Research Article

Unilateral Tentorial Hypoplasia with Ipsilateral Brain Herniation

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Abstract
A case of unilateral tentorial hypoplasia with ipsilateral brain herniation is presented. This was an incidental finding in a 38 years old lady without any other associated findings. Computed Tomographic and Magnetic Resonance showed mild dilation of occipital horn of left lateral ventricle, which is a rare finding.

Keywords: Idiopathic Brain Herniation, Tentorial Hypoplasia, Computed Tomography, Magnetic Resonance Imaging.

Introduction
Idiopathic brain herniations are extremely rare in comparison to brain herniation caused by different intracranial pathologies. Tentorial hypoplasia is one of the rare finding. Clinical findings most probably depend upon its severity and location. Recognition of defective constitution of the dura and its extensions is extremely important to differentiate herniated brain from other intracranial pathologies.

Case
A 38 year old woman was referred from Neuro-Surgery department of our hospital to the department of Radiodiagnosis and Imaging with complaint of headache since 3 months. Computed Tomography(CT) & Magnetic resonance (MR)-imaging was performed and revealed left precuneus and cuneus herniation secondary to a congenital partial hypoplasia of the left tentorium, located mainly along the anterosuperior aspect (Fig-1 & Fig-2), which is a rare incidental finding. No other associated finding was noted in this case.

Imaging
Computed Tomography &Magnetic Resonance Images show precuneus and cuneus gyri of left parietal and occipital lobe respectively herniating in to quadrigeminal cistern through the hypoplastic anterosuperior aspect of left tentorium cerebelli (Fig-1 & Fig-2) posterior to splenium of corpus callosum (Fig-3). There is mild dilation of occipital horn of left lateral ventricle (Fig-4). No other abnormality was detected in the scan.
Fig 1- Coronal CT image showing precuneus (White arrow) and cuneus (White arrow head) gyri of left parietal and occipital lobes respectively herniating in to quadrigeminal cistern through the hypoplastic anterosuperior aspect of left tentorium cerebelli (White stars).

Fig 2- Coronal MRI T2WI showing precuneus (Black arrow) and cuneus (Black arrow head) gyri of left parietal and occipital lobes respectively herniating in to quadrigeminal cistern through the hypoplastic anterosuperior aspect of left tentorium cerebelli (Black stars).

Fig 3- Sagittal MRI T2WI showing precuneus (Black arrow) and cuneus (Black arrow head) gyri of left parietal and occipital lobes respectively herniating in to quadrigeminal cistern through the hypoplastic anterosuperior aspect of left tentorium cerebelli posterior to splenium of corpus callosum.

Fig 4- Axial MRI T1WI showing precuneus (White arrow) and cuneus (white arrow head) gyri of left parietal and occipital lobes respectively herniating in to quadrigeminal cistern through the hypoplastic anterosuperior aspect of left tentorium cerebelli with mild focal enlargement of ipsilateral occipital horn (White stars) of left lateral ventricle.
Discussion

Unlike brain herniation caused by mass effect due to trauma, tumours, infections or inflammatory conditions, idiopathic brain herniations are extremely rare, with a prevalence of 0.073%[1]. Although a very rare condition, it is extremely important to recognize and differentiate idiopathic brain herniations from other intracranial diseases such as mass lesion, encephalocele or dural venous sinus thrombosis to avoid subjecting the patient to unnecessary surgical and medical treatments[1-5].

The tentorium cerebelli, the second-largest dural reflection, is a crescent-shaped dural fold that extends over the posterior cranial fossa, separating the occipital and temporal cerebral hemispheres from the cerebellum and infratentorial brainstem[6, 7]. The tentorium cerebelli is a distinguishing landmark and divides the cranial cavity into the supratentorial and infratentorial spaces[6]. Tentorium cerebelli supports and transfers the weight of the occipital and temporal lobes of cerebrum so as to protect infratentorial cerebellum and brainstem[6]. Extensive central nervous system malformations, such as Dandy-Walker Syndrome and Arnold-Chiari Malformation are usually associated with agenesis or hypoplasia of tentorium cerebelli.

The most accepted hypothesis about the development of the tentorium is an abnormal fusion of the tentorium as described by Tanohata[8]. The tentorium develops as the second dural reflection, first the medial parts develop and secondly the lateral parts. Afterwards the medial parts involute and the lateral parts, which consist of the caudo-lateral and the rostrolateral parts, fuse. Abnormal fusion of the lateral parts can possibly lead to this anomaly. Birth traumas or perinatal insults can also be considered as the cause of this anomaly[8].

Previously few cases with similar findings were also reported including a case by Tanohata[8] in which computed tomography (CT) showed a focal hypoplasia of the left tentorium cerebelli with secondary protrusion of the temporal lobe into the superior cerebellar and quadrigeminal cisterns[8]. The other cases were described by Abi-Jaoudeh and Chevrette[9] that include (i) hypoplasia of the right tentorial leaf with protrusion of the isthmus of cingulate gyrus and the medial occipitotemporal gyrus into the superior cerebellar cistern; (ii) a small hypoplasia of the right tentorial leaf with discrete protrusion of the medial occipitotemporal gyrus and the parahippocampal gyrus into the cerebellar cistern and (iii) an angio-MRI for cerebral aneurysm screening, showed a hypoplasia of the right tentorial leaf with protrusion of the parahippocampal gyrus into the quadrigeminal cistern. [9]

In 2015E.Thomaere et al demonstrated a partial right occipital lobe herniation (inferior precuneus), secondary to a congenital focal hypoplasia of the tentorium, located on the anterosuperior part of the cerebellum[10]. In all these cases the symptoms were unrelated to these imaging findings.

Idiopathic herniation would occur due to localized defective constitution of the dura and dural extensions (e.g., tentorium cerebelli). Spaces confined with the meninges get occupied with herniated brain and it becomes necessary to differentiate them from other intracranial abnormalities such as mass lesion, encephalocele or dural venous sinus thrombosis. Their MR and CT signal characteristics are similar to adjacent brain parenchyma and lack of contrast enhancement & signal abnormalities help to discriminate herniations from intracranial pathologies and avoid unnecessary interventions.

Conclusion

Usually no known clinical symptoms are associated with idiopathic brain herniations but differentiation from other intracranial pathologies is crucial. In this case no treatment was required. Despite the rarity, recognition and differentiation of idiopathic brain herniation, is necessary to avoid needless surgical and medical treatments.
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