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NEUROLOGICAL PICTURE

doi: 10.1136/jnnp.2006.099465

Mid-brain molar tooth sign: expanding the clinical spectrum

A 17-year-old male, a single child of non-consanguineous parentage, presented with non-progressive gait ataxia, which he had had since childhood. He had suffered mild perinatal hypoxic injury, not requiring prolonged intensive care. Motor milestones were acquired late; he walked independently only from the age of three and swayed while walking. Despite moderate mental retardation, he acquired other milestones normally and was independent for activities of daily living. General examination was normal. He had a Mini-Mental Status Examination score of 25/30, slurred, scanning and staccato speech, normal eye movements, hypotonia and significant gait and truncal ataxia.

Magnetic resonance images showed superior vermician dysplasia with agenesis of mid and inferior vermician lobules. Cerebellar hemispheres were apposed to one another in the midline (fig 1A). Abnormally thick and maloriented superior cerebellar peduncles contributed to the classic molar tooth sign (MTS) in transverse images (fig 1B). The fourth ventricle appeared dilated, with "batwing" morphology at the level of the upper pons (fig 1C) and a triangular shape at the level of the mid-brain (fig 1B). The mid-sagittal image showed a high position of the fourth ventricle with a curved roof (fig 1D).

Anomalies at the junction of the mid-brain and hind-brain give rise to the appearance of the MTS in transverse images.¹ MTS is usually seen in Joubert's syndrome, characterised by cerebellar vermis hypoplasia, with the clinical triad of neonatal breathing abnormalities, oculomotor apraxia and hypotonia. However, MTS has been reported in other related, yet clinically and genetically distinct, disorders that show some features of Joubert's syndrome with additional findings.^{2–3} These include Dekaban–Arima syndrome (retinal dystrophy, renal cysts), Senior–Löken syndrome (renal cysts, nephronophthisis), COACH syndrome (cerebellar vermis hypoplasia, oligophrenia, ataxia, coloboma, hepatic fibrosis), Malta syndrome

(hydrocephalus, encephalocele, retinal dystrophy, renal cysts) and Varadi–Papp syndrome (orofaciocigital anomalies).^{2–4} Only a minority of patients with MTS have genetically proved Joubert's syndrome.³ Our patient presented with non-progressive cerebellar ataxia, hypotonia and moderate mental retardation, without abnormalities in eye movement or respiratory disturbances during infancy. Hence, the clinical features were distinct from Joubert's syndrome and related disorders. The high position and curved and dilated shape of the fourth ventricle in our patient is consistent with earlier observations that its presence in patients with MTS predicts a favourable prognosis.³ This report serves to expand the spectrum of clinical disorders associated with the mid-brain MTS.

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Competing interests: None declared.

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Figure 1 (A) Coronal T1 weighted magnetic resonance images showing superior vermician dysplasia with agenesis of mid and inferior vermician lobules and apposing cerebellar hemispheres (arrows). (B) Transverse T2 weighted image at the level of mid-brain interpeduncular fossa showing classical molar tooth sign (arrow) and triangular shape of the fourth ventricle (arrowhead). (C) Transverse T2 weighted image at the level of the upper pons showing a dilated fourth ventricle with bat-wing morphology (arrow). (D) Mid-sagittal T1 weighted image showing a high position of the fourth ventricle (arrow) and curved roof (arrowhead).