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Letter to the Editor: New Observation

Orbital Myositis Preceding Presumed Giant Cell Myocarditis

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We describe a middle-aged man who presented with a semi-acute onset of diffuse limitation of extraocular motility with enlargement of all extraocular muscles on imaging. He was treated for presumed orbital myositis with oral corticosteroids. When prednisone was tapered off 2 months later, he developed shortness of breath and then, shortly after, cardiogenic shock. He was eventually diagnosed with presumed giant cell myocarditis (GCM). This case emphasizes that patients with GCM can rarely present to neurologists with orbital myositis as the initial manifestation of a deadly disease.

A 61-year-old man presented to the emergency department (ED) with a 3-week history of bilateral ptosis and binocular vertical diplopia. There was no associated pain or periorbital edema. His medical history was significant for basal cell carcinoma of the face and mild inflammatory bowel disease. He did not take any medications. CT brain demonstrated enlargement of all extraocular muscles (Figure 1). He was initially thought to have thyroid eye disease versus idiopathic orbital myositis and started on treatment with 50 mg of oral prednisone.

On initial presentation to neuro-ophthalmology 1 week later, vision was 20/20 in each eye. He exhibited severe non-fatigable bilateral ptosis with decreased levator palpebrae superioris function and symmetric limitation of extraocular movements in all directions of gaze. He had a small comitant esotropia on alignment testing. Neurological examination demonstrated normal upper and lower extremity strength, normal gait, normal coordination testing and normal deep tendon reflexes. He reported significant interval improvement in ptosis 3 days after starting oral prednisone. He had mildly elevated C-reactive protein (5 mg/L, normal <3) and thyroid-stimulating hormone levels (6.48, normal <4.78 mIU/L). Levels of T3, T4, anti-thyroid antibody, acetylcholine receptor antibody, anti-muscle-specific kinase, anti-LRP4 and anti-agrin titers were all normal. A CT scan of the chest did not show evidence of thymoma. Serum anti-GB1b level looking for the Miller Fisher variant of Guillain–Barré was weakly positive.

At 1-month follow-up, he demonstrated improvement in extraocular motility in all directions of gaze and had only mild ptosis bilaterally. He otherwise felt systemically well. Prednisone was tapered to 30 mg and then by 10 mg every 2 weeks. One month later, after being on a cruise where he was very physically active and felt well, he presented to a local ED with a new onset of severe dyspnea and lethargy. He was hypotensive with very low oxygen saturation (78%). Troponin level was 44,000 ng/L (normal <26 ng/L), and

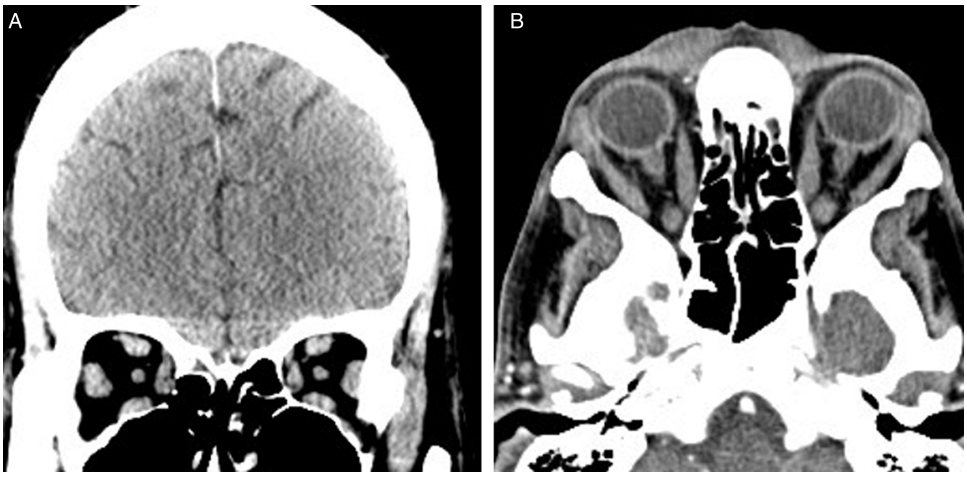


Figure 1. (A) CT brain, coronal sequence, demonstrating mild to moderate enlargement of all extraocular muscles. (B) CT brain, axial sequence, showing enlargement of extraocular muscles sparing the tendons.

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Table 1. Documented cases of giant cell myocarditis with associated ocular involvement

Study	Patient age and gender	Country	Comorbidities	Initial presenting clinical features	Diagnostic findings	Timeline (orbital myositis to GCM)	Cardiac features	Treatment	Final patient outcome
Klein et al. (1989)[5]	65-year-old woman	United States	Not specified	Bilateral ophthalmoplegia	CT orbits: enlarged extraocular muscles Biopsy: Multinucleated giant cells in the myocardium	~ 1 month	Not specified	Not specified	Fatal cardiac arrhythmia
Kattah et al. (1990)[6]	37-year-old woman	Costa Rica	Thyroid disease (unconfirmed)	Bilateral proptosis; ophthalmoplegia; chemosis;	CT orbits: extraocular muscle enlargement with tendon sparing Biopsy: giant cells in cardiac, pharyngeal, and extraocular muscles	~ 18 months	Fatal cardiac arrythmia	Corticosteroids; azathioprine	Ventricular tachycardia and fibrillation leading to death
Stevens et al. (1996)[7]	22-year-old woman	United States	Asthma; vitiligo; several food allergies	Bilateral ophthalmoplegia; periorbital swelling and erythema	CT orbits: edema of extraocular muscles Biopsy: multinucleated giant cells, lymphocytes, monocytes, and eosinophils in the myocardium	~ 1 month	Cardiogenic shock	Steroids; cyclosporine; azathioprine; cardiac transplant 7 days after failure to improve with immunosuppression	Survived post-transplant
Kollmeier et al. (2006)[8]	39-year-old woman	Germany	Chronic pansinusitis	Fluctuating diplopia; pain with eye movements; limitation of supraduction in the right eye; left upper lid ptosis	MRI brain and orbits: pansinusitis Biopsy: multinucleated giant cells in the myocardium	~ 2 months	Right bundle branch block; cardiogenic shock	Corticosteroids; left ventricular assist device	Death following multi-organ failure
Lind-Ayres et al. (2008)[9]	14-year-old girl	United States	None reported	Diplopia; ptosis; pain in the left eye	CT orbits: superior rectus inflammation Biopsy: giant cells, lymphocytes, eosinophils, plasma cells	~ 2 months	Dilated cardiomyopathy; congestive heart failure	Corticosteroids; mycophenolate mofetil; diuretics; milrinone; anticoagulation; thymoglobulin	Discharged in stable condition
Ali et al. (2016)[10]	40-year-old man	United States	None reported	Bilateral eye pain; redness; limitation of eye movements	MRI orbits and lateral rectus biopsy confirmatory of orbital myositis Biopsy: multinucleated giant cells in the myocardium	~ 3 years	Ventricular tachycardia; right bundle branch block; arrhythmogenic right ventricular cardiomyopathy; cardiogenic shock	Corticosteroids; amiodarone; cardioversion; implantable cardioverter-defibrillator; intra-aortic balloon pump; extracorporeal membrane oxygenation	Uncontrollable bleeding from extracorporeal membrane oxygenation cannulation site resulting in death
Garg et al. (2017)[11]	50-year-old man	United States	Rheumatoid arthritis	Diplopia; ophthalmoplegia; ptosis	MRI orbits: extraocular muscle hypertrophy Biopsy: Multinucleated giant cells in the myocardium	~ 2 weeks	Incomplete right bundle branch block; atrioventricular block; syncope; ventricular tachycardia	Plasmapheresis; ventricular assist device; corticosteroids; mycophenolate mofetil; tacrolimus; cardiac transplant	Survived post-transplant

Table 1. Documented cases of giant cell myocarditis with associated ocular involvement (*Continued*)

Study	Patient age and gender	Country	Comorbidities	Initial presenting clinical features	Diagnostic findings	Timeline (orbital myositis to GCM)	Cardiac features	Treatment	Final patient outcome
Kitano et al. (2023)[12] (patient #1)	59-year-old woman	Japan	None reported	Bilateral ophthalmoplegia; fever; presyncope	CT brain: normal Biopsy: multinucleated giant cells, eosinophils, and lymphocytes in the myocardium	~ 2 weeks	Cardiomegaly; ventricular tachycardia; cardiopulmonary arrest	Intra-aortic balloon pump; corticosteroid pulse	Subarachnoid hemorrhage resulting in death
Kitano et al. (2023)[12] (patient #2)	56-year-old woman	Japan	Hashimoto's thyroiditis	Right upper lid ptosis; diplopia	MRI orbits: enlargement of lateral rectus Biopsy: multinucleated giant cells, eosinophils, and lymphocytes in the myocardium	~ 4 days	ST/T-wave abnormalities	Corticosteroids	Stable post-discharge

GCM = giant cell myocarditis.

brain natriuretic peptide level was >70,000 pg/mL (normal <100 pg/mL), and creatinine kinase level was 1446 U/L (normal <240 U/L). Electrocardiogram demonstrated ventricular tachycardia. He was diagnosed with cardiogenic shock. An echocardiogram demonstrated severe left ventricular dysfunction. Coronary angiography demonstrated patency of all cardiac vessels. He went on to develop pump-failure-related fulminant liver failure, acute kidney injury and peripheral limb ischemia; thus, treatment with extracorporeal membrane oxygenation (ECMO) commenced. Given his explosive presentation, patent cardiac vasculature and overwhelming failure of the cardiac pump, primary myocarditis was thought to be the diagnosis, with GCM being the most likely culprit. The patient was on maximal medical therapy over the course of his hospital admission, and 2 weeks later, ECMO was discontinued, and a cardiac biopsy was planned. However, the day after ECMO was stopped (2.5 months after his initial presentation with ocular symptoms), he developed a cardiac arrest and died. His family declined an autopsy. A final presumptive diagnosis of GCM and orbital myositis was made.

GCM is a T-cell-mediated inflammatory condition affecting the myocardium and is almost universally fatal if cardiac transplantation is not performed. It has a predilection for affecting young and healthy individuals; however, several conditions, including orbital myositis, myasthenia gravis, thymoma and inflammatory bowel disease, have all been associated with GCM.[1,2] There is significant variation in the clinical presentation of GCM, with acute myocardial injury being the main feature presenting clinically with progressive heart failure, conduction abnormalities, ventricular arrhythmia and cardiogenic shock.[2] Pathologically, the findings are similar to those seen in giant cell arteritis: lymphocyte infiltration of affected myocardium with plentiful macrophages and giant cells. The mainstay of treatment for GCM includes immunosuppressive therapy (e.g., corticosteroids), with definitive treatment being cardiac transplantation, although the disease can recur in up to 25% of transplants.[2]

Orbital myositis is an inflammatory condition that affects the extraocular muscles and results in varying degrees of ophthalmoplegia, sometimes accompanied by pain.[3] A possible association between orbital myositis and GCM has been previously suggested,[4] although the exact pathogenic mechanism bridging both conditions is unknown.[4] To the best of our knowledge, nine case reports have previously described patients with GCM and preceding orbital myositis (Table 1). The age range was 14–65 years, and seven of the nine patients were women. Most presented with diplopia, ptosis and/or ophthalmoplegia. The timeline between the onset of ocular and cardiac symptoms varied between 4 days and 3 years. Only two patients (22%) underwent cardiac transplantation and survived.

Several studies have attempted to elucidate the underlying pathophysiology of GCM. Both the extraocular muscles and the heart muscle are striated muscles with high metabolic activity. Pathology of GCM is centered on T-lymphocytes infiltrating the myocardium, resulting in the production of interferon-γ and tumor necrosis factor and release of nitric oxide by macrophages.[4] It is plausible that T-cell cross-reactivity may target antigens expressed by both extraocular muscles and myocardial cells, resulting in inflammatory changes affecting both muscles.

In conclusion, this case report underscores the potential association between orbital myositis and GCM. Although GCM is rare, this case brings the association of orbital myositis and GCM to the attention of neurologists who are often the first ones encountering patients with orbital myositis.

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