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Case Report

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Epidural Hematoma Following Ventriculoperitoneal Shunt Revision in Open-Lip Schizencephaly; Case Report.

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ABSTRACT
Schizencephaly is a rare congenital neuronal migration disorder characterized by clefts in the cerebral hemispheres. While hydrocephalus is not a universal feature, it can occur, particularly in open-lip schizencephaly, necessitating ventriculoperitoneal (VP) shunt placement. Epidural hematoma (EDH) is an uncommon but serious complication following VP shunt surgery, especially revision procedures. We present the case of a 4-year-old girl with bilateral open-lip schizencephaly and a history of VP shunt placement who developed a large EDH requiring surgical evacuation following a shunt revision. This report highlights the diagnostic and management challenges in such complex cases and discusses the potential mechanisms of post-shunt EDH formation.

Keywords: Schizencephaly, Open-Lip Schizencephaly, Hydrocephalus, Ventriculoperitoneal Shunt, Epidural Hematoma, Shunt Revision

Introduction

Schizencephaly is an infrequent congenital malformation of the brain. It is defined by the presence of full-thickness, gray matter-lined clefts extending from the pial surface to the lateral ventricles [1]. Schizencephaly is broadly classified into two types: type I (closed-lip), where the cleft walls are in apposition, and type II (open-lip), where the cleft walls are separated, creating a cavity filled with cerebrospinal fluid (CSF) that communicates freely with the ventricular system and subarachnoid space [1]. Clinical manifestations are variable and depend on the type, location, and extent of the clefts, ranging from asymptomatic individuals to those with severe neurological deficits, including seizures, motor impairments, and developmental delay.

Hydrocephalus can be associated with schizencephaly, particularly the open-lip variant [1,2]. The management of hydrocephalus in these patients often involves the insertion of a ventriculoperitoneal (VP) shunt. While VP shunting is a common neurosurgical procedure, it is not without complications, which include infection, obstruction, overdrainage, and, rarely, intracranial hematomas [3,4]. Epidural hematoma (EDH) formation following VP shunt surgery is an unusual occurrence, with an estimated incidence in some series being low but significant given the number of shunt procedures performed [5-7]. Various proposed mechanisms include rapid reduction in intracranial pressure (ICP) leading to dural stripping from the inner table of the skull, particularly in the context of intial shunt insertion or overdrainage, leading to dural and diploic veins to bleed into the epidural space [6-8]. The dural arteries may also tear as the hematoma enlarges and the distance between the dura and the bony arterial channels increases [8].

We report a challenging case of a 4-year-old girl with known bilateral open-lip schizencephaly who developed a significant acute EDH requiring emergency craniotomy and evacuation after a VP shunt revision. This case underscores the importance of vigilance for rare but life-threatening complications in this patient population and explores the potential pathophysiological mechanisms involved.

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Case Presentation

A 4-year-old girl with a known medical history significant for bilateral open-lip schizencephaly (right greater than left), absent corpus callosum, and global developmental delay presented to the emergency department. She had a VP shunt inserted 2 years ago for hydrocephalus. Her other comorbidities included seizures (controlled on levetiracetam), severe physical and mental disability, left-sided spastic hemiplegic cerebral palsy, and strabismus.

She was brought to the ER with a one-day history of lethargy, hypoactivity, and multiple episodes of vomiting, coupled with decreased oral intake over the preceding three days. She had not had a bowel movement for 6 days. There were no other overt signs of raised intracranial pressure. On examination, she was sleepy but arousable, crying, and hypoactive. Her pupils were equal and reactive, and the shunt was compressible with good refilling. There were no signs of shunt infection. The remainder of her examination was consistent with her baseline neurological status. An initial CT scan of the brain showed stable ventricular size with no evidence of acute hydrocephalus or intracranial hemorrhage (Figure 1). A shunt series revealed moderate colonic fecal loading but no shunt disconnection or fracture (Figure 2). She was admitted with an initial impression of shunt malfunction secondary to constipation. General pediatrics team was consulted, and she was started on laxatives. She was also diagnosed with gingivostomatitis and commenced on antibiotics and antiviral medication. An ophthalmology review revealed no papilledema.

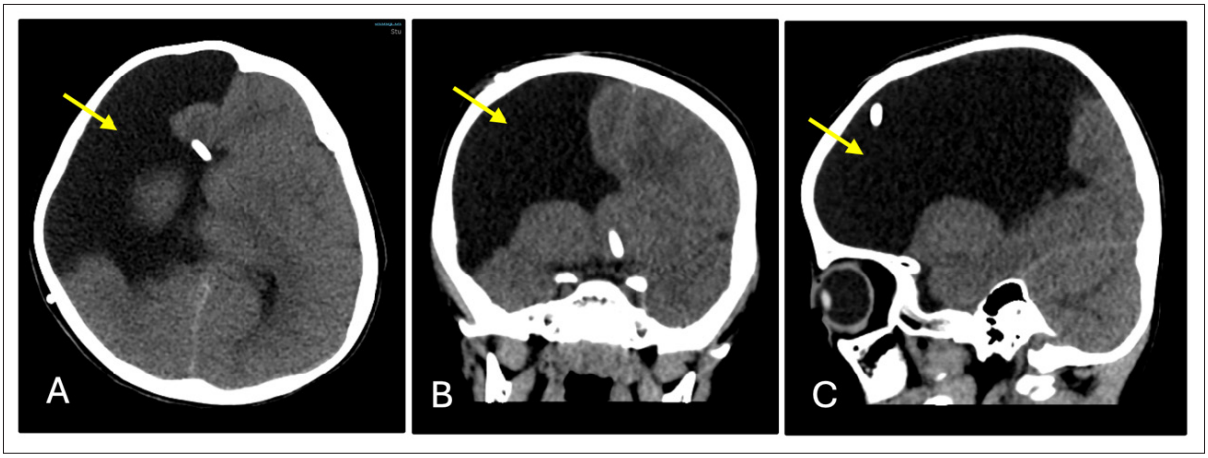

Figure1: Axial (A)/CDespite Supportive Careronal (B)/Sagittal (C) computed tomography (CT) scan of the brain demonstrates the right open-lip schizencephaly (yellow arrow) and corpus callosal dysgenesis, with a right frontal VP shunt in situ and no evidence of acute hydrocephalus or intracranial hemorrhage prior to the acute presentation.

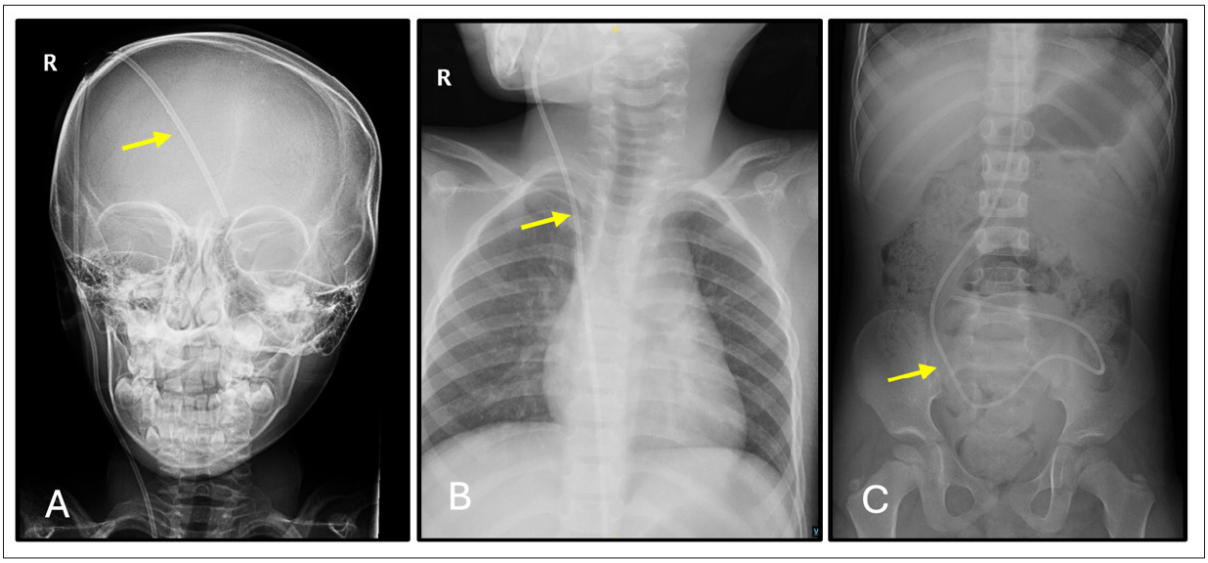

Figure 2: Shunt series radiograph (A-C) confirms the course of the right frontal approach VP shunt (yellow arrow) with the distal intra-abdominal tip projected over the right lower abdomen, with no evidence of kinking or discontinuation.

Despite supportive care, her Symptoms did not improve. Three days later, the patient developed sudden new neurological signs in form of right facial palsy and a left third nerve palsy. The neurology team was consulted to rule out CNS infection versus stroke. A CT scan with contrast was performed, which was unremarkable for any collection or signs of CNS infection, with stable ventricualr size (Figure 3). Concurrently, her shunt was noted to be refilling slowly. A repeat ophthalmology examination revealed Grade 1 papilledema.

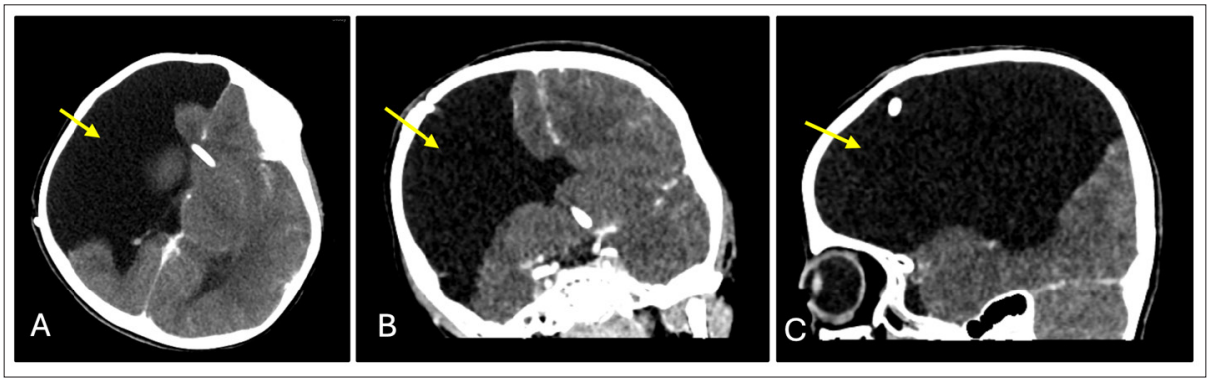

Figure 3: Axial (A)/Coronal (B)/Sagittal (C) computed tomography (CT) scan of the brain with contrast was unremarkable for acute intracranial collection or signs of CNS infection, with the VP shunt tip noted in the left frontal horn with no evidence of any changes from earlier scan (yellow arrow).

Given the clinical suspicion of shunt malfunction, she underwent an emergent VP shunt revision on same day. Intraoperatively, the proximal catheter was found to be nonfunctional and stuck, necessitating the insertion of a new proximal catheter. CSF was noted to be under high pressure upon insertion of the new catheter.

The following day the shunt revision, the patient was irritable and crying but maintained her level of consciousness. A postoperative brain CT scan revealed a large acute right frontoparieto-temporal EDH, measuring 5.5 x 10.5 cm, causing significant mass effect and a 0.3 cm midline shift to the left (Figure 4). Due to these findings, she underwent an emergency right craniotomy and hematoma evacuation on the same day. Intraoperatively, multiple small venous channels draining to the superior sagittal sinus were identified as the source of bleeding and were controlled. Tack-up sutures were placed, and multiple burrholes were created in the bone flap, with a Jackson-Pratt drain inserted upon closure.

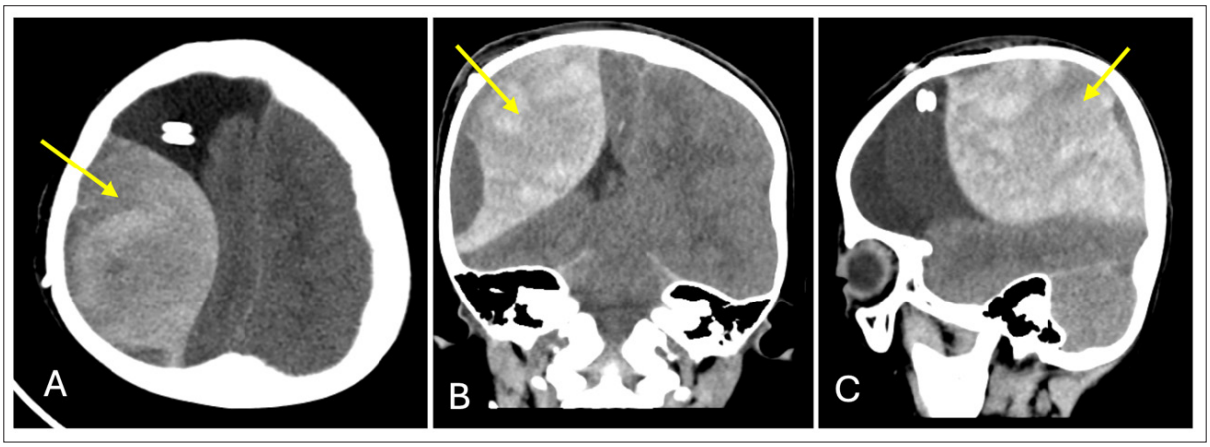

Figure 4: Axial (A)/Coronal (B)/Sagittal (C) computed tomography (CT) scan of the brain, following VP shunt revision, reveals a large acute right fronto-parieto-temporal epidural hematoma measuring 5.5 x 10.5 cm (yellow arrow) causing significant mass effect and a 0.3 cm leftward midline shift. A newly inserted shunt tip is noted.

The patient was managed in the Pediatric Intensive Care Unit (PICU) for neuromonitoring for a few days and was subsequently transferred to the floor in good condition. A follow-up CT scan showed postoperative changes consistent with the hematoma evacuation, with resolution of the midline shift (Figure 5). Upon discharge her home, she was awake, active, and playful, with residual left eye ptosis and right facial weakness, but had returned to her baseline activity level.

Discussion

Schizencephaly represents a complex spectrum of congenital brain malformations. The association of hydrocephalus with open-lip schizencephaly, as seen in our patient, while not ubiquitous, is a recognized phenomenon that often necessitates CSF diversion procedures like VP shunting [1,2]. The management of these patients is inherently challenging due to their underlying neurological comorbidities and altered intracranial anatomy.

The development of an EDH following VP shunt surgery is a rare but potentially devastating complication [5,6]. The incidence is low, and it is more commonly reported after initial shunt insertion rather than revision, although it can occur in both scenarios [8,9]. Several mechanisms have been proposed for post-shunt EDH formation. A leading hypothesis involves the rapid decrease in ICP

following effect CSF shunting [6,10,11]. This sudden pressure change can cause the dura mater to strip away from the inner table of the skull, potentially tearing bridging veins or dural vessels, leading to an epidural collection [8,10,12]. This is particularly relevant in cases of shunt malfunction followed by a rapid restoration of CSF drainage, as occurred in our patient after the revision of a non-functioning proximal catheter where CSF was found to be under high pressure.

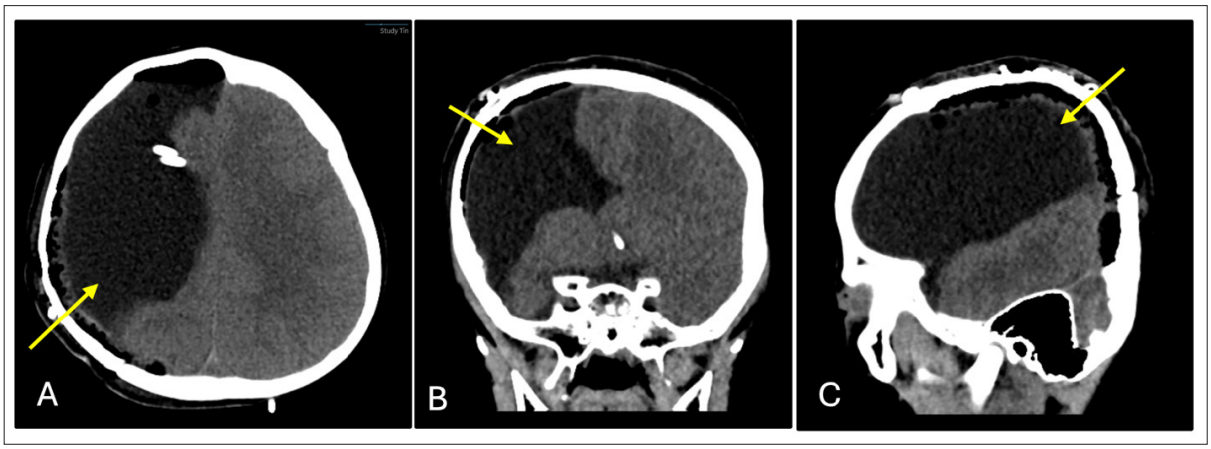

Figure 5: Axial (A)/Coronal (B)/Sagittal (C) computed tomography (CT) scan of the brain, status post right craniotomy and evacuation of the epidural hematoma (yellow arrow), with postoperative changes, including resolution of the midline shift.

In our patient, the EDH developed ipsilaterally to the newly inserted proximal catheter tip. The source of bleeding was identified intraoperatively as multiple small venous channels draining to the superior sagittal sinus. This finding supports the theory of venous bleeding due to dural stripping or tearing of these delicate vessels as the brain shifts or intracranial pressure dynamics change abruptly. The presence of pre-existing brain malformations like schizencephaly might also alter dural attachments or vascular anatomy, potentially predisposing to such complications, although this is speculative.

Other contributing factors to EDH formation can include direct trauma during surgery, or coagulopathy, which was not reported in our patient but has been documented as a risk factor in other cases [9,13]. The creation of burrholes during the initial shunt placement or revision could also, in theory, create a potential space for hematoma formation if dural attachments are disrupted, or if there is bleeding from bone edges or dural vessels injured during the procedure [14,15].

The clinical presentation of post-shunt EDH can be variable, ranging from asymptomatic or mild symptoms like headache to rapid neurological deterioration with signs of increased ICP and focal deficits, as witnessed in our patient with the development of irritability post-revision leading to the diagnostic CT scan [4,14]. Prompt recognition and neuroimaging are crucial for diagnosis. In this case, the postoperative CT scan was pivotal in identifying the life-threatening EDH, leading to timely surgical intervention.

The management of significant EDH is surgical evacuation to relieve mass effect and prevent secondary brain injury. The successful outcome in our patient, with a return to baseline activity despite residual focal deficits, underscores the importance of aggressive management.

This case also highlights the challenges in managing hydrocephalus requiring shunting in the context of open-lip schizencephaly [2]. While CSF pathways are often open to the ventricles, the dynamic balance of CSF production and absorption can still be disrupted, leading to symptomatic hydrocephalus. The decision to shunt, and subsequently revise a shunt, must always weigh the benefits against the potential risks, including rare complications like EDH.

Further literature review indicates that while EDH post-VP shunt is uncommon, it is a recognized entity [5-7]. Reports often emphasize the role of overdrainage or sudden ICP reduction [8,10-12]. The specific context of a shunt revision for a blocked catheter, leading to a sudden release of high-pressure CSF and subsequent EDH, aligns with these proposed mechanisms. The finding of venous oozing from channels draining to the superior sagittal sinus is also consistent with reports where venous sources are implicated.

Conclusion

Epidural hematoma following VP shunt revision is a rare but serious complication that requires a high index of suspicion, particularly in patients with complex underlying congenital brain malformations like open-lip schizencephaly. Rapid changes in intracranial pressure dynamics during shunt revision are a likely contributing factor. Prompt neuroimaging and emergent surgical intervention are critical for a favorable outcome. This case emphasizes the unique challenges in managing patients with schizencephaly and hydrocephalus and adds to the literature on rare but significant complications of VP shunt surgery.

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