CASE REPORT

Bilateral medial rectus aplasia and a modified surgical approach of transposition myopexy of vertical recti

Pradeep Sharma, Shweta Chaurasia, Abhijit Rasal

Department of Ophthalmology,
Dr Rajendra Prasad Centre for
Ophthalmic Sciences, All India
Institute of Medical Sciences,
New Delhi, India

Correspondence to

Dr Shweta Chaurasia,
shweta84omns@yahoo.com

Accepted 30 July 2017

SUMMARY

A 16-year-old girl presented with left eye large-angle exotropia. On examination, we found bilateral limitation of adduction. CT orbit showed hypoplastic medial rectus bilaterally, but intraoperatively we found absent medial recti on both sides. This case report explains discrepancy between the imaging and the intraoperative findings and discusses the management dilemma in view of the risk of anterior segment ischaemia and how marked exodeviation and adduction limitation was tackled by the new technique of transposition myopexy, a modification of the procedure described by Nishida along with recession of lateral rectus to achieve good alignment. This procedure changes the vector forces of the vertical rectus without splitting or tenotomy of the muscles.

BACKGROUND

Medial rectus (MR) aplasia, a rare entity,^{1–3} has been identified as subtype of congenital cranial dysinnervation disorder¹ that affects the normal development of brainstem motor neurons. There is a separate global and orbital development in any muscle,^{3,4} which may present as an intraoperative surprise. However, bilateral absence of MR is rare and is indeed a challenge for the strabismologist to manage such large deviations in view of compromised blood supply. Technically, supramaximal recession⁵ of lateral rectus (LR) will not suffice such large exodeviation and will also limit abduction. Full muscle width vertical recti transposition if done will cause anterior segment ischaemia (ASI) in this case, whereas partial vertical recti muscle transposition (PVRT) may not correct such large-angle squint³ but will accompany increased inflammation due to tenotomy and splitting of fibres and may also risk ASI. We therefore present a rare case of bilateral MR aplasia—a diagnostic and surgical challenge tackled successfully with a modified surgical technique of transposition myopexy.

CASE PRESENTATION

A 16-year-old female patient presented with complaints of outward deviation of left eye since birth associated with diminution of vision. There was a history of limitation of inward movement of each eye. There was no prior history of usage of glasses or occlusion therapy. On examination, her best corrected visual acuity (BCVA) was OD 20/20 and OS 20/120 on ETDRS chart with a refractive error of -2.75 Dioptre cylinder at 90° in the left eye. All

examinations, investigations and surgical intervention were done after her written informed consent. On Hirschberg's test, a left divergent squint $\sim 60^\circ$ was noted (figure 1A) and on prism bar cover test, the deviation measured $>90^\Delta$ BI (120^Δ BI). Bagolini striated glasses test revealed left eye suppression. On motility examination (both ductions and versions), a -2 adduction limitation was noted in both eyes (figure 1A) without any abduction nystagmus, and there was absence of convergence. Force duction test for left eye LR was free, but right eye LR was mildly tight. However, on active force generation test, no tug was felt for MR in both eyes.

INVESTIGATION

Orbital CT revealed mildly hypoplastic MR (figure 2A). Morphometric analysis of the coronal section was done by measuring the diameter of muscle at the posterior aspect of the globe, which showed thinner (1.92 mm) MR compared with the normal population (3.6 mm), and was also thinner than ipsilateral LR (2.3 mm) (figure 2C and D). A provisional diagnosis of both eye MR hypoplasia with anisometropic amblyopia was made.

DIFFERENTIAL DIAGNOSIS

MR hypoplasia/aplasia.

SURGICAL TREATMENT

Intraoperatively, we found an empty muscle sheath with absent MR bilaterally (figure 2B). We planned bilateral transposition myopexy on the vertical recti (a modification of the procedure described by Nishida⁶). The major modification from the original Nishida's procedure was that we passed 5–0 non-absorbable braided polyester suture (Ethibond) 8 mm from muscle insertion in both superior rectus (SR) and inferior rectus (IR) full muscle width (against one-third muscle width done in original Nishida's technique)⁷ similar to recession suture and transposed it just at the site of absent MR border, 14 mm behind limbus without tenotomy or muscle splitting (figure 3A–E). Also, in the right eye, 8.5 mm LR recession was done as its force duction test was mildly tight.

OUTCOME AND FOLLOW-UP

Postoperatively, alignment was achieved with improvement of adduction by 25° measured on synoptophore (figure 1B). Postoperative iris

CrossMark

To cite: Sharma P, Chaurasia S, Rasal A. *BMJ Case Rep* Published Online First: [please include Day Month Year]. doi:10.1136/bcr-2017-220404

Figure 1 (A) Preoperative picture showing large-angle exotropia with bilateral adduction deficit. (B) Postoperative picture showing alignment with improved adduction in both eyes.

fluorescein angiography done immediately and 3 months after surgery did not reveal any sign of ASI. Bilateral transposition myopexy reduced the exotropia from 120Δ BI to 30Δ BI. For the residual exotropia, we performed OD LR recession 8.5 mm. Finally, the patient was aligned with residual exotropia of 10Δ BI and bilateral improvement of adduction by 25°. At 1-year follow-up, patient maintained same BCVA and was well aligned (residual 10 pd BI) with good adduction.

DISCUSSION

MR aplasia is a rare entity, and there are a few reports of absent recti.^{1–3} Although CT scan revealed mild hypoplasia of MR, but to our surprise, intraoperatively, an empty muscle sheath was found bilaterally. This discrepancy between the imaging and clinical finding can be explained by reviewing the muscle embryonic development. Development of extraocular muscles (EOMs) requires early signals from periorbital neural crest and the developing eye, which subsequently provide early signals to cranial mesoderm that are required to drive the morphogenic process of EOM development. There is a separate development of global and orbital fibres of muscles embryologically.³ The global layer extends the full muscle length, inserting into the sclera via a well-defined tendon, while the orbital layer ends before muscle becomes tendinous.⁴ Recent studies have shown that this early termination of the orbital layer is a consequence of its insertion

Figure 2 (A) CT orbit showing presence of medial rectus (MR) bilaterally with mild hypoplasia. (B) Intraoperatively MR found absent (seen from medial side after conjunctival peritomy with eye in abducted position). (C) Intraoperative photograph showing normal eye with MR present at 5.5 mm from limbus (shown with arrow) seen from same position. (D) CT scan showing only orbital layer which abruptly ends at equator and absent global layer. (E) Comparison with normal CT scan showing muscle going up to sclera.

Figure 3 (A and D) 5–0 Non-absorbable suture (Ethibond) shown in green colour being passed 8 mm from muscle insertion of superior rectus both medial and lateral sides. (B) Both ends of Ethibond sutured to sclera 14 mm behind limbus on medial side (at absent upper MR border). (C and E) Intraoperative photographs showing both vertical recti sutured on medial side.

into the muscle pulley, at approximately the equator of the globe.⁸ The active pulley system uses orbital layer motor units to adjust EOM vector forces in different gaze positions making commands independent of initial eye position.⁸ On reviewing axial section scans, both the MR were found to be abruptly ending at the equator. This explains the discrepancy between CT and intraoperative findings that the global layer in our case was absent and only the orbital layer was present, which was seen ending abruptly at the equator, and therefore, intraoperatively no muscle was seen.

The other dilemma was management of this case as we were left with only one muscle in each eye to operate to avoid the risk of ASI, and this would also not correct such a large exotropia of >90 pd BI. In an earlier report of MR aplasia, partial rectus transposition had been used, but only partial success was achieved with residual exotropia in primary position.³ Also, we were hesitant for PVRT because after PVRT, if there is residual exotropia and we plan to operate on LR, there will be a definite risk of ASI in the absence of MR as lateral side of anterior segment is dependent on anterior ciliary artery from LR, SR and IR with minor contribution from long posterior ciliary artery. Half muscle transposition on sclera without tenotomy⁷ had the problem of splitting muscle fibres and thereby inciting inflammation and causing operative damage. So, there was a need for transposition procedure but without tenotomy and splitting of muscle fibres and avoiding ASI. Nishida described a procedure of transferring the vectors in sixth nerve palsy by transposition of vertical recti without tenotomy of muscle.⁶ Non-absorbable suture was passed through each temporal margin of vertical recti at approximately one-third width from the edge at a distance of 8–10 mm behind the insertion. The same suture also was inserted through sclera at a distance of 10–12 mm behind the limbus at the superotemporal or inferotemporal quadrant. Then, the lateral margin of each vertical rectus muscle was transposed superotemporally or inferotemporally and sutured to the sclera. The amount of esotropia corrected using this procedure alone ranged from 24 to 36 PD.

However, in our case, owing to such large exotropia, we wanted greater correction, which one-third muscle transposition would not suffice. So we chose to transpose the vertical recti full muscle width (unlike one-third as in original Nishida) on medial side without tenotomy, modifying the original Nishida's

procedure. Avoidance of muscle splitting thereby avoided associated inflammation. In this technique, first we passed full muscle width 5–0 Ethibond 8 mm behind both vertical recti. We measured 14 mm on medial side on imagined MR upper and lower border and took scleral bites using double arm Ethibond on both lateral and medial sides, thereby transposing both the muscles on nasal side. Bilateral transposition myopexy reduced the amount of exotropia from 120Δ BI (60° by Hirschberg) to 25–30Δ BI and for the residual exotropia 8.5 mm LR recession in the right eye was performed on the subsequent day as it was mildly tight. Finally, the patient was aligned with residual exotropia of 10Δ BI and bilateral improvement of adduction by 25°.

In conclusion, our modified technique of transposition myopexy of the vertical recti successfully corrected large-angle

exotropia and also improved adduction without limiting abduction (as in supramaximal recession) or compromising anterior segment blood supply in absence of MR.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

© BMJ Publishing Group Ltd (unless otherwise stated in the text of the article) 2017. All rights reserved. No commercial use is permitted unless otherwise expressly granted.

REFERENCES

- 1 Houtman AC, Hocking G, Lloyd C, *et al.* Large-angle congenital exotropia due to 'absent' medial recti: a case of congenital cranial dysinnervation disorder. *Eye* 2009;23:1489–90.
- 2 Girard LJ, Neely RA. Agenesis of the medial rectus muscle; correction of a case by transplantation of slips from the vertical recti. *AMA Arch Ophthalmol* 1958;59:337–41.
- 3 Singh A, Sharma A, Sharma P. Congenital bilateral aplasia of medial recti in a family. *JAMA Ophthalmol* 2013;131:798–800.
- 4 Spencer RF, Porter JD. Biological organization of the extraocular muscles. *Prog Brain Res* 2006;151:43–80.
- 5 Chang JH, Kim HD, Lee JB, *et al.* Supermaximal recession and resection in large-angle sensory exotropia. *Korean J Ophthalmol* 2011;25:139–41.
- 6 Muraki S, Nishida Y, Ohji M. Surgical results of a muscle transposition procedure for abducens palsy without tenotomy and muscle splitting. *Am J Ophthalmol* 2013;156:819–24.
- 7 Nishida Y, Inatomi A, Aoki Y, *et al.* A muscle transposition procedure for abducens palsy, in which the halves of the vertical rectus muscle bellies are sutured onto the sclera. *Jpn J Ophthalmol* 2003;47:281–6.
- 8 Demer JL, Miller JM, Poukens V, *et al.* Evidence for fibromuscular pulleys of the recti extraocular muscles. *Invest Ophthalmol Vis Sci* 1995;36:1125–36.

Learning points

- ▶ Such rare congenital cranial dysinnervation disorder cases must be preoperatively suspected, imaged, appropriately planned and surgically staged.
- ▶ We transposed full muscle width without splitting or tenotomy (modification of Nishida's procedure).
- ▶ So this technique promises safety along with greater effectivity by generating greater vector forces with greater adduction range.

Copyright 2017 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow