Non-paralytic pontine exotropia

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A 61-year-old man with a 5-year history of diabetes mellitus presented to the outpatient department with a 1-day history of dysarthria, diplopia, squint, and swaying during walking. Examination showed normal vital parameters. Nervous system examination showed primary position exotropia in the right eye (Fig. 1a), impaired adduction in the left eye with abduction nystagmus in the right eye (left internuclear ophthalmoplegia [INO]) (Fig. 1b). His leftward gaze was normal (Fig. 1c). There was no skew deviation. Convergence was preserved. His pupils were equal and reactive. Other cranial nerves were normal. His motor power in both upper and lower limbs was normal. Deep tendon reflexes were normal. Plantar reflexes were flexor. His sensory system including vibration and joint position sense was normal. He had scanning dysarthria and gait ataxia. His computerized tomography brain was normal. Magnetic resonance imaging of the brain showed acute infarct in the left ponto-mesencephalic junction (Fig. 2). Magnetic resonance angiography of the brain was normal. He was treated with antiplatelets and statin. His squint and diplopia improved and has residual dysarthria on follow-up at 2 weeks.

INO is a disorder of eye movements, characterized by adduction impairment combined with contralateral dissociated abduction nystagmus. Adduction may be preserved during convergence and skew deviation with hypertropia on the side of the lesion is often present. INO is caused by a lesion involving the medial longitudinal fasciculus (MLF) at the brainstem that connects with ocular nuclei. The etiology of INO includes multiple sclerosis, stroke, tumor, infection, hydrocephalus, trauma, nutritional, or metabolic disorders [1]. Involvement of neighboring structures like paramedian pontine reticular formation (PPRF), facial nerve nucleus, or fascicle can result in various one-and-a-half-plus disorders [2]. INO can be associated with exotropia of the ipsilateral eye (wall-eyed monocular INO-WEMINO) [3] or exotropia of the contralateral eye (non-paralytic pontine exotropia [PPE]) [4]. When there is involvement of MLF with PPRF, there is ipsilateral gaze palsy and INO. This combination is called one-and-a-half syndrome. This is often associated with contralateral exotropia [5]. Exotropia in this setting is called PPE.

Patients with classic INO have parallel visual axes in primary gaze. The association of INO with exotropia in contralateral eye was first described and termed as non-paralytic pontine exotropia by Bogousslavsky and Regli in 1983. NPPE is considered by most authors as a milder form of paralytic pontine exotropia (PPE), caused by a partial PPRF lesion[6,7]. In one series of patients with INO, NPPE was transiently noted in 32% and is often overlooked [8].

In patients with INO with limb or truncal ataxia, the responsible lesions were mostly restricted to the paramedian tegmentum at the ponto-mesencephalic junction. In contrast, the lesions causing isolated INO without ataxia were mostly located in the caudal or mid-pontine area. The responsible lesion was located exclusively in the pontine tegmentum affecting unilateral MLF. The ocular movement dysfunction had resolved or improved within <1 month.

Figure 1: (a) right exotropia in primary position; (b) normal gaze toward the left; (c) impaired adduction left eye associated with the right eye abduction nystagmus (left internuclear ophthalmoplegia)

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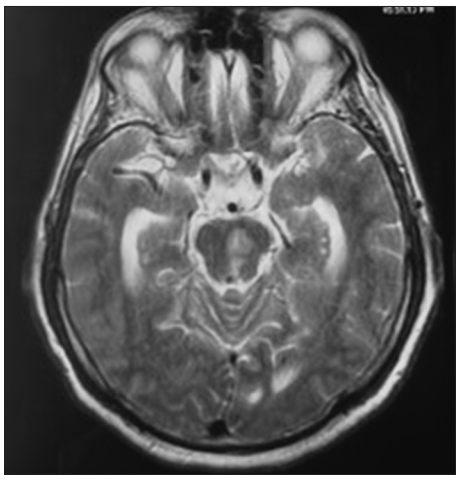

Figure 2: Left paramedian pontine infarct in axial T2-weighted magnetic resonance imaging brain

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