Early Transneuronal Degeneration in Dyke-Davidoff-Masson Syndrome

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Dear Editor,

We report herein a case of a 14-year-old boy who presented to us with a history of right hemiparesis, right focal seizures with secondary generalization, and mental retardation after an episode of meningoencephalitis along with pneumonia at 18 months of age. He had a normal birth history with normal developmental milestones until that time. Examination revealed hemiatrophy and spastic hemiparesis on the right side. EEG revealed asymmetrical slowing with occasional sharp waves arising from the left hemisphere. Brain MRI revealed left cerebral hemiatrophy, dilated sulci with left calvarial thickening (Fig. 1A), hyperaeration of the left frontal sinus (Fig. 1B), and focal encephalomalacia and gliosis in the left parietal lobe. Based on the classical imaging findings, a diagnosis of Dyke-Davidoff-Masson syndrome (DDMS) was reached. The cerebral hemiatrophy was also associated with atrophy of the ipsilateral cerebral peduncle (Fig. 1C) and contralateral cerebellar hemisphere (Fig. 1D). The patient's seizures responded to a combination of carbamazepine (600 mg) and levetiracetam (1 g).

DDMS, which was first described by Dyke et al.¹ in 1933, is characterized by cerebral hemiatrophy, compensatory calvarial changes such as hypertrophy of the skull and sinuses, and elevation of the sphenoid wing and petrous ridge, in association with contralateral hemiplegia, seizures, and mental retardation. The cerebral injury may occur either in utero or early in life, thus being classified into congenital and acquired types, respectively. The compensatory skull changes reflect adaptations to the unilateral reduction in brain substance, and occur when the problem arises either in utero or before 3 years of age.

The peculiar associations in this patient were atrophy of the ipsilateral cerebral peduncle and contralateral cerebellar hemisphere. Although crossed cerebrotocerebellar atrophy in DDMS has been reported previously,² the presence of ipsilateral midbrain atrophy in our patient adds to the evidence supporting transneuronal degeneration. According to this theory, cerebral hemiatrophy produces a diminished cortical output through corticopontine fibers, producing transneuronal atrophy of the homolateral pontine nuclei, which in turn produces tertiary anterograde transneuronal degeneration of the contralateral cerebellar granule cell layer.³ This hypothesis is further supported by the observations of Winkler et al.,⁴ who reported the postmortem findings of DDMS with crossed cerebellar atrophy in a 59-year-old woman. They described neuronal loss and gliosis of the subcortical white matter, thalamus, pontine nuclei, and inferior olivary nucleus, with pyramidal tract atrophy on the side of the cortical atrophy and centrolobular cerebellar sclerosis on the contralateral side. Our patient provides a strong radiological correlation with those pathological findings, and adds valuable information that such changes can be seen as early as the second decade of life.

It is thus possible that our patient experienced late onset of the brain insult due to intracranial infection, which produced cerebral hemiatrophy with dilated sulci and encephalomalacia. This was responsible for his hemiparesis and recurrent seizures. Since the brain insult occurred before 3 years of age, compensatory calvarial changes were present. The diminished

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Fig. 1. A and B: T1-weighted axial brain MRI showing (A) diffuse left cerebral hemiatrophy with dilated sulci and left calvarial thickening and (B) hyperaeration of the left frontal sinus. C: T2-weighted (T2W) fluid-attenuated inversion recovery axial brain MRI showing ipsilateral atrophy of the cerebral peduncle (left). D: T2W coronal brain MRI showing crossed cerebellar atrophy (right).

cortical output from the atrophied hemisphere provoked ipsilateral midbrain and contralateral cerebellar atrophy. Hence, severe and extensive involvement of one cerebral hemisphere in early childhood can produce ipsilateral midbrain and crossed cerebellar changes even early in life. The most plausible explanation for this is disruption of the reciprocal anatomic connections between these areas.

Conflicts of Interest

The authors have no financial conflicts of interest.

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