High Resolution Magnetic Resonance Image of Duane’s retraction syndrome with 3D Fast Imaging Employing Steady-state Acquisition (FIESTA) sequence: three case reports

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Three children of unilateral type I Duane’s retraction syndrome (DRS), aged 8 years, 14 months and 2 years were confirmed by demonstration of absence of ipsilateral abducens nerves on 3D fast imaging employing steady-state acquisition (FIESTA) MR sequence. The two younger patients had isolated abduction deficit without the characteristic globe retraction or upward/downward shooting on attempted adduction. The application of high-resolution MR imaging is useful for differential diagnosis and confirmation of DRS, especially in patients without characteristic presentations.

Duane’s retraction syndrome (DRS) is a rare congenital ocular motility disorder characterized by limited abduction and/or limited adduction. Other diagnostic manifestations include palpebral fissure narrowing, globe retraction and upward/downward shooting on attempted adduction. The most common cause is absence or hypoplasia of abducens cranial nerves and abducens nucleus ipsilateral to the affected eyes. Aberrant innervation of lateral rectus muscle by a branch of oculomotor nerve causes globe retraction when attempted abduction. DRS is classified clinically by Heuber [1] into three types: type I comprised the majority of patients (80%) with limited ability of abduction; type II (7%) with limited ability of adduction and type III (15%) with limited ability of abduction and adduction. Diagnosis of DRS in a typical case is not difficult. However, in young children, correct diagnosis of DRS is more challenging because characteristic globe retraction and up-/downward shooting are not evident [2]. In this situation, image studies by demonstration of absence of abducens nerves on MRI may help for the correct diagnosis of DRS from other diseases such as abducens nerve palsy, Möbius’ syndrome, congenital oculomotor apraxia, and congenital or infantile esotropia. Herein, we presented 3 young children of type I DRS, two of them had only isolated abduction deficit clinically, and all three patients had clear demonstration of absence of ipsilateral abducens nerve by 3D FIESTA sequences.

CASE REPORTS

Between August 2004 and November 2005, three children were referred for congenital abduction deficit. The first patient was an 8-year-old boy with orthotropia in primary gaze, and mild limitation of
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abduction of the right eye. Characteristic feature of upshoot of the affected eye on adduction was noted. He was referred for MRI study from ophthalmologist under the impression of Duane's retraction syndrome. The second patient (Fig. 1), a 14-month-old boy, and the third patient, a 2-year-old girl, had only abduction deficit of right eye and left eye respectively. Both of them were esotropia in primary gaze and were referred from neurosurgeons. None of these three patients had globe retraction or palpebral fissure narrowing on adduction, which is a typical finding for DRS. The MR studies were performed on a 1.5-T clinical scanner using an 8-channel phased-array head coil (Signa Excite; General Electric Medical Systems, Milwaukee, W1). In addition to routine spin echo T1-weighted images (T1WIs), fast spin echo T2-weighted images (T2WIs), fluid attenuation inversion recovery (FLAIR), thin-section 3D Fast Imaging Employing Steady-state Acquisition (FIESTA) sequence at brain stem level was performed using the following parameters: repetition time/echo time = 4.9/1.4 ms, flip angle: 65°, acquisition number: 2, FOV: 240 × 240, matrix size: 448x256, image thickness: 0.7 mm (after interpolation from 1.4mm scanning thickness), scan time: 4 minutes. There is no gross intracranial or intraorbital abnormality identified on routine T1WIs, T2WIs and FLAIR in all three patients. On the 3D FIESTA images, cisternal portion of the abducens nerve on affected side could not be observed in all three patients while the abducens nerves on unaffected side were well demonstrated. The diagnosis of Duane’s retraction syndrome was established for these three patients.

DISCUSSION

The abducens nerve fibers originate from a nucleus in the floor of the fourth ventricle, then emerges from the medullo-pontine sulcus on the anterior aspect of the brainstem. It runs upwards, forwards and laterally in the pontine cistern and then pierces the dura mater inferolaterally to the dorsum sellae at the opening of Dorello’s canal. Sometimes, CSF evagination into Dorello’s canals occurs with T2 hyperintensity at the opening. Although the abducent nerve passes a long distance from the brain stem to the lateral rectus muscle, it is difficult to identify this nerve reliably and consistently by using conventional radiologic techniques. In the initial two reports addressing the MR findings of DRS [3, 4], high-resolution thin-section T1-weighted images were used to observe presence or absence of abducent nerves in the patients with DRS. Low contrast between abducent nerve and CSF on T1-weighted images leads to inconsistent demonstration of abducent nerves on normal subjects [4], rendering the imaging studies of DRS less reliable. The later developed 3D T2-weighted images using either FSE or fast asymmetrical spin-echo (FASE) had conspicuously improved contrast between cranial nerves and CSF. Detection rate of cisternal portion of abducent nerves in normal subjects could be raised to 94.7% (540/570) [5] ~ 97.9% (47/48) [6]. However, imaging degradation by phase ghosting and CSF pulsation artifacts is common in both sequences.

3D FIESTA is the generic name by GE for the 3D Steady State Free Precession (SSFP) pulse sequence. Similar pulse sequence will be 3D true FISP by Siemens and by 3D-bFFE by Philips, and 3D b-TFE by Toshiba. The 3D SSFP is a gradient echo pulse with extremely short TR and completely balanced gradients in all three dimensions. The resultant images are T2 over T1 weighted with fat, CSF, and blood appearing bright. The images are characterized by short acquisition time, high contrast to noise ratio, high tissue-fluid contrast and almost
completely devoid of flow artifact [7], ideal for MR cisternography demonstrating the cranial nerves [8] including abducens nerves. The abducens nerve could be identified reliably and consistently by 3D SSFP up to 98%–100% in normal subjects [8, 9]. On the other hand, the differentiation between DRS and isolated abducens palsy is more challenging for patients with isolated abduction deficit and esotropia in primary position. Surgical treatment options for these conditions are different and misdiagnosis could have serious consequences. For patients of type I DRS with unacceptable head turn, significant misalignment, severe retraction and anomalous vertical movement, recession of medial rectus is the mainstay of surgical treatment [10]. On the contrary, resection and transposition of lateral rectus is the treatment for 6th nerve palsy but the contraindication for DRS because it increases retraction and does not improve abduction. With the advantage of high detection rate of abducens nerve by 3D FIESTA, non-visualization of the 6th nerve in patients with DRS implies congenital aplasia or even severely hypoplasia without normal innervation of ipsilateral lateral rectus muscle and confirms the diagnosis of Duane’s retraction syndrome.

Abducens nerve aplasia was documented by 3D FIESTA in our series of three patients with DRS of Huber’s type I, concordant with that of previous report by Kim using the same pulse sequence [11], in which absence of abducens nerve on the affected side is demonstrated in all 16 patients of type I DRS. In the same report, three of five patients of type III DRS also had aplasia of abducens nerve while none of two type II DRS had aplasia of the nerve. A most recent report by Denis [12] demonstrated abducens nerve aplasia in both of DRS type I and type II (one patient of each category). Identification of abducens nerve by MRI in type II and type III Duane’s syndrome and absence of the nerve in type I imply heterogeneity of the pathogenesis of ocular motility disorder diagnosed as DRS clinically. Further MRI study with 3D SSFP of larger cohort of patients should help to clarify the diagnosis and classification.

In summary, we presented three cases of unilateral DRS type I with aplasia of ipsilateral abducens nerve demonstrated exclusively by 3D FIESTA. In uncertain cases without characteristic presentation of Duane’s retraction syndrome, 3D FIESTA is a robust tool for diagnosis and differentiation.

**REFERENCE**

10. Kubota N, Takahashi H, Hayashi T, Sakaue T, Maruo T. Outcome of surgery in 124 cases of Duane's Retraction Syndrome (DRS) treated by intraoperatively graduated recession of the medial rectus for esotropic DRS, and of the lateral rectus for exotropic DRS. Binocul Vis Strabismus Q 2001; 16: 15-22
本文利用 3D FIESTA 清楚確定三位單側 Duane’s retraction syndrome（DRS）的兒童（年紀分別為 8 歲、14 個月、2 歲）缺乏同側的外旋神經。其中年紀較小的二位兒童只有單純的外旋神經麻痺而沒有典型眼球內縮的表現。利用 3D FIESTA 高解析度影像對於 DRS 的診斷確立及鑑別診斷很有幫助，特別是針對沒有典型表現的病患。