Case Report

Case-based review of isolated tentorial hypoplasia with herniation: Applying Occam's Razor

Mona Tiwari[1], Shreyashi Jha[2], Santosh K Pendyala[2]

From [1]Head, Department of Neuro Radiology, [2]Consultant Neurologist, Department of Neurology, Institute of Neurosciences, Kolkata, West Bengal, India

ABSTRACT

Descriptions of isolated tentorium hypoplasia (TH) without any major clinical correlates have been confined to scarce clinical reports. This report entails the first case of an adult with a bilateral tentorial defect leading to herniation of bilateral occipital lobes, without any alarming neurological sequel. A 43-year-old gentleman presented to the outpatient department with a headache for 6 months. Neurological examination was normal. On neuroimaging, a "mass" was noted in the quadrigeminal and left superior cerebellar cistern. On careful evaluation, herniation of cuneus and precuneus in quadrigeminal plate cistern was observed bilaterally, more on the right side secondary to a bilateral defect in the superior part of tentorium cerebelli evident by non-visualization of the superior part associated with dilatation of posterior part of lateral ventricles. This case report aims to increase awareness about TH to prevent misdiagnoses along with careful follow-up so as not to dismiss the finding as clinically irrelevant. The sparse literature and vague clinical symptomatology contribute to inadequate identification of this entity.

Key words: Cuneus, Headache, Hypoplasia, Precuneus, Tentorial

The spectrum of tentorial defects has been described ranging from small apertures to aplasia, and hypoplasia, the latter of which are frequently associated with severe malformations such as Dandy-Walker malformations Dandy-Walker, Arnold-Chiari, and Gorlin syndrome [1]. Descriptions of isolated tentorium hypoplasia (TH) without any major clinical correlates have been confined to scarce clinical reports (Table 1) [1-10]. A Turkish study estimated the prevalence of TH among 5,000 odd patients screened during a period of 1 year as 2.22% [11].

This report entails the first case of an adult with a bilateral tentorial defect leading to herniation of bilateral occipital lobes, without any alarming neurological sequel.

CASE REPORT

A 43-year-old gentleman presented to the outpatient department with a headache for 6 months. The headache was bifrontal, dull aching in nature, mild to moderate in intensity, present on 2 days of the week, lasting for 1 h, without any other associated features, and not incapacitating his activities of daily living. He did not have any associated medical co-morbidities or any significant past neurological history.

Access this article online	
Received - 21 November 2024 Initial Review - 09 December 2024 Accepted - 22 March 2025	Quick Response code
DOI: 10.32677/ijcr.v11i6.4924	

General examination including vitals and neurological examination were normal. On neuroimaging, a "mass" was noted in the quadrigeminal and the left superior cerebellar cistern (Fig. 1a and c).

On careful evaluation, herniation of cuneus and precuneus in quadrigeminal plate cistern was observed bilaterally, more on the right side secondary to a bilateral defect in the superior part of tentorium cerebelli evident by non-visualization of the superior part associated with dilatation of posterior part of lateral ventricles (Fig. 1b and d). No other associated abnormality was noted. Headache responded to analgesics. He is currently under follow-up. At 6 months of follow-up, he did not have any neurological deficits.

DISCUSSION

The tentorium cerebelli is the second largest dural reflection, containing the transverse sinus and dividing the intracranial space into supratentorial and infratentorial compartments [5]. Isolated TH is very rare with little knowledge of its clinical relevance, evolution, and etiopathogenesis. The widely accepted etiology has been attributed to abnormal fusion of the medial and lateral aspects of the tentorium during embryogenesis [5,7]. While birth trauma and perinatal hypoxia have been postulated to accentuate TH, evidence supporting this hypothesis is scarce.

Correspondence to: Mona Tiwari, Institute of Neurosciences Kolkata, 185/1 AJC Bose Road, Kolkata - 700017, West Bengal, India. E-mail: drmonatiwari@gmail.com

Table 1: Published literature on tentorial hypoplasia and associated herniation

S. no.	Author, year	Age/sex	Clinical symptoms/ associations	Side of tentorial defect	Brain structures herniated	Associated radiological abnormalities
1.	Gun 1985 [1]	8/F	Occipital mass	Right	Occipital lobe	NA
2.	Tanohata *et al*., 1995 [2]	68/F	Galactorrhea	Left	Temporal lobe	-
3.	Abi-Jaoudeh and Chevrette 2006 [3]	2 females	Loss of consciousness and panic attacks	Right	Cingulate gyrus and medial occipitotemporal gyrus	NA
		1 Male	Dizziness, nausea, vomiting Headaches secondary to Valsalva	Right		Previous benign
				Right	Parahippocampal gyrus	Cerebral tumor
					Parahippocampal gyrus	NA
4.	Thomaere *et al*., 2015 [4]	50/F	Vertigo, headache	Right	Occipital Lobe	-
5.	Sun *et al*., 2018 [5]	5/M		Right	Temporal lobe	Optic Atrophy
		43/F	Tremor	Right	Cuneus	-
		42/F	Left-sided homonymous hemianopia	Right	Medial occipital gyrus	-
6.	Agrawal *et al*., 2020 [6]	38/F	Headache	Left	Left cuneus and precuneus	-
7.	Shoyab 2022 [7]	11/F	Headache, vertigo, vomiting	Bilateral	Left precuneus and cuneus	-
					Lingual gyrus of the right occipital lobe	
8.	Tanaka *et al*., 2000 [8]	22/F	Left trigeminal neuralgia	Left	Medial temporal lobe	Trigeminal nerve compression caused by distortion of the superior cerebellar artery by the herniated temporal lobe
9.	Jain and Jana 2021 [9]	25/F	Headache	Left	Left precuneus and cuneus	-
10.	Bernardo *et al*., 2020 [10]	9/F	Precocious puberty	Bilateral	Bilateral occipital lobes and straight sinus	Small posterior fossa, and hypoplasia of the straight sinus, the distal half of the corpus callosum
11.	Present case	43/M	Headache	Bilateral	Bilateral herniation of cuneus-precuneus in quadrigeminal plate cistern, more on right side.	-

An extensive literature review (Table 1) identified only nine cases with TH, with most of the defects being unilateral tentorial defects and the two patients with bilateral defects being in the pediatric age group. We describe the first adult with a large bilateral defect resulting in herniation of bilateral occipital lobes. Interestingly, the patient remained mildly symptomatic, thus poorly correlating with his sinister imaging.

As noted in Table 1, the symptoms attributed to TH remain largely non-specific. Among the nine cases, two cases alone had symptoms that could be correlated with the affected functional anatomy that included left homonymous hemianopia consequent to herniated medial occipital gyrus [5] and trigeminal neuralgia secondary to superior cerebellar artery compression by herniated temporal lobe [8]. While the association of TH with other malformations demands intervention, this clinico-radiological dissociation in isolated TH is of uncertain significance, mandating close follow-up. Our patient did not have any additional neuroimaging abnormalities.

Although there is no consensus on management, it is important to raise awareness of ITH as being potentially symptomatic. A useful tool to estimate the accurate extent of clinical symptomatology of isolated TH is diffusion tensor imaging (DTI) to identify trapped tracts and fibers within the herniation [5]. These findings indirectly suggest that symptoms depend on the severity and location of the tentorial defect. However, our case study digresses and demonstrates a significant disparity between the clinical examination and radiological findings. In spite of a large defect, the patient remained clinically mildly symptomatic. However, the size ranges require further exploration since previous literature does not mention any cut-off measurements differentiating small, moderate, and large defects.

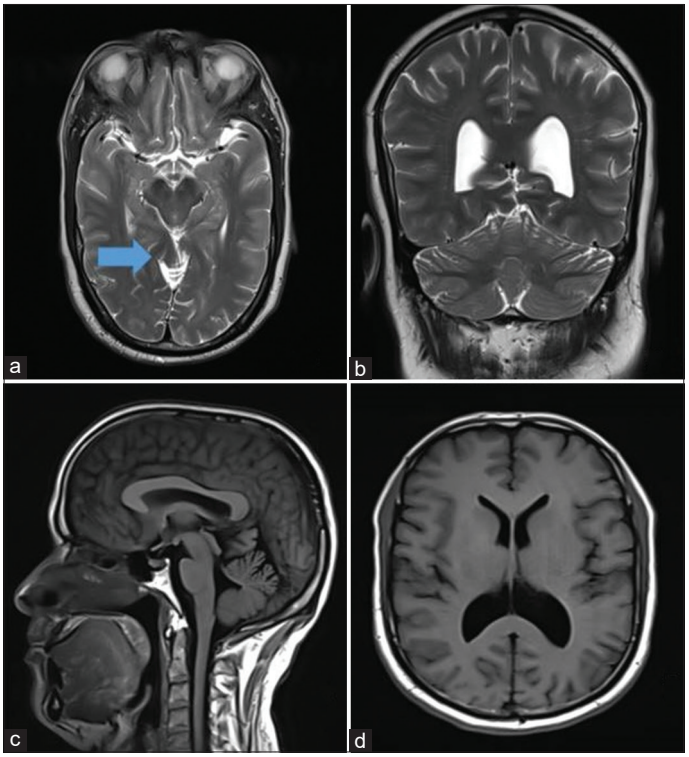

Figure 1: (a): Axial T2 weighted image showing herniation of cuneus-precuneus in quadrigeminal plate cistern, more on the right side; (b) Coronal T2 w image showing non-visualization of the superior part of tentorium cerebelli; (c) Sagittal T1 w image showing herniation of gyrus in quadrigeminal plate cistern; and (d) Axial T1 weighted image showing dilatation of posterior part of lateral ventricles

CONCLUSION

This case report aims to increase awareness about TH to prevent misdiagnoses along with careful follow-up to not dismiss the finding to be clinically irrelevant. The sparse literature and vague clinical symptomatology contribute to inadequate identification of this entity. Usage of DTI in large tentorial defects should be part of routine radiology practice to identify subclinical compression of vital structures.

REFERENCES

1. Gund A. Hypoplasia of the tentorium with pseudohernia of the occipital lobe into the posterior cranial fossa. Mod Probl Paediatr 1976;18:155-7.
2. Tanohata K, Okoshi T, Iwasawa T. Focal hypoplasia of the tentorium with temporal lobe herniation: CT demonstration. J Comput Assist Tomogr 1991;15:863-7.
3. Abi-Jaoudeh N, Chevrette E. Isolated hypoplasia of the tentorium. J Comput Assist Tomogr 2006;30:131-4.
4. Thomaere E, Schepers S, Termote B, Vanwyck R, Souverijns G. Tentorium hypoplasia with partial occipital lobe herniation. JBR-BTR 2015;98:96.
5. Sun Y, Bobra S, Kurian C, Ahluwalia-Singh B, Thomas A, Mehta H. MR and diffusion tensor imaging of isolated tentorial hypoplasia. Neurol Clin Pract 2018;8:79-81.
6. Agrawal SK, Premi V, Babu CR. Unilateral tentorial hypoplasia with ipsilateral brain herniation. J Med Sci Clin Res 2020;8:368-71.
7. Shoyab M. Bilateral occipital lobe herniations due to asymmetric tentorial defects: Incidental Computed Tomography (CT) findings and literature review. Cureus 2022;14:e32117.
8. Tanaka T, Nakazaki H, Hida T, Nakajima M, Abe T. Trigeminal neuralgia associated with tentorial agenesis and temporal lobe herniation--case report. Neurol Med Chir (Tokyo) 2000;40:124-7.
9. Jain R, Jana M. Rare case of brain herniation secondary to tentorial hypoplasia: Be careful while reporting! Neurol India 2021;69:1629-30.
10. Bernardo K, Roychowdhury S, Bhise V. Tentorium and straight sinus hypoplasia in a child: A case report. SN Compr Clin Med 2020;2:974-7.
11. Ünal Ö, Şara Hİ. The prevalence of isolated tentorial hypoplasia on magnetic resonance imaging. Pol J Radiol 2022;87:e506-9.

Funding: Nil; Conflicts of interest: Nil.

How to cite this article: Tiwari M, Jha S, Pendyala SK. Case-based review of isolated tentorial hypoplasia with herniation: Applying the Occam's Razor. Indian J Case Reports. 2025; 11(6):242-244.