

appropriate and CT and MRI findings are suspicious for facial or cranial tumor. MNTI secretes high levels of vanillylmandelic acid and catecholamines in the urine, which can be used to make a preoperative diagnosis [3–5, 9, 10].

In the current patient, CT and MRI 3D reconstructed images, enhanced with color, aided neurosurgical recognition of the calvarial relationships of MNTI to cerebral anatomy.

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Locked-in Syndrome with Bilateral Peduncular Infarct

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ABSTRACT

Locked-in syndrome due to midbrain infarct is very rare. This report describes a patient with locked-in syndrome whose magnetic resonance images showed bilateral infarcts in the cerebral peduncle. Cerebral angiography disclosed bilateral irregular thrombotic narrowing of the vertebral and proximal basilar arteries with rostral basilar artery occlusion, a previously unreported finding.

Park SA, Sohn YH, Kim WC. Locked-in syndrome with bilateral peduncular infarct. *J Neuroimag* 1997;7:126–128

Locked-in syndrome (LIS), defined as complete paralysis of bulbar muscles and extremities with preserved consciousness, is caused almost exclusively by bilateral ventral pontine infarcts [1–3]. An extensive MEDLINE search of the English literature found only 4 cases of LIS with bi-

Received Jun 16, 1996, and in revised form Nov 4. Accepted for publication Nov 5, 1996.

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lateral midbrain stroke [4–7]. Although a previous autopsy study suggested rostral basilar artery thrombosis to be a main vascular lesion responsible for midbrain LIS [4], this has not been confirmed by cerebral angiography before death.

Case Report

A 64-year-old woman with a diabetic history of 6 years had an abrupt onset of vertigo, dysarthria, and quadriplegia. Ten days earlier, she had transient left hemiparesis.

Her blood pressure, pulse, and temperature were normal. She was mute but alert, with isocoric pupils reacting to light. Horizontal gaze to the left side was impaired, but she could move her eyes to the right on request. Corneal reflexes were present, but gag reflexes were severely depressed. Other than eye movements, there were no voluntary movements. She grimaced but did not withdraw in response to noxious stimuli. Tendon reflexes were increased with extensor plantar responses bilaterally.

Initial brain computed tomography (CT) scans appeared normal. Chest x-ray films revealed a consolidation in the right middle lung field. On the second hospital day, she could look to the left, but other neurological signs were not changed. Heparin was administered intravenously for a week, followed by aspirin and ticlopidine. Axial T2-weighted magnetic resonance imaging (MRI) of the brain on the twenty-fourth hospital day demonstrated focal high-signal lesions involving the middle and lateral portion of the cerebral peduncle bilaterally (Fig 1A) and a small high-signal spot in the flow-void area of the basilar artery (Fig 1B), but other parts of the brainstem appeared normal except for two small high-signal spots in the pons. The T1-weighted parasagittal images demonstrated a linear structure isointense with the brainstem in the pontine cistern (Fig 1C). Cerebral angiography on the thirty-third hospital day disclosed an irregular thrombotic narrowing of the vertebral and proximal basilar arteries with rostral basilar artery occlusion (Fig 2).

Two weeks after admission, the neurological deficits gradually improved. Four months later, she could partially open and close her mouth, speak with a comprehensible slur, move her right extremities against some resistance, and raise her left extremities against gravity. Swallowing difficulties persisted.

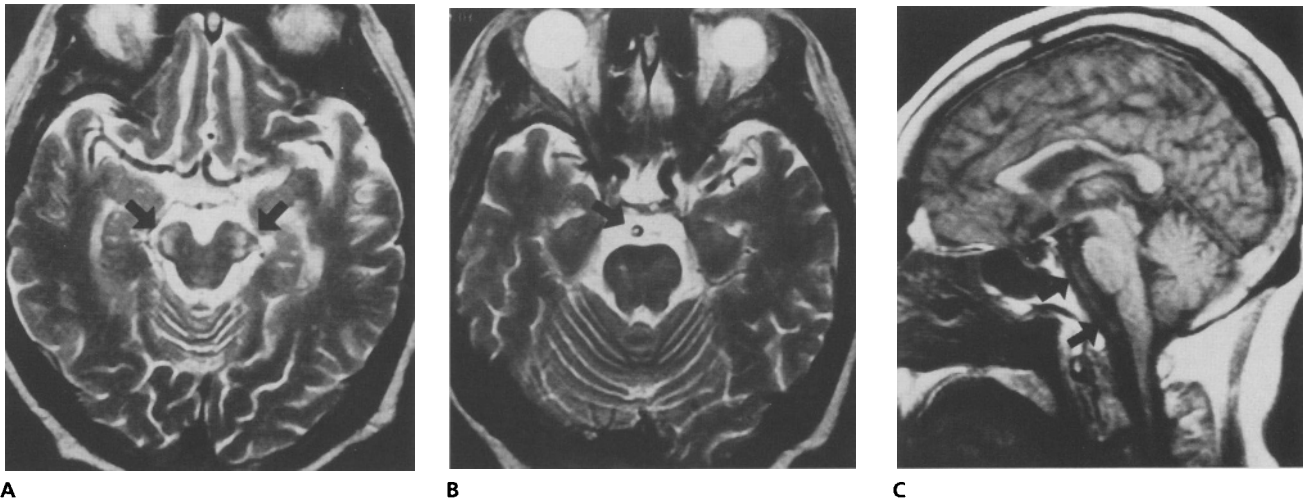


Fig 1. (A) Axial T2-weighted (TR 4000/TE 103) MRI demonstrated bilateral infarcts (arrows) that involved the cerebral peduncles. (B) Axial T2-weighted (TR 4000/TE 103) MRI demonstrated a high-signal spot (arrow) in the flow-void area, suggesting basilar artery thrombosis, and a small high-signal lesion in the right side of the basis pontis. (C) Parasagittal T1-weighted (TR 500/TE 12) MRI demonstrated a linear structure (arrows) in the pontine cistern, suggesting basilar artery thrombosis.

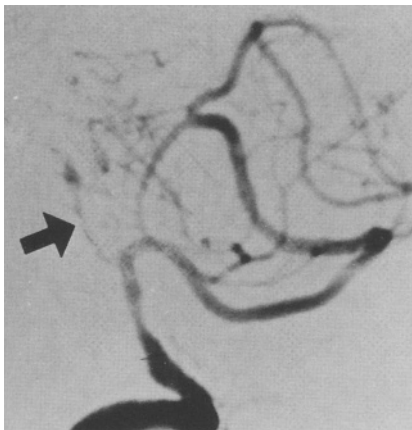


Fig 2. Angiography (lateral view after left vertebral arterial injection) demonstrated irregular thrombotic narrowings in the vertebral and proximal basilar artery with rostral basilar artery occlusion. The arrow points to the stenotic basilar artery.

Discussion

The report here of quadriplegia and bulbar palsy with preserved consciousness and communication by blinking is consistent with the original description of LIS [1]. Some pontine LIS patients had lesions that extended to the midbrain [2, 8, 9], but LIS due to isolated midbrain lesion is extremely rare. To the authors' knowledge, only 4 such patients whose lesions were confirmed by autopsy [4–6] or brain MRI [7] have been reported in the

English literature. Meienberg and coauthors [10] used the term “mesencephalic locked-in syndrome” to describe a quadriplegic patient with bilateral oculomotor palsy and ptosis; however, the responsible lesion was not confirmed.

The reported midbrain lesions responsible for LIS were quite similar to that of the present patient, which was confined bilaterally to the cerebral peduncle. Three patients including the present one had lesions mainly involving the central and lateral portion of cerebral peduncle and intact eye movements [4, 7]. One patient [6] had lesions involving the medial two-thirds of each cerebral peduncle and impaired horizontal eye movements. One patient had total oculomotor palsy due to bilateral infarcts in the cerebral peduncles; however, the boundary of the lesion was not clearly described [5]. The corticopontine neurons responsible for eye movements are thought to pass through the medial portion of the peduncle, while those for bulbar and limb muscles pass through the other portions [5, 7]. Thus, sparing of the medial portion of the cerebral peduncle is presumably related to the preservation of eye movements. In this patient, T2-weighted MRI also revealed two small high-signal lesions in the right side (see Fig 1B) and the paramedian region of the basis pontis, but these lesions were too small to be of clinical significance.

Rostral basilar artery occlusion was demonstrated by autopsy in a midbrain

LIS patient [4], but was never confirmed neuroradiologically. In the present patient, the MRI findings, a linear isosignal structure on T1-weighted parasagittal images and a high-signal spot in the flow-void area of the basilar artery on T2-weighted axial images, strongly suggest the presence of basilar artery thrombosis [11, 12]. These findings were confirmed by cerebral angiography showing an occlusion of the rostral portion of the basilar artery with irregular thrombotic narrowing involving its proximal portion and the distal portion of the vertebral arteries. In other patients, LIS with pontine infarcts usually resulted from basilar artery occlusion [5, 8, 9], but their occlusion levels exclusively occurred more proximally compared with that in the present patient.

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torticollis. MRI and ultrasound were performed to exclude neoplasm. MRI showed a diffusely enlarged, right sternocleidomastoid muscle that was isointense to normal muscle on T1-weighted conventional spin-echo images. The muscle became progressively brighter and more heterogeneous in signal intensity on proton density- and T2-weighted conventional spin-echo sequences. A thin rim of bright signal intensity on the T2-weighted images surrounded most of the muscle. Ultrasound demonstrated fusiform enlargement with patchy inhomogeneous areas of increased echogenicity. At 14 weeks there were signs of clinical improvement, with softening of the mass and decreased torticollis.

Entel RJ, Carolan FJ. Congenital muscular torticollis: magnetic resonance imaging and ultrasound diagnosis. *J Neuroimag* 1997;7:128–130

Congenital Muscular Torticollis: Magnetic Resonance Imaging and Ultrasound Diagnosis

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ABSTRACT

The magnetic resonance imaging (MRI) and sonographic appearances of congenital muscular torticollis were studied in a 6-week-old female infant who presented with a firm mass in the right sternocleidomastoid muscle and clinical signs of

Congenital muscular torticollis is a well-recognized disorder characterized by a unilateral contracture of the sternocleidomastoid muscle (SCM), often associated with head tilt and a palpable mass [1]. Although several prenatal theories have been proposed, it is often thought to result from stretch injury occurring during delivery. This condition usually resolves with conservative treatment in 4 to 8 months but refractory cases persisting for 1 year may require surgery [2]. There are only a few reports describing the magnetic resonance imaging (MRI) appearance of torticollis. This case study correlates both MRI and sonographic findings in an infant with congenital muscular torticollis.

Case Report

A 6-week-old female infant presented with a firm mass in the right SCM. There were mild clinical signs of torticollis but the remainder of the physical examination revealed no abnormalities. She was born 5 days after maturity by cesarean section, and the birth weight was 2,615 gm. At birth, an area of "slight reddening" was noted on the right side of the neck, along with a small bruise on the left thigh. No medical, surgical, or physiotherapeutic intervention was needed.

MRI with a 2-T high-field-strength unit (Elscent, Haifa, Israel) performed at 6 weeks showed a diffusely enlarged right

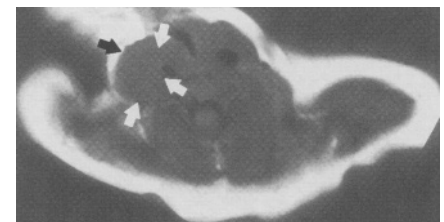
SCM, which was isointense to normal muscle on axial T1-weighted conventional spin-echo images (Fig 1A). Axial T2-weighted conventional spin-echo sequences revealed heterogeneously increased signal with a well-defined thin rim of bright signal surrounding most of the muscle (Fig 1B).

Ultrasonography (Acuson, Mountain View, CA) showed diffuse fusiform enlargement of the right SCM without focal mass or calcifications. Echogenicity of the area was inhomogeneous and increased compared to that of normal muscle (Figs 2A, 2B). The differential diagnosis for the neck mass included congenital torticollis, cervical adenopathy, neuroblastoma, lymphoma, cystic hygroma, branchial cleft cyst, and hematoma.

By 9 weeks there was persistence of the firm, enlarged right SCM. By 14 weeks the neck mass had softened and decreased in size. The patient also showed clinical improvement with decreased torticollis.

Discussion

Congenital or infantile muscular torticollis is a neck deformity also known as fi-



A



B

Fig 1. (A) Axial T1-weighted MRI showing an enlarged right sternocleidomastoid muscle (arrows) that is isointense to normal musculature. Also note the abnormal position of the head. (B) Axial T2-weighted MRI of the neck showing enlargement and heterogeneous areas of increased signal intensity of the right sternocleidomastoid muscle (arrows). Also note the rim of bright signal partially surrounding the muscle. (Courtesy of All Children's Hospital, St. Petersburg, FL.)

Received Jun 19, 1996, and in revised form Aug 21 and Sep 17. Accepted for publication Sep 18, 1996.

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