JOURNAL OF NEUROSURGERY:
Case Lessons

J Neurosurg Case Lessons 3(2): CASE21565, 2022
DOI: 10.3171/CASE21565

A red herring CVA with unexpected outcome: illustrative case

Charles Swanson, DO,[1] Raju Z. Abraham, MD,[2] Michael Ruebhausen, MD,[3] and Juan Jimenez, MD[4]

Departments of [1]Internal Medicine, Graduate Medical Education, [2]Pulmonary and Critical Care Medicine, [3]Plastic and Reconstructive Surgery, and [4]Neurosurgery, Riverside Medical Center, Kankakee, Illinois

BACKGROUND Disruptions to the integrity of the inner table and trabeculae of the calvaria are rare phenomena. Increasingly rare is the phenomenon of herniation of brain parenchyma through the defects in the skull causing neurological deficit. Surgical intervention is commonly performed but is fraught with risk of brain tissue loss.

OBSERVATIONS The authors present a case of a 78-year-old White male presenting with strokelike symptoms who was found to have an intradiploic encephalocele that was successfully treated with surgical intervention and neuroplastic reconstruction of the anatomical deficit. The patient had a marked recovery and had near-complete resolution of symptoms.

LESSONS This notably rare phenomenon resolved with neurosurgical intervention, sparing the parenchyma, and provided the patient with perceivably normal contour of the head using a collaborative approach with neuroplastic intervention.

https://thejns.org/doi/abs/10.3171/CASE21565

KEYWORDS intradiploic encephalocele; neuroplastics; craniectomy

The purpose of this report is to describe a case of a 78-year-old White right-handed male who experienced symptoms synonymous with a cerebrovascular accident and was found to have an intradiploic encephalocele that was treated with neurosurgical intervention and neuroplastic reconstruction of the skull. After undergoing surgery with both techniques, the patient had a full recovery and is free of visible defect to the afflicted side of his head.

Illustrative Case

Preoperative

A 78-year-old right-handed White male with bronchiectasis and pulmonary fibrosis, bronchial artery hemorrhage status post–bronchial artery embolization in the past, and benign prostatic hypertrophy presented as a code cerebrovascular accident with right-handed weakness and minor dysarthria. The patient had been experiencing weakness for approximately 3 weeks preceding presentation with a sudden worsening in his speech and right distal forearm and hand weakness. He had a right-sided facial droop, was dysarthric, and exhibited right pronator droop and right hand weakness. The patient underwent computed tomography (CT) (Fig. 1) of the head without contrast, revealing an expansile lytic bone lesion involving the left frontal bone measuring 4.6 × 1.3 × 4.7 cm. Herniation of the adjacent left frontal cortex was seen through a defect in the inner table on initial CT. Neurosurgery was consulted, and a frontotemporoparietal craniotomy was scheduled.

Operation

The patient was taken to the operative theater. Preoperative magnetic resonance imaging (MRI) of the brain (Fig. 2) was loaded to the Brainlab curve navigation system. Three-dimensional computer models of the patient's skull lesion were developed. The patient was placed in a DORO three-point head holder. Then, using Airo intraoperative CT, a spin was performed. The position of the skull lesion was identified. The lesion was noted to be directly behind the left frontoparietal convexity. Merging was performed with the preoperative MRI with an excellent overlay. Next, a free flap craniectomy was planned with the navigation system. There was visible thinning of the outer table of the skull to a translucent degree. The cystic component of the lesion was outlined. There

ABBREVIATIONS CSF = cerebrospinal fluid; CT = computed tomography; ICU = intensive care unit; MRI = magnetic resonance imaging.

INCLUDE WHEN CITING Published January 10, 2022; DOI: 10.3171/CASE21565.
SUBMITTED October 1, 2021. **ACCEPTED** October 19, 2021.

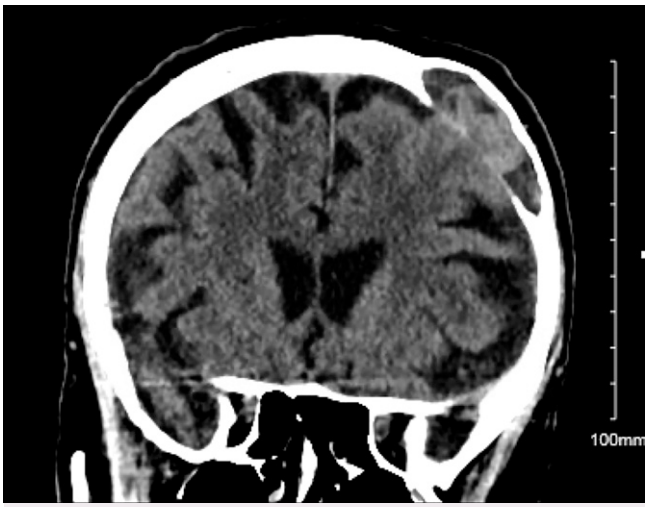

FIG. 1. CT without contrast showing cranial deficit and lesion.

was elevation of the outer table. Once the margins were determined, an area of cyst without herniated brain was selected to enter the diploe. The outer table was paper thin. Once a window into the diploe was created, the herniated cortex was visualized. The outer table was drilled to establish the full extension of the herniated brain. The exposed brain was continuously irrigated. It was evident that the inner table and dura were calcified. Dissection was performed, and a wider margin of resection was needed. A matchstick burr was used to establish a margin to normal skull surrounding the cystic skull lesion. The craniectomy was performed in a freehand manner to ensure no dural violation. A dural margin was established. There was evidence of hyperostosis, likely reactive.

Once a full circumferential margin was established, it was possible to remove the outer table. There was dural erosion with a defect at the base of the herniated brain. There was pallor of the cortex that was herniated. Meticulous dissection of the dura was performed, the calcified layer around the herniated brain was outlined to allow safe removal of the inner table to access the lesion, and the herniated brain with normal dura was established. An inner and outer margin of normal dura was established. Drilling was performed

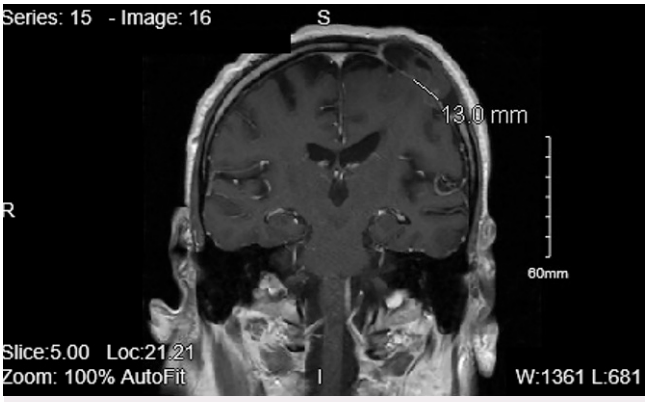

FIG. 2. MRI of the brain with and without contrast showing cranial deficit and lesion, further depicting herniation.

between normal inner and outer dural margins, and this allowed further space to dissect the dura from the inner table and calcified margin above the dura. A hook microdissector and a 1-mm Kerrison rongeur were used to allow the piecemeal resection of the inner table complex. The free flap craniotomy was disconnected off the dura in several fragments. All bone fragments and the inner cystic wall were submitted for pathological analysis. Next, the dura was inspected and found to be intact around the herniated brain (Fig. 3). The dura was seen to be relatively slack. The dura was opened widely in cruciate fashion and was reflected, and fragments were removed from around the herniated brain. There was an inner rim of dura around the herniated brain. The size of the original dural defect was small. The microscope was brought into the field, and, with microsurgical technique, the herniated brain was dissected off the dural ring. Bipolar was used for hemostasis, and the inner rim of the dura was reached. This allowed a circumferential release of the dural ring. The dura was resected and reflected in piecemeal fashion. All dural fragments were submitted for pathological analysis. Once the dural rim was removed, the herniated brain was less taut.

A sulcus was identified, and dissection of the arachnoid allowed the safe egress of cerebrospinal fluid (CSF), which further relaxed the herniated brain. Electrostimulation was performed at this time. The herniated brain was stimulated, and activity was obtained at 20 mA. The cortex around the herniated brain was stimulated at 4 mA inferior and lateral to the brain. This indicated primary motor cortex. At this point, due to the identification of motor cortex and the dense number of cortical vessels involved with the thickened arachnoid, no further dissection was pursued due to the risk of devascularization of the exposed primary motor cortex. It was believed that due to the wide decompression and improved perfusion of the herniated brain, the edema would subside over time and the herniated cortex would naturally regress. The

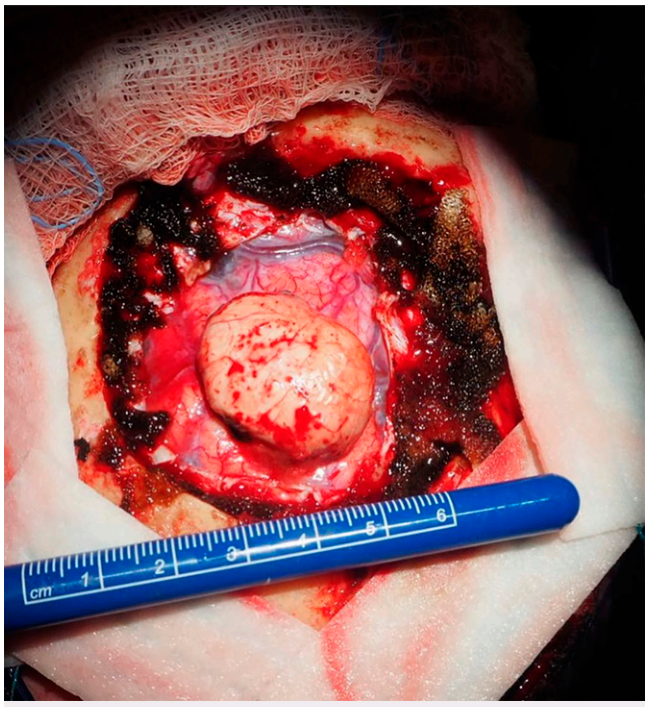

FIG. 3. Cerebral cortex herniating through a dural deficit after the outer table was removed.

brain was lax, as was the encephalocele, and, due to the presence of stimulation, the brain stump was left intact.

At this point, additional irrigation was applied. Meticulous hemostasis was secured with topical hemostatic agents. No significant contusion or swelling of the brain was noted. The dura was then closed using a 3 × 3–inch Duragen onlay dural graft. Stryker green dural sealant was applied to secure a watertight closure. At this point, the case was turned over to the plastic surgery service. Due to the brain relaxation and relaxation of the herniated brain, we agreed that proceeding with closure of the skull defect was safe.

Plastic surgery then undertook reconstruction using a prebent Stryker gold titanium mesh with 0.6-mm thickness to template the defect (Fig. 4). This was then cut to size and bent with the inflection Stryker plate bender in order to create more of a dome over the top of the defect so that there was no pressure placed on the part of the brain that was herniating. The edges of this plate were then carefully cut around and then contoured to the edges of the bone. Sharp edges were removed as much as possible in order to make them flush and smooth. This was then secured in place using self-drilling, self-tapping screws monocortically. The implant was smoothed with direct injection of on-demand hydroxyapatite cement.

A 10-French Blake drain was then placed inferiorly, and the wound was closed stepwise in the usual fashion, first with the galea by 3-0 Vicryl suture in a figure-of-eight fashion, followed by the skin with 3-0 Vicryl suture in a deep dermal fashion for the entirety of the horseshoe-shaped incision extending from just posterior to the superficial temporal artery, posterior to the occiput, and then anterior just to the lateral portion of the midline. The skin edges were then closed using 3-0 nylon suture in a horizontal mattress fashion.

The incision line was anesthetized with Marcaine, and the wound was dressed with Xeroform, bacitracin, and Telfa over the incision. Unfolded fluffs were then placed globally over the top of the head to provide some level of compression to avoid a potential space for serous or sanguineous fluid to accumulate. This was then wrapped with a 4-inch Kerlix roll at this time. Neurosurgery then removed the bolts from the halo. The patient was then wrapped with a 6-inch stockinette that was placed over the top of the face, and a hole was cut for the face so that he had a chin strap. The drain was then brought out of the end of this. This was turned 180°, and a second overlap was then placed over the top of this, tucking the drain into the area between the two folded-over stockinettes. The patient was then sent to the intensive care unit (ICU) for recovery.

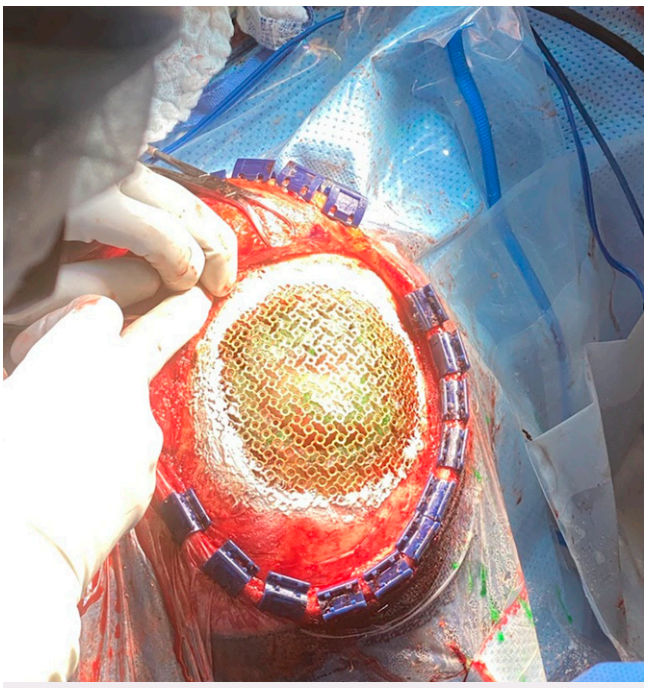

FIG. 4. Neuroplastic reconstruction of the deficit after gold titanium mesh was in place.

Postoperative Course

The patient was transferred to the ICU. He was successfully extubated but had persistent right upper extremity deficits initially. He was treated with intensive therapy services, including physical, speech, and occupational therapies. He had repeat head CT scans (Fig. 5), which indicated the reduction in the encephalocele size that corresponded with reduction in size of the lesion. After 6 months, the patient regained full strength of the right upper extremity, and his speech problems fully resolved. The patient is currently asymptomatic without any appreciable neurological deficits.

Discussion

Observations

Intradiploic encephaloceles are relatively rare phenomena that predominately occur in children and few adults.[1] An encephalocele is the herniation of cerebral contents through the dura or skull.[1] Craniomeningoceles contain leptomeninges and CSF but not parenchyma. They are typically described by the area in which they present: posterior fossa, occipital, basal, and frontoethmoidal and/or parietal regions.[1–4] Causes of such defects are either congenital or usually due to infection or trauma. Primary encephaloceles are described as inherited imperfections in the meninges of the skull precipitating the erosion and herniation. This is caused by neural tube defects that occur during development hypothesized to be due to failure of these structures to close during neurulation.[5–7] Commonly, the defects occur along a cranial suture. Secondary encephaloceles occur more commonly in adulthood and are precipitated by infections, trauma, tumor, or surgery. Classification of such

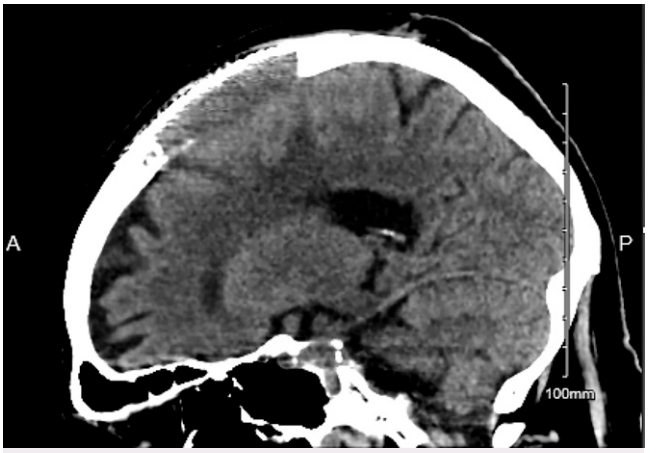

FIG. 5. Postoperative CT of the head after release of dura and reconstruction.

lesions can be made by either the Matson and Ingraham or Nager classification.[5] According to the Matson and Ingraham classification, defects are characterized by four main groups: basal, sincipital, convexity, and atretic. The Nager classification is more complex in terms of the number of metrics.[5]

Typical presentations of patients with encephaloceles include palsies, seizures, CSF leakage, and meningitis.[5,8–11] Neurological deficits are relatively uncommon, and those that have been cited include hearing loss, visual deficits, facial nerve palsy, and hemiparesis. This case was unique in that our patient was older and experienced only neurological deficits with the expressive aphasia and right upper extremity deficits. The patient also did not have known congenital, traumatic, or infectious etiologies or prior neurosurgery. The patient notes that he was aware of a growing "bump" on the left side of his skull for several years but was unable to characterize the duration. The standard of care would have been to resect the affected area of the cerebral cortex because it has a propensity for recurrence, but in this case, the lesion consisted of the primary motor cortex and was unable to be resected.

Lessons

A patient presented with an intradiploic encephalocele with right-sided upper extremity deficits and expressive aphasia. This rare lesion was thoroughly investigated with CT and MRI, resulting in the optimal treatment plan being surgical exploration and repair. The primary motor cortex was identified intraoperatively. After surgical intervention, the lesion was noted to decrease in size, and the patient's symptoms abated. This case is rare in the character of the presentation of the lesion and in the frequency of this anatomical abnormality in the literature.

References

1. Shi C, Flores B, Fisher S, Barnett SL. Symptomatic parietal intradiploic encephalocele—a case report and literature review. *J Neurol Surg Rep.* 2017;78(1):e43–e48.
2. David DJ. Cephaloceles: classification, pathology, and management – a review. *J Craniofac Surg.* 1993;4(4):192–202.
3. David DJ, Proudman TW. Cephaloceles: classification, pathology, and management. *World J Surg.* 1989;13(4):349–357.
4. Suwanwela C, Suwanwela N. A morphological classification of sincipital encephalomeningoceles. *J Neurosurg.* 1972;36(2):201–211.
5. Valci L, Dalolio M, Kuhlen D, Pravatà E, Gobbi C, Reinert M. Intradiploic encephalocele of the primary motor cortex in an adult patient: electrophysiological implications during surgery. *J Neurosurg.* 2018;128(3):871–874.
6. Dias MS, Partington M. Embryology of myelomeningocele and anencephaly. *Neurosurg Focus.* 2004;16(2):E1.
7. Ertas B, Aksoy EA, Unal OF. Nasopharyngeal mass diagnosed as transsphenoidal encephalocele in an adult patient. *J Craniofac Surg.* 2015;26(8):e793–e794.
8. Agladioglu K, Ardic FN, Tumkaya F, Bir F. MRI and CT imaging of an intrasphenoidal encephalocele: a case report. *Pol J Radiol.* 2014;79:360–362.
9. Harada N, Nemoto M, Miyazaki C, et al. Basal encephalocele in an adult patient presenting with minor anomalies: a case report. *J Med Case Rep.* 2014;8(1):24.
10. Jain A, Tullu MS, Agrawal M, Jadhav DU. Occult encephalocele causing recurrent meningitis. *Pediatr Neurol.* 2015;53(3):270–271.
11. Zoli M, Farneti P, Ghirelli M, et al. Meningocele and meningoencephalocele of the lateral wall of sphenoidal sinus: the role of the endoscopic endonasal surgery. *World Neurosurg.* 2016;87:91–97.

Disclosures

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author Contributions

Conception and design: Ruebhausen, Jimenez. Acquisition of data: Ruebhausen, Jimenez. Drafting the article: Swanson, Abraham. Critically revising the article: all authors. Reviewed submitted version of manuscript: Abraham, Ruebhausen. Approved the final version of the manuscript on behalf of all authors: Swanson. Administrative/technical/material support: Ruebhausen.

Correspondence

Charles Swanson: Riverside Medical Center, Kankakee, IL. cswanson@rhc.net.