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The cough/laugh syndrome: MR evaluation.

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The Cough/Laugh Syndrome: MR Evaluation

The cough/laugh syndrome is characterized by the onset of severe headache immediately after an episode of coughing, laughing, or straining. It has been associated with pathologic changes in the posterior fossa, in particular, cerebellar tonsillar herniation or ectopia [1-3]. MR imaging is important in the evaluation of craniocervical anomalies, particularly Chiari malformations [4, 5]. We present the MR findings in two patients who had headache associated with the cough/laugh syndrome.

Case Reports

Case 1 is a 46-year-old man who had a history of severe episodic headaches consistently precipitated by laughing or coughing. Case 2 is a 16-year-old girl who also had a history of severe occipital headaches that occurred immediately after episodes of coughing or laughing or when she performed the Valsalva maneuver. Neither patient had had gross ataxia, but the teenager had noticed a subtle deterioration in gymnastic coordination and in academic performance. Neurologic examination of this patient showed horizontal nystagmus on downward gaze and bilateral hyperreflexia. Both patients had symptomatic relief with analgesic therapy.

In case 1, MR imaging showed cerebellar tonsillar ectopia (Fig. 1); tonsils extended approximately 8 mm inferior to the foramen magnum. In case 2, MR also showed cerebellar tonsillar herniation, with the tonsils extending approximately 11 mm inferior to the foramen magnum (Fig. 2). No syrinx could be detected, but mild dilatation of the lateral ventricles and fourth ventricle was observed.

Discussion

The headache associated with the cough/laugh syndrome typically occurs after an episode of increased intraabdominal pressure, and it usually persists for a relatively brief period [1-6]. It has been suggested [1-3] that the headache is caused by a valvelike cerebellar tonsillar obstruction of CSF flow at the level of the foramen magnum that precipitates a CSF pressure differential between the spinal subarachnoid space and the ventricles. Disruption of craniospinal CSF circulation also has a possible role in the formation of a syrinx [1]. The dynamic-pressure-gradient theory is supported by the ob-

servation that cerebellar tonsillar surgical decompression may result in symptomatic relief.

The degree of tonsillar ectopia remains highly variable in the Chiari I malformation. Similarly, a broad spectrum of severity of clinical expression exists for cerebellar tonsillar ectopia [5, 7]. If symptoms develop, they tend to do so during the second to fourth decade of life, and clinical manifestations typically include signs of cranial nerve or cerebellar impairment [7].

Because of the association between the headache caused by the cough/laugh syndrome and tonsillar herniation or ectopia, patients who have this symptom complex should be considered candidates for neuroradiologic imaging. MR, by virtue of its sagittal imaging capability, offers an excellent means for assessing craniocervical relationships in these patients [4, 5]. Furthermore, MR permits evaluation of associated intramedullary disease, such as syringomyelia and compressive myelopathic edema or myelomalacia. Because of the potential for successful neurosurgical treatment of the cough/laugh syndrome, the recognition of specific associated MR findings has important therapeutic implications, and cerebellar tonsillar ectopia should be suspected in patients who have this unusual symptom complex.

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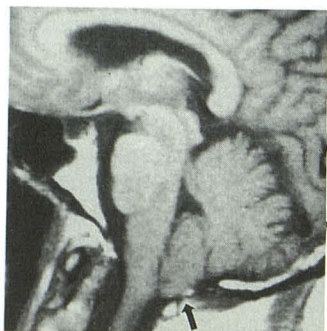
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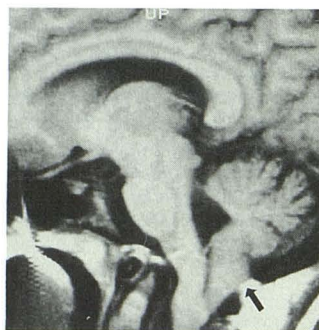
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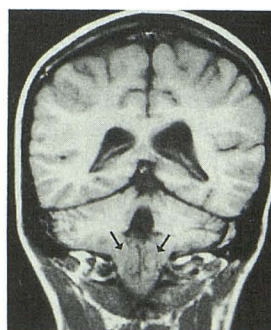
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1



2A



2B

Fig. 1.—Sagittal T1-weighted midline MR image, 450/20, shows cerebellar tonsillar herniation (arrow) and mild deformity of caudal aspect of fourth ventricle.

Fig. 2.—A and B, Sagittal (A) and coronal (B) T1-weighted midline MR images, 450/20, show cerebellar tonsillar herniation (arrows) accompanied by dilatation of fourth ventricle and compression of dorsal cervicomedullary junction.