Brain herniation with surrounding CSF into the skull

TO THE EDITOR: I read the article by Valci et al.7 with great interest (Valci L, Dalolio M, Kuhlen D, et al: Intradiploic encephalocele of the primary motor cortex in an adult patient: electrophysiological implications during surgery. J Neurosurg [epub ahead of print April 28, 2017. DOI: 10.3171/2016.11.JNS162426]). The authors reported a case of spontaneous right frontal lobe parenchyma herniation with surrounding CSF into the right parietal bone in a 70-year-old man who was experiencing a spastic progressive paresis of his left lower limb. The authors described this entity as a spontaneous intradiploic encephalocele.7 As I have a special interest in herniations of brain parenchyma into calvaria and/or dural venous sinuses (DVS),1–3 I would like to contribute regarding naming, imaging, and clinical features of this recently recognized entity to prevent confusion that may occur in readers.

In their paper, Valci et al.7 reported that there was a bone defect on the right parietal bone with herniation of right precentral gyrus parenchyma through this osseous defect into the intradiploic area, without extension to the outside of the calvaria. Consequently, the authors noted that this entity was consistent with a spontaneous intradiploic encephalocele. However, in the relevant case there was lysis of the internal table and extension into the diploic region of the parietal bone, and only thinning of the external table without any total defect as seen on the presented MR and CT images and mentioned by the authors in the operation section of the case report.7 Herniations of brain parenchyma with surrounding CSF into the calvaria or DVS without apparent complete bone defects in the adjacent skull were recently described on MRI and CT images and mentioned by the authors in the operation section of the case report.7 Initially the descriptions, possible etiologies, and clinical significances of this entity were determined differently in various reports. Chan et al.4 described this entity as focal cerebellar hemisphere herniation into a giant arachnoid granulation located in occipital bone. Çoban et al.,5 in a similar case of left temporal lobe parenchyma herniation (surrounded with CSF) into the left transverse sinus without an apparent bone defect, described it as an occult temporal lobe encephalocele. Battal and Castillo1 reported 5 cases in their report and described this entity as brain parenchyma herniation with the surrounding CSF into the calvaria or DVS. They reported that this entity was probably an incidental finding that might be more common than previously recognized.1 This entity was described as brain herniation with surrounding CSF into the calvaria or DVS in the most comprehensive research about this topic reported by Battal et al.3 I believe that brain herniations into the skull have different features from classic encephaloceles. Central skull base encephaloceles remain occult unless there is a CSF leak, meningitis, seizure, or headache. Encephaloceles are documented as masses composed of meninges and brain extruding outside the skull.1–3 Conversely, the brain herniation that was described by Valci et al.7 did not occur through a complete calvarial defect. Instead, it occurred through a dural defect into calvaria and thus does not completely meet the criteria for encephaloceles. Moreover, as mentioned by the authors in the discussion section, the herniated brain parenchyma was not covered by the dura, and there was not a history of trauma, infection, or previous cranial operations as an underlying cause for acquired encephaloceles. Because of these reasons, this entity should be described as brain herniation with surrounding CSF into the calvaria instead of an encephalocele.

Arachnoid granulations are protrusions of arachnoid through the dura mater. A neck of arachnoid penetrates an aperture in the dura and expands to form the core of the granulation. Arachnoid granulations may enlarge with age or in response to increased CSF pressure.1–3 Although the features of brain parenchyma herniations into calvaria are also different from classic arachnoid granulations and arachnoid pits, it appears reasonable to postulate that the presence of preexisting arachnoid granulations or pits may facilitate the formation of brain herniation into the adjacent calvaria or DVS.1–3 Therefore, I believe these structures are not covered by the dura, as mentioned by Valci et al.7, and are a different entity from the classic acquired encephalocele.

Battal et al.3 reported that brain parenchyma herniations with surrounding CSF into the calvaria and/or DVS are more common than previously assumed, with a prevalence rate of 0.32%. These herniations are encountered more frequently in posterior-inferior parts of the intracranial cavity and the most common locations were the transverse sinus and occipital bones.3 Recently, a very similar case was reported by Rodrigues and Santos in the parasagittal convexity area. They presented a case of parietal lobe parenchyma herniation with surrounding CSF into the incomplete parietal bone defect, with a narrow neck and atrophy, partial strangulation, and hyperintensity of...
herniated brain parenchyma on MR images. Consequently, brain herniation into the calvaria has been reported to be very rare and its etiology, symptomatology, and clinical significance are not well-known. I believe that this entity is more common than expected and should be considered in the differential diagnosis of encephaloceles and lytic skull lesions. The continuity of the external table of the calvaria (although it may be very thin) and no brain tissue extruding outside the skull can be distinguishing features of this entity from encephaloceles.

Bilal Battal, MD
Private İzmiryolu Sevgi Hospital, Balıkesir, Turkey

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Disclosures

The author reports no conflict of interest.

Response

We read with interest the letter to the editor from Dr. Battal and are happy to provide an extensive response. The details provided by Dr. Battal certainly help to define the characteristics of anatomical “peculiarities,” which are hardly classifiable because of their rarity. Battal also suggests interesting ideas about the mechanisms of their formation.

Encephaloceles can present in various ways and, as noted in our article, they are not well classified yet. In the literature there are many classifications according to distinct aspects of the encephalocele, such as content, location, and pathogenesis. The extradiploic encephalocele—when brain, meninges, and CSF protrude out of the skull, whether occult or not—is an intuitive and defined entity. In contrast, the intradiploic encephalocele, which is a herniation into the calvaria through an incomplete bone defect with or without dural defect, remains complex and difficult to classify. Nevertheless, we believe that our case should be defined as an encephalocele for two main reasons.

First, the etymology of the term itself helps. The term “encephalocele” comes from ancient Greek, “enkefálē” and “kele,” meaning brain herniation. A brain herniation is a displacement of cerebral mass, with or without CSF and meninges, from the usual position to a different one, in most cases through a narrow passage (i.e., tonsillar, uncal, or transtentorial herniation). So, according to the actual classification and in our opinion, the encephalocele is a type of brain herniation in which brain protrudes out of an osseous defect within or outside the diploe. We can argue that in the case of brain herniation into the calvaria (so called intradiploic), the partial osseous defect plays an effective role in pathogenesis. The brain herniation in DVS, as described by Battal et al., appears to be a different entity in which the osseous defect has no role in its pathogenesis.

Second, the reason why we did not find any dura layer covering the herniated brain could correlate with the presence of meningioma discovered during pathological examination of the bone margins. The tumor, therefore, could have induced a reabsorption of the dura layers. An acquired encephalocele can be the consequence of trauma, infection, or previous cranial operations, but also because of tumors. In the literature, another case of an intradiploic encephalocele not covered by the dura layer has been reported. In that case, the patient experienced a previous trauma that over time induced the protrusion of the brain parenchyma through an osseous-dural defect.

In conclusion, the main focus of our article was to emphasize that the herniated parenchyma was functional. We also supposed that the meningioma caused a progressive reabsorption of both bone and dura, inducing the parenchyma herniation by intermittent physiological pulsation of the brain. Despite the long duration of this process, a reorganization of the primary motor area, with exclusion of the herniated brain parenchyma, did not occur. This is certainly rare, considering the cases of encephalocele already published, as we discuss in our article. Beyond definitions and classifications, the clinical features, involvement of the primary motor area in the herniated parenchyma, and possibility of mapping it and thus preserving it, represent the peculiar and surprising aspects of our case.

Luca Valci, MD
Marina Dalolio, MD
Dominique Kuhlen, MD
Emanuele Pravatá, MD
Claudio Gobbi, MD
Michael Reinert, MD
Neurocentro della Svizzera Italiana, Ospedale Regionale di Lugano, Switzerland

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