Acquired reversible Brown syndrome caused by focal abscess of the superior oblique muscle

Julius T. Oatts, MD, MHS,[a]
Jefferson J. Doyle, MD, PhD,[b]
Mary-Magdalene U. Dodd, MD,[c]
Iason S. Mantagos, MD,[d] and Linda R. Dagi, MD[d]

A 16-year-old boy with a history of relapsed acute myeloid leukemia and a right lower lobe lung abscess confirmed to be Aspergillus presented for a baseline eye examination prior to consideration of bone marrow transplantation. He noted double vision in up-and-left gaze, and his examination was consistent with an acquired right-sided Brown syndrome. Magnetic resonance imaging revealed a 4 mm rim-enhancing inflammatory focus in the right superior oblique muscle. His Brown syndrome resolved after treatment with systemic antimicrobials.

Case Report

A 16-year-old boy presented at Boston Children's Hospital for a baseline eye examination prior to consideration of bone marrow transplantation (BMT). He had a history of acute myeloid leukemia, which had been diagnosed 8 months prior to presentation when he developed fatigue, jaundice, fever, and was found to be pancytopenic. He underwent induction chemotherapy with cytarabine, daunorubicin, and etoposide. Two months later, he was found to have relapsed disease and underwent reinduction with mitoxantrone and cytarabine. This was complicated by a polymicrobial necrotizing infection of his right hand, which grew *Aspergillus fumigatus*, Bacillus nonanthracis, *Pseudomonas boreopolis*, and Staphylococcal nonaureus, requiring wound debridement and prolonged antimicrobial therapy.

One month prior to his eye examination, he was admitted to the hospital for planned BMT but was found to have a right lower lobe abscess, which required a right lower lobectomy, the pathology showing numerous septate branching hyphae consistent with Aspergillus. He was treated with voriconazole. Days prior to his eye examination, he was noted to have new lung nodules, which were found to grow methicillin-sensitive *Staphylococcus aureus* (MSSA) on tissue culture following biopsy. He was also found to have a lesion on the mitral valve concerning for endocarditis. His blood cultures remained negative while he was on broad-spectrum antimicrobials, including vancomycin, micafungin, and voriconazole.

On presentation, he reported 2-3 days of diplopia in up-and-left gaze. His uncorrected visual acuity was 20/20 in each eye. Ductions showed a −2 deficit in elevation on adduction of the right eye but were otherwise full (Figure 1A). He was orthotropic in all positions of gaze with the exception of upgaze, with left hypertropia of 8^{Δ}, up-and-left gaze with left hypertropia of 10^{Δ}, left gaze, with left hypertropia of 10^{Δ}, and left head tilt, with left hypertropia of 4^{Δ}. Examination results were consistent with a right-sided Brown syndrome. The remainder of his examination, including dilated fundus examination, was normal.

The patient underwent magnetic resonance imaging (MRI) of the brain and orbits with and without contrast, which revealed a mildly expansile T2 hyperintensity within the right superior oblique muscle belly consistent with edema. Within the central portion of this edema, there was a 3 × 2 × 4 mm (anterior-posterior × transverse × sagittal) rim-enhancing focus, with intermediate T2 signal and low diffusivity and minimal extraconal fat stranding surrounding the right superior oblique muscle (Figure 2). This was presumed to be a focal abscess, given the patient's history of new pulmonary nodules and likely MSSA endocarditis.

He was started on oxacillin for the treatment of presumed disseminated MSSA infection with endocarditis and focal right superior oblique muscle abscess. Surgical drainage was considered, but, given the relatively small size and posterior location in the orbit, the decision was made to observe on medical treatment. Three weeks following his initial examination, his complaints of diplopia in up-and-left gaze had resolved, and his ductions and versions returned to normal (Figure 1B).

Discussion

Brown syndrome can be congenital or acquired and refers to the inability to elevate the eye in adduction. Acquired Brown syndrome has been reported in the setting of inflammatory disorders, such as rheumatoid arthritis,[1] systemic lupus erythematosus,[2] scleroderma,[3] and local injury or irritation to the muscle such as sinusitis, peribulbar anesthesia,[4] blunt orbital trauma,[5] or glaucoma valve implantation.[6] Our patient's strabismus examination was consistent with a right-sided Brown syndrome, but he had no known risk factors, which prompted MRI evaluation.

Patients with a history consistent with Brown syndrome with a known precipitating event are often followed without imaging, although MRI findings for Brown syndrome have been reported in the literature and are variable, ranging from no findings to enlarged superior oblique tendon or fat enhancement surrounding the trochlea.[7] In

Author affiliations: [a]Department of Ophthalmology, University of California, San Francisco, California; [b]Department of Ophthalmology, Wilmer Eye Institute, Johns Hopkins University, Baltimore, Maryland; [c]Department of Ophthalmology, Alberta Childrens Hospital, Calgary, Alberta, Canada; [d]Department of Ophthalmology, Boston Children's Hospital, Harvard Medical School, Boston, Massachusetts

Submitted June 22, 2018.
Revision accepted December 29, 2018.
Published online January 30, 2019.

Correspondence: Julius T. Oatts, 10 Koret Way, San Francisco, CA 94131 (email: julius.oatts@ucsf.edu).

J AAPOS 2019;23:172-174.

1091-8531/$36.00
https://doi.org/10.1016/j.jaapos.2018.12.003

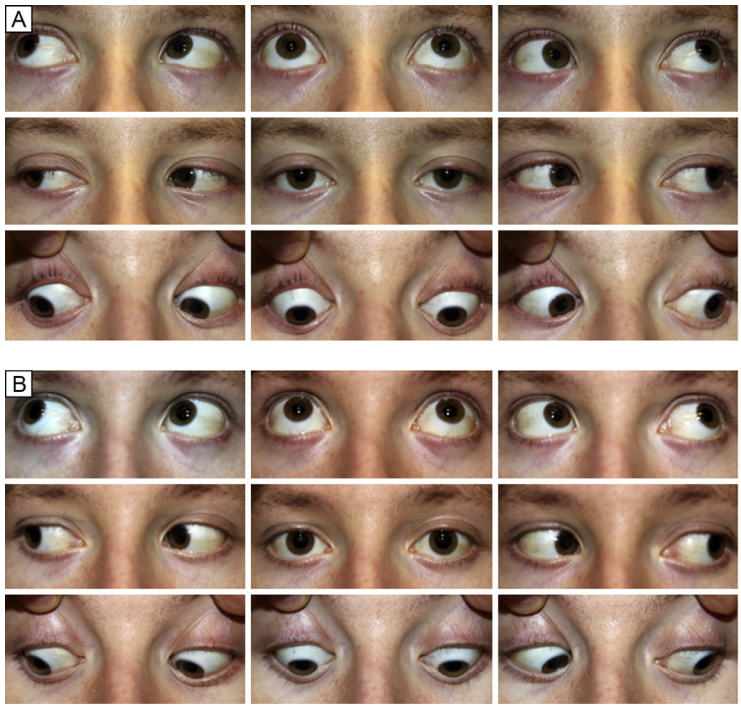

FIG 1. Nine positions of gaze on presentation (A), showing right eye elevation deficit in adduction consistent with a right-sided Brown syndrome, and 3 weeks after presentation and treatment with oxacillin (B), showing normalization of right eye elevation in adduction.

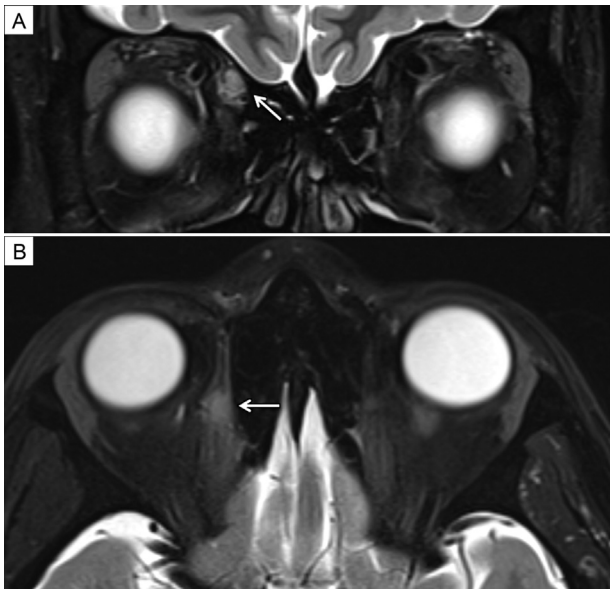

FIG 2. Coronal (A) and axial (B) T2-weighted fat-suppressed magnetic resonance images demonstrating mildly expansile T2 hyperintensity within the right superior oblique muscle belly consistent with edema (arrow). And a 3 × 2 × 4 mm (anterior-posterior × transverse × sagittal) rim-enhancing focus with intermediate T2 signal with minimal extraconal fat stranding.

our case, imaging was essential to investigate the etiology of our patient's Brown syndrome. The abscess caused focal muscle edema and enlargement, likely preventing it from sliding normally through the trochlea.

Treatment for Brown syndrome is often dictated by the underlying etiology. Surgical approaches include superior oblique tenotomy, tenectomy, or spacer with suture or silicone. Nonsurgical management includes local corticosteroid injection and systemic nonsteroidal anti-inflammatory medications or oral steroids. In this case, surgical exploration with possible incision and drainage was considered, but given the relatively small size and posterior location, the patient was observed on antimicrobial therapy, and his Brown syndrome resolved with appropriate systemic antibiotics.

To our knowledge, this is the first reported case of Brown syndrome due to a focal abscess of the superior oblique muscle. This case demonstrates the importance of imaging in cases of acquired Brown syndrome with no clear etiology as well as the efficacy of systemic antimicrobials in the treatment of a focal abscess of the superior oblique muscle.

Literature Search

PubMed was search on June 15, 2018, without date or language restrictions, using the following terms and combinations: *Brown syndrome*, *superior oblique muscle* AND *abscess*.

References

1. Wang FM, Wertenbaker C, Behrens MM, Jacobs JC. Acquired Brown's syndrome in children with juvenile rheumatoid arthritis. Ophthalmology 1984;91:23-6.
2. Alonso-Valdivielso JL1, Alvarez Lario B, Alegre López J, Sedano Tous MJ, Buitrago Gómez A. Acquired Brown's syndrome in a patient with systemic lupus erythematosus. Ann Rheum Dis 1993;52:63-4.
3. Barnett AJ, Griffiths JC, West RH. Acquired Brown's syndrome. Ann Rheum Dis 1993;52:835.
4. Erie JC. Acquired Brown's syndrome after peribulbar anesthesia. Am J Ophthalmol 1990;109:349-50.
5. Baker RS, Conklin JD Jr. Acquired Brown's syndrome from blunt orbital trauma. J Pediatr Ophthalmol Strabismus 1987;24:17-21.

6. Prata JA Jr, Minckler DS, Green RL. Pseudo-Brown's syndrome as a complication of glaucoma drainage implant surgery. Ophthalmic Surg 1993;24:608-11.
7. Wright KW. Brown's syndrome: diagnosis and management. Trans Am Ophthalmol Soc 1999;97:1023-109.

Increased restriction from an accessory lateral rectus in exotropic Duane syndrome

Luiza Macieira de Almeida Neves, MD,[a,b] and Ian Curi, MD[a]

Supernumerary extraocular muscles can cause restrictive strabismus, unusual ocular movements, and a persistent positive forced duction test. Even among patients with clinically typical strabismus, intraoperative testing and surgical exploration may reveal the presence of supernumerary extraocular muscles. We report the case of a patient with exotropic Duane syndrome found intraoperatively to have an accessory lateral rectus muscle, with histopathologically confirmed striated fibers.

Case Report

An otherwise healthy 11-year-old girl presented at the Hospital Federal dos Servidores do Estado, Rio de Janeiro, Brazil, with right head turn and abnormal left eye movements since birth. The family denied any previous ocular surgery. On ophthalmological examination, her visual acuity was 20/20 in each eye, and there was a 15° right head turn, in which stereopsis was 40 arcsec. Motility examination revealed −3 limitation of adduction and −3 limitation of abduction of the left eye as well as a great retraction of the bulbi in adduction and intense up- and downshoot (Figure 1). In forced primary position, she had a left exotropia of 30$^{\Delta}$. She was diagnosed with exotropic Duane syndrome in the left eye. Periosteal fixation of the lateral rectus muscle in the orbital wall was planned to try to eliminate or alleviate all of her anomalies—retraction, exotropia, anomalous head position, up- and downshoot—in a single surgery.

Intraoperatively there was no sign of a previous surgery, and forced duction testing still revealed a mild restriction to adduction of the left eye after lateral rectus disinsertion. We explored further and found an accessory muscle 4 mm behind to the original insertion of the lateral rectus muscle that continued far posterior to the globe (Figure 2). The muscle was extirpated and sent for histopathological analysis, which showed striated muscle fibers (Figure 3). After removal of the accessory muscle, forced duction testing was negative.

At postoperative month 3 (Figure 4) the patient was orthotropic, without anomalous head position or significant retraction, there was full adduction, no up- or downshoot, but the abduction remained very weak. At final follow-up, 1 year postoperatively, the patient remained orthotropic with no head turn or up- or downshoot.

Discussion

Supernumerary muscles can cause anomalous attachments between structures of the eye and the orbit. According to Duke-Elder,[1] anatomical abnormalities of extraocular muscles are relatively common. Khriti and Demer[2] found that 2.4% of the population had structures consistent with supernumerary extraocular muscles on magnetic resonance imaging. There are few reported cases of accessory muscles in Duane syndrome.[2-5] To our knowledge, this is the first report of an accessory lateral rectus muscle confirmed by histopathological examination in a patient with exotropic Duane syndrome.

Orbital imaging examination are indicated in patients with unusual ocular movements to investigate anomalous structures.[2,3,5] Because our patient had a typical presentation of exotropic Duane syndrome, we did not perform MRI before the surgery.

Intraoperative forced duction testing is particularly important, given the restrictive nature of anomalous structures.[2,3] Lueder[3] suggests that the persistence of restriction after disinsertion of a rectus muscle is usually a clue to the presence of an anomalous muscle. Accordingly, in our patient forced duction testing showed continued limitation after disinsertion of the the lateral rectus muscle, and thus we continued surgical exploration and discovered an anomalous structure adherent to the sclera. After its extraction, we could finally move the eye freely. The patient became orthotropic, with neither anomalous head position nor significant ocular retraction or up- or downshoot. Other authors have also described similar favorable outcomes.[3-8]

Lueder[3] classified anomalous orbital structures into three types. The first includes structures arising from the extraocular muscle and inserting in abnormal locations. The second includes abnormal fibrous bands located under the extraocular muscle. The third type arises in the posterior orbit and inserts on the globe or on the extraocular muscle. Pineles and Velez[4] presented a similar case of exotropic Duane with two accessory bands posterior to the lateral rectus muscle that resembled fibrous tissue with scattered muscle fibers as a combination of Lueder's first two types. The accessory muscle described in our case resembles Lueder's third type, because its insertion was

Author affiliations: [a]Hospital Federal dos Servidores do Estado do Rio de Janeiro, Rio de Janeiro, Brazil; [b]Instituto Nacional de Saúde da Mulher, da Criança e do Adolescente Fernandes Figueira – Fundação Oswaldo Cruz, Rio de Janeiro, Brazil

Submitted July 17, 2018.
Revision accepted January 9, 2019.
Published online February 5, 2019.

Correspondence: Ian Curi, MD, Sacadura Cabral Street, 178, Saúde, Rio de Janeiro, Brazil (email: ianbonotto@gmail.com).

J AAPOS 2019;23:174-176.

1091-8531/$36.00
https://doi.org/10.1016/j.jaapos.2019.01.003