Congenital sixth nerve palsy with associated anomalies

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Congenital abduction deficit is most likely due to Duane's retraction syndrome as congenital abducens nerve palsy is very rare. We report two cases of infantile abduction deficit due to sixth nerve palsy associated with other anomalies to highlight the importance of including neuroimaging in the evaluation of an infant presenting with a limitation of abduction.

Key words: Congenital abducens palsy, Duane's syndrome, anomalies

Congenital sixth nerve palsy is a rare condition that can be transient and usually resolves within a few weeks. It is commonly due to a relative delay in the myelination of the sixth nerve when compared to the third nerve or as a result of perinatal cranial trauma.[1] Some cases with spontaneous resolution occurring within a few weeks with occasional relapses may also result from a post viral syndrome.[2] Congenital sixth nerve palsy may simulate Duane's retraction syndrome (DRS), a common cause for congenital abduction deficit. A careful clinical evaluation is essential to differentiate the two.

Case Report

Two infants with abduction deficit presented to our strabismus clinic. The first patient, a 6-month-old male infant, was brought to our clinic with a history of abnormal head posture noticed since 3 months of age. Clinical examination revealed a large right face turn [Fig. 1a] with a relatively small 20 prism diopter esotropia and abduction deficit of −4 in the right eye (graded on a 9-point scale of −4 through to +4) detected by the doll's eye maneuver. Gadolinium-enhanced brain magnetic resonance imaging (MRI) showed a 3.4 cm × 2.0 cm encephalomeningocele involving the right lower pons through the unfused petrosquamous suture of right temporal bone, with the neural tissue tethered to an intracranial dermoid [Fig. 1b]. The abducent nerve was intact. Corresponding high-resolution computerized tomogram of the temporal bone showed right inner ear malformation with cochlear and vestibular aplasia and poorly formed semicircular ducts [Fig. 1c]. The patient underwent right medial rectus recession, after which the face turn improved.

The second patient, a 4-month-old female infant, was referred for esotropia since birth. She had left 30 prism diopter esotropia in the primary position and a marked abduction deficit of −4 in the left eye revealed by doll's eye maneuver [Fig. 2a]. Brain stem evoked potentials were diminished in the right ear. She also had a left foot deformity since birth. She underwent botulinum toxin 2.5 IU injection to the left medial rectus under general anesthesia. Postinjection, her esotropia in primary gaze improved but abduction deficit persisted. Contrast-enhanced MRI of the brain showed anterior expansion of the subarachnoid spaces in the temporal poles and anterior to the frontal lobes. The abducent nerve was intact, and there was hypoplasia of the right cochlea with malformed vestibule and lateral semicircular canal [Fig. 2b]. Neither patient showed any upshoots, downshoots, or globe retraction on attempted adduction.

Discussion

Infants with a limitation of abduction are frequently considered to have DRS, which also includes other diagnostic signs such as palpebral fissure narrowing, upshoots, or downshoots on adduction. Sometimes, it is difficult to distinguish an esotropic Duane sine retraction from congenital sixth nerve palsy in

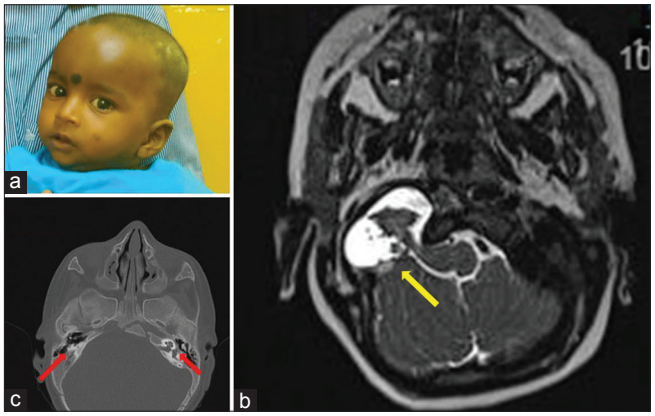

Figure 1: (a) Clinical photograph of the infant showing a large right face turn. (b) Axial T2 scan (magnetic resonance imaging) showing an encephalomeningocele involving the right lower pons with the neural tissue tethered to an intracranial dermoid (yellow arrow). (c) High resolution computerized tomogram of the temporal bone showing a cochlear and vestibular aplasia with poorly formed semi-circular ducts in the right inner ear when compared to the left (red arrow)

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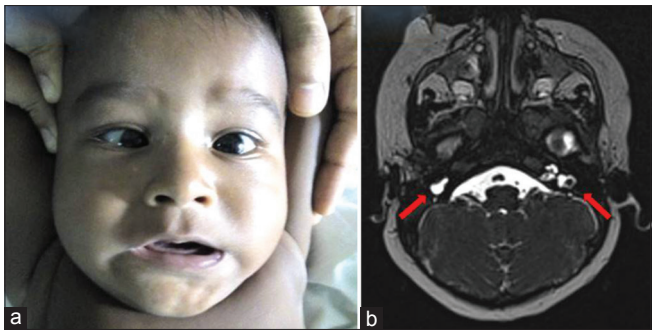

Figure 2: (a) Clinical photograph of the infant showing – 4 abduction deficit in the left eye. (b) Axial T1 image of magnetic resonance imaging brain showing hypoplasia of the right cochlea with malformed vestibule and lateral semicircular canal when compared to the left (red arrow)

an infant with no other diagnostic signs. It is important to differentiate the two because the evaluation and surgical management of these conditions are different. A confirmed case of DRS does not require any further elaborate investigations and has a good prognosis. In DRS, the primary position esotropia is relatively small compared to the lateral rectus underaction and a small head turn results in alignment, when compared to sixth nerve palsy.[3,4] Small recession of the medial rectus muscle is more effective in treating DRS which is unlikely in a case of abducens nerve palsy. Lateral rectus resection or vertical recti transposition procedure may worsen the upshoots and the eyeball retraction in DRS but are effective in the management of sixth nerve palsy.[5,6] The sixth cranial nerve on the affected side is absent in Type 1 DRS and a few Type 3 patients. This can be verified with high-resolution MRI which also shows sparing of lateral rectus muscle along with aberrant innervation by an extra branch of the oculomotor nerve. In true congenital abducens nerve palsy, it reveals a profoundly atrophic lateral rectus and present abducens nerve.[7-9] In our cases, we have found intact abducens nerves with associated cranial nervous system anomalies which have not been previously reported. Both infants also had malformed auditory ossicles which can lead to impaired speech and language development. Therefore, MRI resolves the clinical ambiguity between DRS with severe abduction deficit and abducens palsy and also helps with the classification of suspected cases of DRS. Furthermore, if neuroimaging in a patient with abduction deficit shows intact abducens nerves, further study to exclude associated anomalies, mass lesions, and hearing assessment should be performed.

Conclusion

Sixth nerve palsy should be carefully looked for in children with congenital abduction deficit. Cases with clinically diagnosed sixth nerve palsy need to be investigated with neuroimaging to exclude associated anomalies.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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