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Locked-In Syndrome

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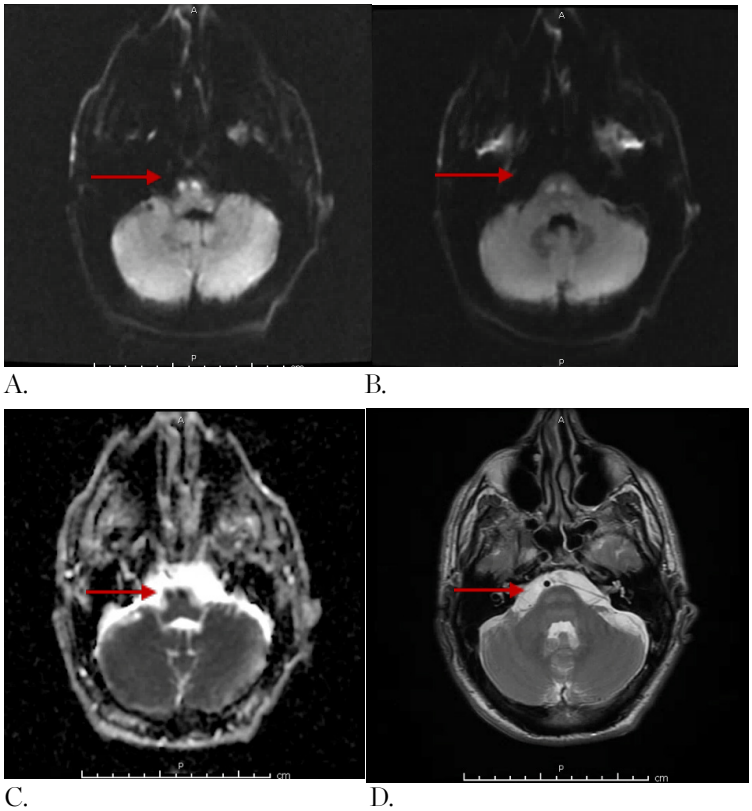

Figure 1. Axial MRI of the brain obtained 36 hours after admission demonstrates (by red arrows) symmetric areas of increased signal in diffusion weighted imaging (A, B) with decreased signal in apparent diffusion coefficient (C) and high T2 signal (D) at the level of the medullary pyramids and caudal pons involving predominantly white matter tracts.

CASE DESCRIPTION

A 56-year-old male with history of alcohol abuse was admitted to our trauma service following a motor vehicle collision, where he was a passenger. The patient was found unresponsive with a Glasgow Coma Score of 3 (E1, V1, M1). Initial brain imaging demonstrated small subdural hematoma along the falx cerebri as well as the frontal convexities. A cervical spine magnetic resonance imaging (MRI) demonstrated severe post-traumatic changes at C5 - C6 level without evidence of cord compression. The patient regained consciousness 36 hours after admission. His neurological examination one week later was remarkable for an awake patient able to blink on command. Pupils were equal and reactive and extraocular movements had restricted bilateral abduction. Remaining bilateral vertical eye movements were intact. He had bilateral facial, lingual, and palatal weakness that impaired articulation, swallowing, and respiratory ability. He had flaccid quadriplegia with hyper-reflexia and bilateral Hoffman signs were present, and he required full ventilator support through tracheostomy. His neurological examination was consistent with "Locked-in Syndrome",[1] suggestive of a lesion at the level of the pons. MRI of the brain demonstrated symmetric areas of subtle restricted diffusion at pons and medulla involving corticospinal and corticobulbar fibers, suggestive of brainstem traumatic axonal injury (Figure 1).

DISCUSSION

Locked-in Syndrome is described clinically as quadriplegia and anarthria with preserved consciousness.[2] Disruption of the corticospinal, corticopontine, and corticobulbar tracts in the brainstem lead to these clinical features. There are multiple etiologies to Locked-in Syndrome, most commonly due to ischemic or hemorrhagic stroke, pontine hemorrhage related to hypertension of vascular malformations, or trauma.[3] We described a patient with Locked-in Syndrome due to traumatic axonal injury (TAI) or diffuse axonal injury (DAI), a common finding in patients with severe traumatic brain injury.[4] Angular and rotational acceleration and deceleration forces to the head lead to stretching of brain tissue and subsequent damage to white matter. After traumatic brain injury, axonal degeneration from DAI is considered progressive. Disruptive axonal transport results in axonal swelling followed by secondary disconnection and anterograde degeneration (more common) or even orthograde degeneration (less common).[5] MRI can characterize these lesions typically located in the white matter of cerebral hemispheres, corpus callosum, and brainstem.[6]

REFERENCES

[1] Patterson JR, Grabois M. Locked-in syndrome: A review of 139 cases. Stroke 1986; 17(4):758-764. PMID: 3738962.
[2] Plum F, Posner JB. The Diagnosis of Stupor and Coma. Philadelphia: FA Davis Company, 1966. ISSN 0069-9446.
[3] León-Carrión J, Van Eeckhout P, Domínguez-Morales Mdel R, Pérez-Santamaría FJ. The locked-in syndrome: A syndrome looking for a therapy. Brain Injury 2002; 16(7):571-582. PMID: 12119076.
[4] Skandsen T, Kvistad KA, Solheim O, Strand IH, Folvik M, Vik A. Prevalence and impact of diffuse axonal injury in patients with moderate and severe head injury: A cohort study of early magnetic resonance imaging findings and 1-year outcome. J Neurosurg 2010; 113(3):556-563. PMID: 19852541.
[5] Johnson VE, Stewart W, Smith DH. Axonal pathology in traumatic brain injury. Exp Neurol 2013; 246:35-43. PMID: 22285252.
[6] Scheid R, Preul C, Gruber O, Wiggins C, von Cramon DY. Diffuse axonal injury associated with chronic traumatic brain injury: Evidence from T2*-weighted gradient-echo imaging at 3 T. AJNR Am J Neuroradiol 2003; 24(6):1049-1056. PMID: 12812926.

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