Abstract

Brain herniations predominantly result from increased intracranial pressures due to various etiologies and are usually acute. Chronic medial temporal lobe [uncal and parahippocampal] herniation has been rarely reported in literature. Unilateral and bilateral idiopathic temporal lobe [uncal and parahippocampal] herniations have been even more rarely reported. It's important to diagnose idiopathic temporal lobe herniations, as these are “do not touch” lesions. In this paper we presented two cases with nonemergent medial temporal lobe herniations. Standard MRI of the brain was performed in a 3T Siemens magnet with a standard head coil, to make the diagnosis. First patient is a 40-year-old female with a longstanding history of headaches and bilateral idiopathic uncal herniations. Second patient is a 21-year-old female with shunt revisions for hydrocephalus since a young age. She had unilateral right uncal and parahippocampal herniation, and an ipsilateral dural based right frontal calcified small mass without significant mass effect on the nearby structures.

Keywords

Idiopathic uncal herniation, Chronic uncal herniation, Chronic parahippocampal herniation

Abbreviations

PH: Parahippocampal Gyrus; MRI: Magnetic Resonance Imaging; LDTH: Lateral Descending Transtentorial Herniation

Introduction

Medial temporal lobe (uncal) herniation is usually thought of as a life-threatening situation in an acute setting. However, cases of idiopathic and chronic uncal and parahippocampal herniations have been rarely reported [1, 2]. In this paper we present two rarely reported cases, one case of idiopathic and one case of chronic medial temporal lobe descending transtentorial herniation. These cases demonstrate that significant herniation can be present without accompanying neurologic consequences [3]. So far, there has been one reported case in literature, where idiopathic significant uncal and PH herniation was surgically approached [1]. Hence radiologists and neurosurgeons should be aware of this entity before aggressively pursuing it. Given the significance of this awareness, we are presenting our cases with a detailed review of literature.
Nonemergent Descending Transtentorial Temporal Lobe Herniations - Case Reports of Idiopathic [Bilateral Uncal] and Chronic [Unilateral Uncal and Parahippocampal] Herniations with Review of Literature

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Case Reports

Patient 1:

40-year old female presented to the Neurology department with a long-standing history of headaches from the age of two. Her headaches were severe and occurred 3-4 times per week. They lasted up to 4 or 5 days and were centered behind her right eye with pain radiating to the back side of the ear. The headaches were associated with nausea, photophobia, phonophobia, double vision and dizziness. There wasn’t any known specific trigger or history of head trauma. Detailed neurological examination did not show any neurological disabilities or dysfunction. She had no significant laboratory data. MRI was obtained to evaluate for migraine.

Patient 2:

21-year old female presented to the Neurology department with a history of headaches and seizures. She was born prematurely. At 4 months of age she was diagnosed with hydrocephalus and had undergone shunt placement in the right lateral ventricle with 2 shunt revisions over the next 2 months. Neither the patient nor her family recalls history of congenital abnormalities or prior intracranial mass including hematoma, in the last 20 years. Clinically she now has poor memory retention and functions at about the level of a 9th-10th grade student. On annual physical examination she had dysmetria, dysrhythmia, positive Romberg’s sign, intermittent dysconjugate gaze, dystonia of the left hand and intermittent bilateral hemisensory inattention. MRI was obtained to evaluate the current status of the brain.

Methods

MRI of the brain was performed on a 3T Siemens Spectra scanner [Siemens Medical Systems, Malvern, Pennsylvania, USA] using a standard head coil. Standard 4 mm thick Axial T1, Axial T2, Fat Suppressed Axial T2 FLAIR, Axial DWI and ADC brain pulse sequences were acquired with 2.5mm thick Coronal spin echo T2 images. In the two cases we are presenting, bilateral uncus in patient 1 and right uncus, right parahippocampal gyrus [PH] in patient 2, are clearly overhanging the tentorial edge and touching the cerebral peduncle, and fulfilling the generally accepted criteria for true herniation.

Imaging Findings

Patient 1:

MRI of the brain [Figures 1A, 1B, 2A and 2B] showed bilateral uncus herniation, right greater than left, with normal signal of the uncus. The right uncus extended closer to the nearby right oculomotor nerve and the right cerebral peduncle of the midbrain. As there was no other intracranial abnormality to cause the herniation, a diagnosis of idiopathic bilateral uncus herniation was made. No prior imaging was available.

Patient 2:

MRI of the brain [Figures 3, 4A and 4B] showed prominent medial herniation of the right uncus and right PH, with normal signal. It caused mild mass effect on the nearby right cerebral peduncle. T2 FLAIR image [Figure 4A] also demonstrates mild medial left uncus displacement, with normal signal. There was a small dural based right frontal smoothly margined calcified mass which abutted the right side of superior sagittal sinus without narrowing it. This mass is presumed to be a calcified old subdural hematoma probably related to prior shunt revisions as an infant. Other images, not shown here, demonstrate a right frontal burr hole adjacent to the anterior edge of the calcified hematoma. The calcified hematoma is chronic with only minor local mass effect on the immediately adjacent superior right frontal gyrus. There are no other associated acute lesions with edema or mass effect to contribute to the visualized uncus and parahippocampal herniation, confirming their chronic nature. Given the mild left uncus medial displacement, the possibility of incidental initial presence of mild medial displacement of the right uncus, which eventually worsened due to ipsilateral right dural based lesion like hematoma, cannot be excluded. There are no prior comparison imaging studies.
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**Definition**

The extension of brain beyond its dural confines is called brain herniation. Herniations can be intracranial or extracranial. There are three basic types of intracranial herniations. They are subfalcine herniation, transtentorial herniation and cerebellar tonsillar herniation. Transtentorial herniation has two subtypes. They are the ascending [superior vermian] and descending transtentorial herniations [4]. Second most common cerebral herniation is unilateral descending transtentorial herniation [5]. The descending transtentorial herniations have two types, the central and lateral. In the central descending transtentorial herniation, the diencephalon, midbrain and pons descend through the tentorial incisura. In the lateral descending transtentorial herniation [LDTH], the medial temporal lobe descends through the tentorial incisura. The LDTH has two further subtypes. They are the anterior LDTH with the uncus descending down the tentorial incisura into the ipsilateral cranial cistern and the posterior LDTH with the parahippocampal gyrus descending down the posterolateral part of the tentorial incisura [6]. The various types of herniations can be acquired or idiopathic. While varying types of acquired herniations are commonly seen [7], Idiopathic brain herniations are rare.

**Etiology**

Brain is protected within the rigid skull and is cushioned by CSF. The Monro-kellie hypothesis postulates that the total volume of brain, intracranial blood and intracranial CSF is constant in an intact skull. If there is increase in one component, then there will be decrease in one or both of the remaining two components [5, 8]. When there is superimposed space occupying mass or other edematous lesions within the already tightly packed skull, the soft brain parenchyma is secondarily forced to traverse from one compartment to another [4, 9]. Depending on the location of the inciting lesion, nearby secondary brain herniation is seen.

Medial temporal lobe [uncal and PH] transtentorial descending herniations can be secondary or rarely idiopathic [1, 3]. They can be acute or rarely chronic [2, 10]. Secondary causes of uncal and PH herniations are related to space occupying supratentorial lesions like tumor, trauma, infection, infarction, hemorrhage, etc. Some of the tumors of the temporal region are astrocytoma, meningioma, ganglioglioma, hamartoma, neurilemoma, epidermoid and dermoid. The most common infection in the medial temporal lobe is due to Herpes Simplex Virus. Subdural, epidural and parenchymal hematomas can also cause LDTH based on their location and size. All these lesions increase supratentorial pressure and when it crosses a certain threshold, it results in herniation. Chronic secondary LDTH could be caused by associated trapping or adhesion of the herniated structure. It was postulated in chronic LDTH occurring in children with Dandy walker syndrome or following cistoperitoneal shunting of the large posterior fossa cyst that communicates with fourth ventricle, that negative pressure gradient in the posterior fossa relating to placement of shunts and shunt malfunction, can cause downward medial temporal lobe herniation. Wider incisura, high tentorial position and smaller cerebellum can be predisposing factors [2, 10]. We suspect that the angle of the tentorium cerebelli with respect to the tentorial incisura, along with a wide tentorial incisura could be predisposing factors for idiopathic uncal herniation.

**Clinical Presentation**

Idiopathic medial temporal lobe herniation has been found incidentally without signs or symptoms associated with
Chronic medial temporal lobe herniation can result from residual herniation following improvement of a significant space occupying lesion, that caused it. Chronic medial temporal lobe herniation has been reported to be associated with poor prognosis [2]. Significant uncinal herniation compresses the nearby midbrain, sylvian aqueduct, posterior cerebral and anterior choroidal arteries, and third cranial nerve [oculomotor], against the free edge of the tentorium [9]. Compression of the ipsilateral cerebral peduncle causes contralateral hemiparesis. When the sylvian aqueduct is compressed, it can result in hydrocephalus. Compression of posterior cerebral and choroidal arteries result in ischemia and infarction of ipsilateral occipital and medial temporal lobes. Third cranial nerve compression results in oculomotor nerve palsy with ipsilateral pupillary dilation. Oculomotor nerve palsy is usually accompanied by decreased level of consciousness due to interruption of the Ascending Reticular Activating System (ARAS) [11], but a case has also been reported without decrease in level of consciousness [12]. Uncal herniation through the transtentorial incisura can occasionally result in shift of mesencephalon to the other side with contralateral cerebral peduncle impingement on the contralateral tentorium cerebelle. This is associated with false localizing ipsilateral hemiparesis, known as the Kernohan-Woltman notch phenomenon [KWNPN] [13]. This phenomenon is associated with a poor prognosis [14]. Pressure necrosis of the medial temporal lobe can occur with severe unilateral or bilateral descending temporal lobe herniation [5]. Severe bilateral hemiparesis, coma, and even death can result from severe medial temporal lobe herniation [1].

Theoretically, acute herniation can superimpose on pre-existing idiopathic or chronic herniation and can cause acute catastrophe. Hence awareness and adequate reporting of the herniation findings are essential. Symptomatic patients require closer surveillance and should be treated promptly [10].

**Radiological Findings**

**CT:** There is medial displacement of the uncus and PH gyrus. It has the same density as the normal brain parenchyma. Mass effect on the adjacent midbrain is based on the adjacent herniation. Prominent herniations are better seen than smaller ones on CT. There is no abnormal enhancement.

**MRI:** MRI is more sensitive and specific than CT. There is medial displacement of the uncus, with or without the PH gyrus. They have normal signal characteristics on all sequences. There is no abnormal enhancement with contrast. Based on the degree of herniation, there will be mass effect on the adjacent ipsilateral cerebral peduncle of midbrain, sylvian aqueduct, posterior cerebral and anterior choroidal arteries, and third cranial nerve [oculomotor], with possible compression against the free edge of the tentorium. If there is significant displacement of the mesencephalon to the opposite side, the contralateral cerebral peduncle will impinge on the contralateral tentorium cerebelle. This can cause signal changes in the contralateral cerebral peduncle.

In the general population, coronal imaging occasionally reveals small degrees of uncinal extension beyond the edge of the tentorium, which historically has been considered to be within normal limits given the lack of corresponding clinical manifestation.

**Similar cases in literature**

Very few cases of Idiopathic uncinal herniation have been reported in literature so far, to our knowledge. The salient articles are the following. Initially Horowitz et al, reported a misinterpretation of parahippocampal herniation for a posterior fossa tumor with subsequent surgery [1]. Yavarian et al, then published a case report on herniation of uncus and parahippocampal gyrus as an accidental finding on MRI of cerebrum [3]. Idiopathic herniations of left inferior temporal gyrus and precuneal gyrus [15], and idiopathic herniation of cuneus through focal dural defect [16], have also been reported in literature.

Chronic uncinal herniation has been discussed in 2 infants with Dandy-Walker malformations by Naidich et al. [12]. A case report on chronic uncinal herniation secondary to posterior fossa shunting on a 12 year old, has been reported by Udayakumaran et al. [10].

Our cases clearly represent idiopathic and chronic herniation with high specificity, as detailed in patient imaging findings.

**Conclusion**

Findings of idiopathic LDTH are pathognomonic on MRI. If there is suspicion for temporal lobe tumor, sometimes radiologic imaging alone cannot distinguish benign from malignant processes. In those instances, biopsy and surgery might be needed. Correct identification of idiopathic uncinal and PH herniations by both the radiologist and neurosurgeon, are important to avoid unnecessary biopsy [1]. Given the advanced imaging techniques today, it’s easy to identify this entity. Chronic herniations have to be identified along with residual primarily pathology. In the presence of idiopathic and chronic descending transtentorial temporal lobe herniations, prompt surveillance is needed with new or worsening symptoms to prevent catastrophic consequences.

**Conflict of Interest**

None.

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None.

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