

PARTIAL RIGHT TENTORIAL HYPOPLASIA WITH FOCAL BRAIN HERNIATION- A RARE INCIDENTAL FINDING

Radiodiagnosis

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ABSTRACT

Partial tentorial hypoplasia with ipsilateral focal brain herniation is an extremely rare cause of idiopathic intracranial herniation with very few cases reported in the literature. This is an essentially benign condition which may mimic a sinister pathology to a radiologist unaware of this rare entity. We present a case of 58 year old male presenting with giddiness and headaches. An MRI brain was performed which revealed this rare incidental finding which on the first look simulated a mass.

KEYWORDS

Idiopathic Brain Herniation, Tentorial Hypoplasia, parahippocampal gyrus, cingulate gyrus and medial occipitotemporal gyrus herniation.

INTRODUCTION

Idiopathic brain herniation is extremely rare with a prevalence of only 0.073%¹ with congenital tentorial hypoplasia being an even rarer cause. Agenesis or hypoplasia of tentorium is usually associated with other congenital posterior fossa malformations like Dandy Walker spectrum, Arnold Chiari malformation. Isolated hypoplasia of tentorium with adjacent brain herniation is a rare entity which usually does not present with any clinical symptoms.^{3,4}

Knowledge about this entity is essential as it can be misinterpreted for a pathology due to its unusual appearance.

CASE

A 58 year old male patient presented to the casualty with symptoms of giddiness and headache. There was no previous imaging. Blood reports were within normal limits. An MRI Brain was performed which revealed soft tissue in the quadrigeminal cistern on the right side on T2WI. Further evaluation with other sequences revealed that the soft tissue was the parahippocampal gyrus, isthmus of cingulate gyrus and medial occipitotemporal gyrus which had herniated into the quadrigeminal and superior cerebellar cistern. The herniation was secondary to a congenital defect in the right tentorial leaflet. No other abnormality was detected.

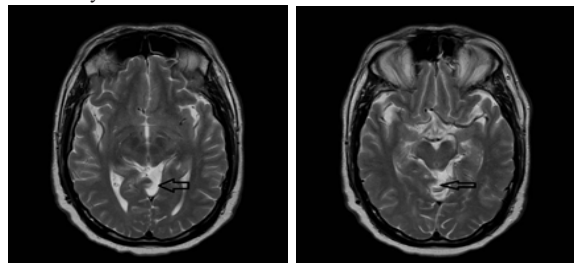


Fig 1 and 2 - Axial T2WI reveals soft tissue signal in the quadrigeminal cistern on the right side which is the herniated parahippocampal gyrus, isthmus of cingulate gyrus and medial occipitotemporal gyrus, herniating into quadrigeminal and superior cerebellar cistern.

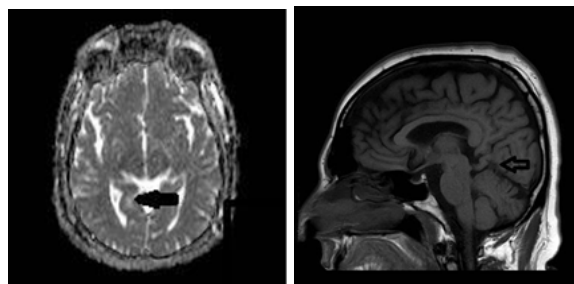


Fig 3.-DWI reveals no abnormal restriction. **Fig 4.-**Sagittal T1WI confirms the findings.

DISCUSSION

The tentorium is the second largest dural reflection and is a dural fold

that divides the brain into the supra and infratentorial neuroparenchyma. The tentorial hypoplasia is due to incomplete fusion of the lateral aspect of the tentorial leaflet and is associated with other CNS malformations like Dandy walker, Gorlin syndrome and Arnold Chiari malformation². Isolated tentorial hypoplasia with focal brain herniation is a very rare entity with only a few cases described in the literature. The clinical symptoms are usually unrelated to this, although it may be a plausible cause of headache. Although benign it is extremely important to identify this condition as it may mimic other intracranial diseases such as mass lesion, encephalocele or dural venous sinus thrombosis. Horowitz et al had reported a similar case where a parahippocampal herniation was misinterpreted as a lesion and the patient was subjected to biopsy and surgical exploration only to reveal normal herniated brain parenchyma¹. Few similar cases were reported by Tanohata et al in their study³ as well as by Abi-Jaoudeh and Chevrette⁴. In our case also there was soft tissue noted in quadrigeminal and superior cerebellar cistern. This soft tissue was herniated parahippocampal gyrus, isthmus of cingulate gyrus and medial occipitotemporal gyrus secondary to focal hypoplasia of right tentorial leaflet. The similar signal intensity, its continuity with the adjacent normal parenchyma and lack of abnormal enhancement helps in establishing the diagnosis.

CONCLUSION AND TEACHING POINTS

Congenital tentorial hypoplasia with focal brain herniation is a rare but benign entity which needs no treatment. It can be a diagnostic dilemma for a radiologist unaware of this entity and thus can be mistakenly presumed to be a mass and biopsied as noted in some rare instances⁵. Hence identifying this entity is crucial.

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