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MRI Findings Differentiating Tonsillar Herniation caused by Idiopathic Intracranial Hypertension from Chiari I Malformation

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Purpose

Patients with Idiopathic Intracranial Hypertension (IIH) could have cerebellar tonsillar herniation ≥ 5 mm mimicking Chiari malformation I (CMI), which can result in misdiagnosis and unjustified treatment. The goal of this study was to identify imaging findings that could distinguish CMI from IIH.

Similarity between CMI and IIH:

- Both can cause intractable headache
- Both can cause headache in the back of the head
- Both can show tonsillar herniation on MRI
- Both can present in young adults

Methods

Ninety-eight patients with IIH, 81 patients with CMI, and 99 controls were retrospectively assessed. Two neuroradiologists blindly reviewed MR images. Patients with IIH and tonsillar herniation ≥ 5 mm (IIH_{TH}) were compared with patients with CMI and controls regarding the extent of tonsillar herniation (ETH), bilateral transverse sinus stenosis (BTSS), empty sella >50%, and bilateral tortuosity of optic nerve (BTON).

Results

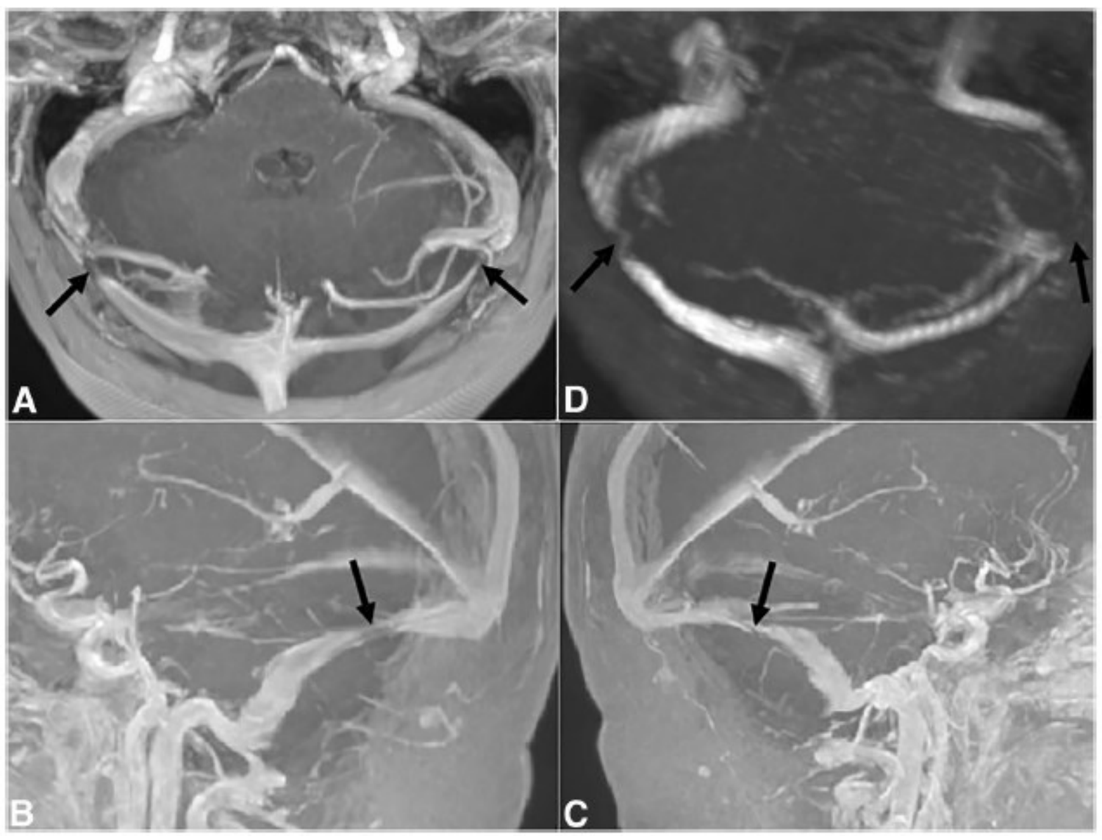
13/98 (13.2%) patients with IIH had tonsillar herniation ≥ 5 mm (IIH_{TH}). They were significantly younger and had higher BMI compared with CMI patients and controls. ETH was significantly less in the patients with IIH_{TH} than CMI (6.5 ± 2.4 mm vs. 10.9 ± 4.4 mm; $p < 0.001$). BTSS and empty sell > 50% were more common in patients with IIH_{TH} than CMI ($p < 0.001$ and $p = 0.003$, respectively). No differences were seen between CMI and controls. BTON was significantly more common in patients with IIH_{TH} compared to control ($p = 0.01$) but not to the CMI ($p = 0.36$). Sensitivity and specificity to differentiate IIH_{TH} from CMI were 69.2% and 96.1% for BTSS and 69.2% and 75.3% for empty sella > 50% .

Conclusions

IIH and CMI can have similar clinical presentations. The differentiation of these two entities is important because the treatment is different. The presence of BTSS and/or empty sella > 50% in patients with ETH ≥ 5 mm should suggest further evaluation to exclude IIH before considering CMI surgery.

Sensitivity, Specificity and Likelihood Ratios of MRI Findings in Differentiating IIH_{TH} from CMI

	sensitivity	specificity	Positive likelihood ratio
Bilateral transverse sinus stenosis	69.2%	96.3%	18.7
Empty sella > 50%	69.2%	75.3%	2.8
Bilateral tortuosity of optic nerve (BTON).	23.1%	88.9%	2.1



A) Axial post-contrast MRI of the posterior fossa shows more than 50% narrowing in bilateral transverse sinus in a patient with IIH (arrows). B&C) Sagittal post-contrast MRI demonstrates the stenosis on both sides (arrows). D) Axial Phase Contrast MRV of the posterior fossa shows bilateral transverse sinus stenosis.