Congenital Absence of Bilateral Inferior Rectus Muscles: A Case Report

Pei-Yu Lin, MD; and May-Yung Yen, MD

INTRODUCTION

Cases of congenital absence of the inferior rectus muscle are rare, and bilateral ones are even rarer. Such a case combined with multiple anomalies of extraocular muscles is discussed in this article.

Congenital absence of the inferior rectus muscle is quite rare. Cooper reported the eighth case, not associated with systemic diseases, in 1971, and five other isolated cases were subsequently reported. Congenital absence of bilateral inferior rectus muscles is even rarer and is often found in craniofacial dysplasia or combined with an absence of other extraocular muscles.

The first bilateral case was presented in 1908 by Stier, whose patient could not depress either eye below midline, and absence of the inferior rectus muscle was noted intraoperatively in one eye, however, the other eye was not explored surgically. Mets, in 1987, reported two cases of bilateral absence of the inferior rectus muscles found intraoperatively, but coronal CT scanning demonstrated the presence of bilateral inferior rectus muscles in the posterior orbit.

Adhesion between the lateral rectus and inferior oblique muscle has been reported and produces a picture of abducens palsy. A case is reported in this article of congenital absence of bilateral inferior rectus muscles combined with abnormal insertion of inferior oblique muscle to the lateral rectus muscle. Instead of abducens palsy, however, the lateral rectus muscle shows marked restriction.

CASE REPORT

A 44-year-old Asian woman came to our clinic with the complaint of poor vision in the left eye since childhood. Upon examination, her left eye deviated outward 45 prism dipters (D) and upward 16 prism dipters on primary gaze. This deviation had occurred for as long as she could remember. She denied having had any ocular examination or treatment, even glasses, in the past. Visual acuity was 20/30 in the right eye and CF/20 cm in the left eye. The vision of the right eye could be corrected to 20/20 with lens –3.0 D x 170°.

However, the vision of the left eye showed non-correcetable with lens –11.0 D –5.0 D x 10°. Duction testing revealed that the left eye could not adduct or infraduct to pass the vertical and horizontal midline, respectively. The right eye showed a limitation of infraduction on abduction (Fig 1). Forced duction test demonstrated that the lateral rectus muscle of the left eye was so tight that the eyeball would not move nasally passing the midline. The superior rectus muscle showed no restriction. The forced duction test of the right eye was normal.

In view of the unusual eye movement, CT scanning was ordered prior to any treatment. Surprisingly, an absence of bilateral inferior rectus muscles was found throughout the coronal section of the orbit (Fig 2). Next, recession of the superior rectus muscle 6 mm and disinsertion of the lateral rectus muscle were performed in the left eye under subconjunctival injection of 2% xylocaine and intraoperative adjustment.

More muscular abnormalities were noted while performing the operation. First, the lateral rectus muscle showed marked fibrosis, and the inferior oblique muscle was attached to the muscle sheath of lateral rectus muscle instead of sclera (Fig 3). The medial rectus muscle became very thin and lax, and could be easily pulled out with a muscle hook (Fig 4). Next, a 360° peritomy was performed, but no tissue similar to the inferior rectus muscle could be traced. Instead, a prominent anterior ciliary artery was located directly on the sclera where the inferior rectus muscle should have been found (Fig 5). The patient exhibited 12 prism dipters of exotropia and no vertical deviation on primary gaze one month postoperatively, but depression of the left eyeball was still limited.

DISCUSSION

Surgical procedures including tenotomy or recession of the superior rectus muscle, myectomy of the inferior oblique muscle, Knapp's procedure, and tenotomy of the contralateral inferior rectus muscles have all been reported to correct congenital absence of the inferior rectus muscle. Horizontal muscle transplantation was not performed in this case because the lateral rectus muscle was so tight it had to be disinserted to correct the exotropia. Furthermore, the medial rectus muscle was so weak we believe it would not function if transplanted inferiorly. We were also afraid of anterior ischemic syndrome occurring if we operated on three rectus muscles at the same time, especially when the only
unoperated rectus muscle was absent.

Congenital absence of the inferior rectus muscle had been reported to combine with an absence of other extraocular muscles, including the superior rectus muscle, lateral rectus muscle, inferior oblique muscle, superior rectus plus superior oblique muscles, and superior rectus plus two oblique muscles. Sevel in 1981, after reappraisal of 54 specimens, concluded that the extraocular muscles developed from a superior and inferior mesodermal complex. The superior oblique, superior rectus, levator palpebrae superioris muscles, and upper part of two horizontal rectus muscles were developed from the superior mesodermal complex. The inferior rectus, inferior oblique, and lower part of two horizontal rectus muscles were developed from the inferior mesodermal complex.

Our patient displayed multiple abnormalities of extraocular muscles, including an absence of the inferior rectus muscle, malinsertion of the inferior oblique muscle to the lateral rectus muscle, fibrosis and restriction of the lateral rectus muscle, and extreme weakness of the medial rectus muscle, which, according to Sevel’s theory, represent inferior mesodermal complex maldevelopment.

**SUMMARY**

A case has been presented in this article of congenital absence of bilateral inferior rectus muscles combined with restriction of the lateral rectus muscle and malinsertion of the inferior oblique muscle to the lateral rectus muscle. The surgical procedures for correction of the absence of the inferior rectus muscle and embryogenesis of extraocular muscles were also reviewed.

**REFERENCES**

Fig 3: (A) The inferior oblique muscle (arrow) inserts directly to the muscle sheath of lateral rectus muscle. (B) The inferior oblique muscle (arrow) also recedes while performing recession of the lateral rectus muscle. (C) The insertion site of the lateral rectus muscle becomes a sheet of scar tissue.

Fig 4: The medial rectus muscle becomes very thin and lax and could be retracted easily with a muscle hook.

Fig 5: A prominent anterior ciliary artery located on the area where the inferior rectus muscle should normally be inserted.