS off all drops (Fig. 4A). A 24-2 humphrey visual field (HVF) test showed significant constriction to the central 10 degrees of vision in all meridians (Fig. 4B).

DISCUSSION

Retrobulbar hemorrhage is itself an uncommon entity, but spontaneous nontraumatic orbital hemorrhage is even less common.[2] In a large review of 115 cases of nontraumatic orbital hemorrhages, the majority (90%) had a predisposing vascular lesion while only 3.5% had an underlying coagulopathy.[3] Cases of spontaneous retrobulbar hemorrhage due to underlying coagulopathy have been reported in patients with Vitamin K deficiency, dabigatran use, idiopathic thrombocytopenic purpura, hemophilia A, scurvy, and warfarin use.[4–7] The majority of reported cases are unilateral. This is, to our knowledge, the first reported case of spontaneous bilateral retrobulbar hemorrhage due to warfarin misuse.

In this patient, it is very likely the initial bleeding was triggered by Valsalva-like maneuvering from aggressive nose-blowing following nasal lavage. While a small amount of bleeding may not be dangerous in most patients, this patient's supratherapeutic INR (above the maximum lab threshold) prevented appropriate clotting which led to the accumulation of blood in the retrobulbar space. By the time the patient presented to the ED, surgical intervention was required given the development of bilateral orbital compartment syndrome.

In addition to presenting a previously unseen complication of warfarin misuse, this case also reinforces the importance of vigilant INR monitoring in all patients on warfarin. Current guidelines from the American Heart Association, recommend INR monitoring every 4 weeks in patients stable on warfarin and even more frequently when warfarin is first initiated.[8] With the rise of direct oral anticoagulants and their improved safety profile compared to warfarin regarding major bleeding events, this case should also remind providers that, where possible, direct oral anticoagulants should be considered in patients with atrial fibrillation requiring anticoagulation.[9]

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Tendon-Sparing Extraocular Muscle Enlargement Associated With Chronic Inflammatory Demyelinating Polyradiculoneuropathy

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Abstract: **Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) is an inflammatory, sensorimotor**

polyneuropathy. It has presented with a variety of orbital and neuro-ophthalmic manifestations, including cranial nerve hypertrophy and a single case of extraocular muscle enlargement. The authors present a second case of tendon-sparing, extraocular muscle enlargement, resulting in new-onset diplopia and strabismus in a teenager with CIDP. The workup ruled out alternative causes of extraocular muscle enlargement, such as hyperthyroidism, inflammation, or malignancy. As with other cases of CIDP, management involved a combination of immunoglobulin therapy and anti-inflammatory medications. The patient experienced resolution of his symptoms, and radiologic improvement was noted in the muscle enlargement. As many CIDP patients have a favorable treatment response and long-term prognosis, awareness of this rare disease with an early and accurate diagnosis is important.

Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) is an acquired, immune-mediated sensorimotor polyneuropathy that presents to ophthalmologists with ophthalmoplegia, proptosis, papilledema, optic neuropathy, and pupillary abnormalities associated with cranial nerve hypertrophy.[1] The authors present a case of diplopia and esotropia with tendon-sparing extraocular muscle (EOM) enlargement in a teenager with CIDP.

CIDP is characterized by symmetric, progressive, or relapsing proximal and distal muscle weakness with peripheral sensory loss.[1] Clinical examination, nerve conduction studies, lumbar puncture, MRI, and nerve biopsy are used to make the diagnosis.[2] CIDP is highly responsive to treatment with immunosuppressants or immunomodulation, making early diagnosis important.[3]

Only one other case of CIDP-associated EOM enlargement has been reported, and this was in a 67-year-old man treated with decompression surgery.[3] To date, this is the first report of CIDP-associated EOM enlargement in a pediatric patient successfully managed without surgical intervention. This report adhered to the ethical principles outlined in the Declaration of Helsinki as amended in 2013 and is compliant with the Health Insurance Portability and Accountability Act.

CASE PRESENTATION

A 15-year-old male with a history of CIDP on weekly immunoglobulin therapy and monthly dexamethasone presented with 4 days of constant binocular diplopia.

His CIDP was diagnosed 5 years prior when the patient presented with upper and lower extremity weakness. Lumbar puncture demonstrated elevated cerebrospinal fluid protein levels along with a normal cerebrospinal fluid white blood cell count—a finding known as albuminocytologic dissociation that can be seen in acute inflammatory demyelinating polyradiculoneuropathy. MRI brain and lumbar spine revealed no neurologic abnormalities. Nerve conduction studies demonstrated evidence of diffuse sensorimotor polyneuropathy with predominantly demyelinating features. The patient was diagnosed with Guillain-Barre syndrome, a form of acute inflammatory demyelinating polyradiculoneuropathy. Intravenous immunoglobulin (IVIG) was initiated but then discontinued due to severe headaches and nausea. Treatment was changed to oral prednisone. Over the next few months, while tapering the steroids, the patient experienced repeat episodes of weakness in his upper and lower extremities, most prominent in the distal extremities. Following these episodes, the family sought an outside provider, who treated the patient with approximately 2 years of antibiotics for Lyme disease, though laboratory testing for this was negative. During this time, the patient's distal upper and lower extremity weakness persisted.

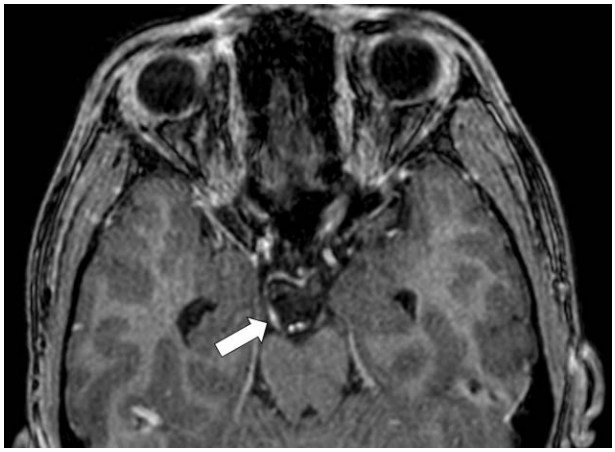

FIG. 1. MRI. Axial post-gadolinium fat saturated T1-weighted MRI demonstrating enhancement of right oculomotor nerve (white arrow). Image quality is limited due to motion artifacts.

After this period, he returned to his original pediatric neurologist due to the persistent distal weakness. Repeat nerve conduction studies were consistent with CIDP and subcutaneous immunoglobulin of 0.2 g/kg/week was trialed. Laboratory tests at this time revealed normal thyroid function testing, negative extractable nuclear antigens, nuclear antibody screen, thyroid peroxidase antibodies, thyroglobulin antibody, and thyroid stimulating immunoglobulin. Antiacetylcholine receptor antibodies and antimuscle-specific kinase antibodies were also noted to be negative. A few months into treatment with subcutaneous immunoglobulin, the patient experienced worsening weakness and balance, as well as a transient episode of horizontal, binocular diplopia. A dose of IVIG (2 g/kg total) and a pulse dose of oral dexamethasone (4 days of 40 mg/day), followed by resumption of subcutaneous immunoglobulin at a higher weekly dose (0.4 g/kg/week) and monthly pulse dexamethasone (4 days of 40 mg/day) was started. A few weeks following the 2nd pulse dexamethasone treatment, the patient then developed a constant horizontal, binocular diplopia.

The patient's full ophthalmic examination was unremarkable, except for esotropia with a mild abduction restriction of the OS. MRI orbits demonstrated abnormal enhancement of the following cranial nerves: right oculomotor, bilateral trigeminal, left abducens, bilateral facial, and bilateral vestibulocochlear (Fig. 1). There was evidence of spindle-shaped enlargement of all rectus and superior oblique muscles with sparing of the myotendinous junctions (Fig. 2). Mild infiltration of bilateral retrobulbar fat and crowding of the orbital apices, resulting in venous congestion, was also noted.

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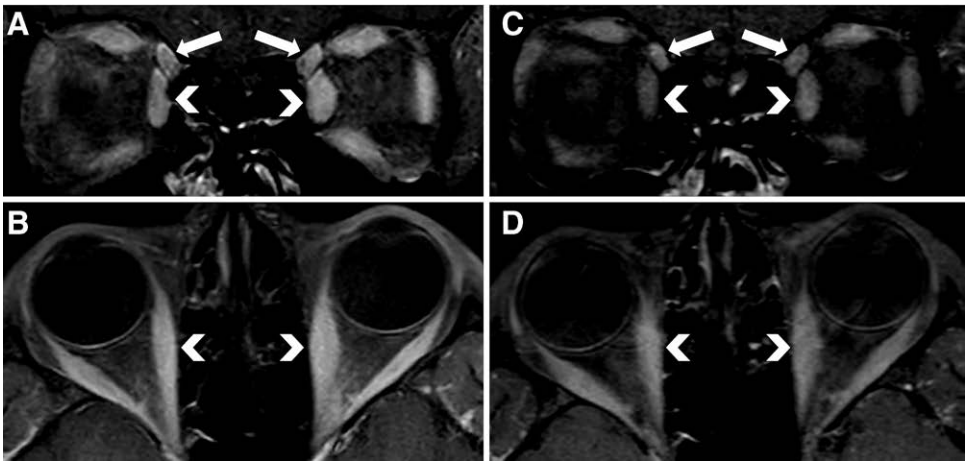

FIG. 2. MRI. Post-gadolinium T1-weighted spectral presaturation with inversion recovery MRI demonstrated enlarged superior oblique and medial rectus muscles prior to treatment with IVIG and pulse-dose steroids: (**A**) coronal (**B**) axial. **C**, Coronal postgadolinium T1-weighted fat suppression MRI showed a decrease in size of the superior oblique and medial rectus muscles following treatment. **D**, Axial postgadolinium T1-weighted fat suppression MRI demonstrated a decrease in size of the medial rectus muscle (chevron arrow indicates medial rectus, thick arrow is superior oblique).

Workup for other systemic causes including thyroid eye disease, IgG4-related disease, granulomatosis with polyangiitis, and sarcoidosis remained negative. The patient's family deferred a definitive biopsy of EOM.

Treatment was switched to IVIG 1 g/kg total every 3 weeks, with pulse-dose steroids consisting of 40 mg of dexamethasone daily for 4 days every 4 weeks, resulting in the resolution of the double vision over several months. A repeat MRI 5 months later demonstrated decreased size of medial, inferior, and lateral rectus muscles and persistent, but decreased enhancement of the right 3rd cranial nerve (Fig. 1). Follow-up examination 6 months later was significant for the return of full extraocular movements and resolution of esotropia. He continued to receive treatment with IVIG and pulse-dose steroids for 1 year, followed by IVIG alone for an additional year. All medical therapy was then discontinued and the patient has remained relapse-free now nearly one full year later.

DISCUSSION

Extraocular muscle enlargement is commonly associated with thyroid eye disease, IgG4-related ophthalmic disease, sarcoidosis, and other orbital inflammatory, infectious, and neoplastic processes. Muscle tendon involvement is a significant distinguishing feature; in thyroid eye disease, the EOM enlargement is tendon-sparing, similar to the case presented here. However, in thyroid eye disease, the rectus muscles are affected most commonly, while oblique muscle involvement is rarer. In the presented case, there was superior oblique involvement in addition to all the rectus muscles. In addition, all thyroid antibody testing was negative. Although proptosis, extraocular motility restriction, and diplopia have been noted to occur in CIDP, these were the result of cranial nerve hypertrophy rather than due to EOM enlargement.[1]

In a large retrospective study of 132 patients with CIDP, the most commonly involved cranial nerves were as follows: facial and glossopharyngeal at 9%, oculomotor and abducens at 5%, trigeminal at 3%, vestibulocochlear at 2%, and optic nerve at 1%.[4] In our case, the following nerves were involved: right oculomotor, bilateral trigeminal, left abducens, bilateral facial, and bilateral vestibulocochlear. Clinically, the left abduction limitation in our patient likely reflected both abducens nerve involvement and EOM enlargement.

The only other case of CIDP-related EOM enlargement and proptosis was in a 67-year-old male.[3] The etiology was postulated to be due to inflammation of the nerve endings and adjacent muscle fibers resulting in tissue expansion, specifically muscle belly enlargement that spared the tendon.[3] Biopsy of EOMs was not performed due to concern of further worsening the clinical presentation.[3] The patient was treated with bilateral orbital decompression and plasma exchange.[3] Orbital decompression consisted of the lateral and medial walls and the medial orbital floors, with significant improvement in the proptosis and resolution of globe subluxation.[3] However, there was no change in strabismus and diplopia, for which the patient deferred additional surgery.[3] In this case and ours, no optic nerve compromise was noted.

Many patients with CIDP have a favorable treatment response and long-term prognosis. However, 13%–24% demonstrate severe disability despite treatment with about 40% of these more severe cases requiring continuous immunosuppressants.[5] In our case, the patient's symptoms and inflammation resolved on IVIG and pulse-dexamethasone therapy. One year after discontinuing all treatment modalities, he is still doing well. This is the second reported case of CIDP-related EOM enlargement and the first in a pediatric patient. In addition to the more commonly reported cranial nerve hypertrophy, CIDP can also present with EOM enlargement and diplopia. In our case, the patient was successfully managed medically without any surgical intervention.

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Superior Ophthalmic Vein Thrombosis Associated With Asymptomatic COVID-19 Infection

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Abstract: **Superior ophthalmic vein thrombosis is a rare condition scarcely described in clinical literature with potentially severe consequences including permanent vision loss. This report details the case of a 70-year-old woman who presented with acute binocular horizontal diplopia, relative proptosis of the OD by 4 mm, and pain OD. On exam, visual acuity was 20/20 OD and 20/30 OS with full extraocular movements. CT revealed proptosis OD with a thrombosed superior ophthalmic varix. Evaluation for etiology of hypercoagulability was unremarkable, although the patient did have an asymptomatic COVID-19 infection 1 month prior. To the authors' knowledge, this is the first reported case of superior ophthalmic vein thrombosis secondary to an asymptomatic COVID-19 infection.**

Superior ophthalmic vein thrombosis (SOVT) is a rare condition with potentially vision-threatening complications with early diagnosis and intervention necessary to prevent irreversible complications. A previous report has described a case of SOVT secondary to a severe COVID-19 infection with large saddle pulmonary embolus. Unlike the previous case, this patient only had an asymptomatic COVID-19 infection without any other signs of systemic hypercoagulability.

The collection and evaluation of protected patient health information were compliant with the Health Insurance Portability and Accountability Act, and this report adheres to the ethical tenets of the Declaration of Helsinki.

CASE PRESENTATION

A 70-year-old woman presented with acute onset binocular horizontal diplopia, moderate OD pain, and relative proptosis of the OD. Visual acuity was 20/20 OD and 20/30 OS, intraocular pressure 14 OD and 11 OS, and there was no relative afferent pupillary defect. Extraocular movements were full bilaterally. Hertel exophthalmometry measured relative OD proptosis of 4 mm. On alternate cover testing, 10 prism diopters of esotropia were measured in primary and lateral gazes. Dilated fundus exam was unremarkable.

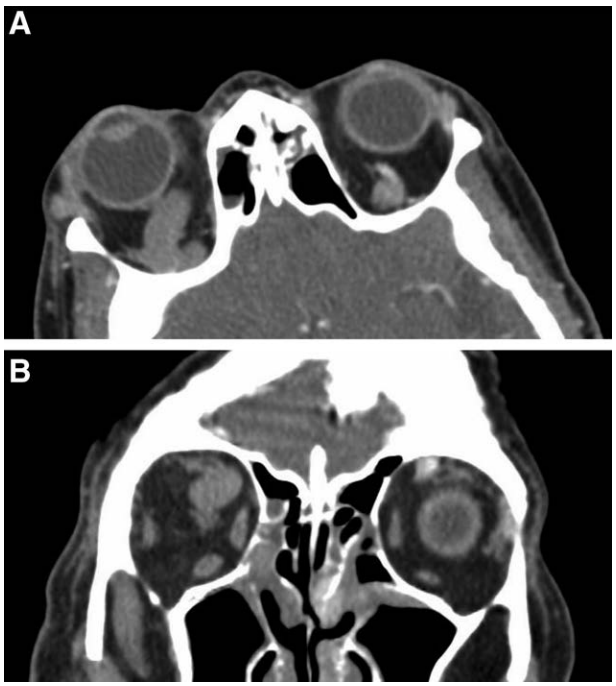
The patient's past medical history included glaucoma suspect, hypertension, and chronic obstructive pulmonary disease. Her medications were amlodipine, chlorthalidone, lisinopril, omeprazole, and vitamin D3. She was an active smoker and had smoked 1 pack per day for 45 years. She denied headaches, nausea, vomiting, and changes in visual acuity. One month prior, she had a COVID-19 infection that did not necessitate medical treatment.

CT scans revealed proptosis OD, a thrombosed superior ophthalmic vein (Fig.), and an incidental 1.1 cm left middle cerebral artery aneurysm. Further, the workup had an erythrocyte sedimentation rate elevated to 62 mm/hour. C-reactive protein, Angiotensin-converting enzyme level, rheumatoid factor, lupus anticoagulant, Antineutrophilic cytoplasmic antibody, antithrombin III activity, prothrombin, factor V Leiden, protime-international normalised ratio, and thyroid stimulating hormone were negative. The patient had not had any prior head imaging for comparison.

Further characterization of the right orbital varix and middle cerebral artery aneurysm was done with CT angiography, revealing an enlarged superior ophthalmic vein without enhancement compatible with thrombosis and an unchanged left middle cerebral artery aneurysm. A week after the initial presentation, she reported improving diplopia and resolution of eye pain. Exam showed orthophoria and decreased proptosis. Anticoagulation was deferred in the setting of middle cerebral artery aneurysm and clinical improvement of the patient. At the final follow-up, 7 months after the initial presentation, the patient reported complete resolution of symptoms. Hertel measured OD proptosis of 1.5 mm; the exam was otherwise unchanged. CT head done for another indication 9 months following the initial presentation revealed a slightly decreased caliber of the thrombosed right superior ophthalmic vein.

DISCUSSION

SOVT is a rare condition with only 93 total cases in literature from 1975 to 2019.[1,2] Impaired venous drainage typically manifests as sudden onset unilateral orbital pain, proptosis,



Orbital CT scan with contrast. The axial (**A**) and coronal (**B**) views demonstrate the large superior ophthalmic vein thrombosis.

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