PEDIATRIC ONCALL
CHILD HEALTH CARE

CASE REPORTS

ADOLESCENT PTOSIS UNMASKED: ANTI-PM/SCL MYOSITIS TARGETING THE SUPERIOR RECTUS

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ABSTRACT

Orbital myositis, a subtype of idiopathic inflammatory myopathy, rarely presents with isolated superior rectus involvement in adolescents, particularly associated with anti-PM/Scl antibodies.[1,2] We report a 16-year-old male with a 3-week history of left eye ptosis and ipsilateral headache. MRI revealed thickening of the left superior rectus muscle with proptosis of the left globe, mimicking infectious or neoplastic processes. Diagnostic workup suggested superior rectus myopathy. Autoimmune profiling identified positive myositis-specific anti-PM/Scl antibodies, consistent with PM/Scl myositis overlap syndrome. Prompt initiation of intravenous methylprednisolone (1 g/day for 3-5 days) led to rapid improvement in ptosis, headache, and proptosis, with near-complete resolution on follow-up imaging. This case underscores the diagnostic utility of myositis specific antibodies in steroid-responsive orbital inflammation, particularly in young patients without systemic features. Early recognition prevents misdiagnosis such as thyroid eye disease or infection, averting unnecessary biopsies or antimicrobials. PM/Scl-associated myositis typically features proximal weakness and interstitial lung disease in adults, but isolated orbital involvement in pediatrics is exceptional, highlighting the need for expanded antibody testing in atypical myopathies. This report adds to sparse pediatric literature on anti-PM/Scl orbital myositis, advocating for multidisciplinary care involving pediatric neurology, ophthalmology, and rheumatology.

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KEYWORDS

Superior rectus, Anti-PM/Scl, Orbital myositis.

Introduction

Anti-PM/Scl antibodies target the PM/Scl ribonucleoprotein complex, defining a rare myositis overlap syndrome with features of polymyositis, systemic sclerosis, and arthritis.[1,9] Predominantly affecting adults (mean age 50-60 years), it manifests with proximal muscle weakness (70-80%), sclerodactyly (50%), and mechanic's hands (40%), alongside elevated creatine kinase and myopathic electromyography.[2] Orbital involvement is exceedingly rare, comprising <1% of cases, and typically involves extraocular muscles diffusely rather than isolated superior rectus enlargement.[10] In pediatrics, incidence of myositis is 2-4 per million, with anti-PM/Scl positivity in <5% of juvenile cases, often linked to calcinosis or lung fibrosis.

Ptosis with proptosis evokes thyroid-associated orbitopathy (80% of cases), myasthenia gravis (MG), or infection, but steroid responsiveness and antibody specificity differentiate autoimmune orbital myositis. This case illustrates an adolescent presentation, emphasizing serological diagnosis amid negative infectious/neuromuscular tests. Early immunosuppression halts fibrosis, preserving vision and ocular motility.

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Case Report

A 16-year-old male presented with a 3-week history of progressive left eyelid drooping and headache, worse on eye movement. No diplopia, fever, trauma, or systemic symptoms like rash, dysphagia, or joint pains. Examination revealed left ptosis (margin-reflex distance -1.5 mm), proptosis (3 mm), and tender superior eyelid swelling; extraocular movements were full but painful; visual acuity and fundus were normal bilaterally. Laboratory tests showed normal CBC, ESR (2 mm/hr), CRP (<0.6 mg/L), Antinuclear antibody panel including ANA global, pANCA and cANCA were negative, only anti-PM/Scl was positive. Repetitive nerve stimulation test was normal (no decrement), visual evoked potentials normal, pseudocholinesterase 8.5 U/ml (normal). CSF analysis: 2 cells/μL, protein 75 mg/dL, acid fast bacilli for tuberculosis and herpes simplex virus PCR was negative. Orbital MRI demonstrated diffuse homogeneous enhancement and the thickening of the superior rectus muscle, which is likely representing inflammatory orbital pseudotumor; no intraconal mass or cavernous sinus involvement. Brain MRI was unremarkable. IgG4 levels were done to rule out IgG4 orbitopathy, which was normal. Intravenous methylprednisolone (1 g/day x 3 days) was administered, yielding dramatic improvement: ptosis resolved by day 5, headache abated. Oral prednisolone (1 mg/kg) was tapered and continued at discharge. At 2 weeks, child was asymptomatic. However, at 1 month follow up, child had a recurrence of symptoms on tapering dose of prednisolone (0.1 mg/kg). Hence the child was readmitted for

intravenous methylprednisolone which was continued for a total of 3 days and then switched over to oral prednisolone 1 mg/kg/day. Neurology review was taken and advised continuing the oral steroid and to consider immunosuppressants like azathioprine and mycophenolate mofetil. As the child's clinical condition improved, child was discharged on oral steroids.

Discussion

Isolated superior rectus myositis in adolescents is rare, often idiopathic orbital inflammatory syndrome (IOIS), but anti-PM/Scl positivity implicates systemic autoimmunity.[1,2] PM/Scl antigens (PM/Scl-75/100) are nucleolar, driving Th17-mediated muscle inflammation with perivascular CD8+ infiltrates and MHC-I upregulation, akin to polymyositis.[5] Orbital MRI findings (enlarged tendon-sparing muscle) distinguish from thyroid eye disease (diffuse involvement, sparing tendons) or infection (abscess, sinusitis).[10] Negative neuromuscular tests excluded myasthenia gravis (prevalence 1/10,000 adolescents) and botulism; normal cerebrospinal fluid ruled out Herpes simplex virus and /Tubercular orbitopathy.[3] Steroid response confirmed inflammatory etiology, with relapse risk 20-30% necessitating the consideration of immunosuppression.[6] Unlike adult PM/Scl (Interstitial lung disease 30%-50%, Raynaud's 40%), pediatric cases skew myopathic without scleroderma, possibly due to immature fibrosis pathways.[7] Differential includes IgG4 orbitopathy (biopsy-proven) and granulomatosis with polyangiitis, but negative ANCA/ serum IgG4 favored isolated myositis.[4] Prognosis excels with early therapy (90% remission), though chronicity warrants monitoring for systemic evolution. This case expands phenotype to unilateral pediatric orbital-predominant disease, urging myositis panels in steroid-responsive ptosis.[8]

Conclusion

Anti-PM/Scl-positive orbital myositis should be considered in steroid-responsive pediatric ptosis with superior rectus thickening, even sans systemic features. Serological diagnosis expedites targeted immunosuppression and averting morbidity.[1]

Compliance with Ethical Standards
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Conflict of Interest: None

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