

Case Report

Posterior reversible encephalopathy syndrome in a patient with a Chiari I malformation

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Abstract

Background: The authors describe a unique case of a patient who developed posterior reversible encephalopathy syndrome (PRES) following postoperative treatment of a Chiari I malformation.

Case Description: A 25-year-old female presented with complaints of left upper and lower extremity paresthesias and gait disturbances. A magnetic resonance imaging (MRI) of the brain and cervical spine showed a Chiari I malformation with tonsillar descent beyond the level of the C1 lamina. She underwent a suboccipital craniectomy and C1 laminectomy with cerebellar tonsillar cauterization and duraplasty. Postoperatively, an MRI showed bilateral acute infarcts of the cerebellar vermis. She was initially treated for cerebellar ischemia with hypertensive therapy with a subsequent decline in her neurologic status and generalized tonic-clonic seizure. Further workup showed evidence of PRES. After weaning pressors, the patient had a significant progressive improvement in her mental status.

Conclusion: Although the mechanism of PRES remains controversial given its diverse clinical presentation, several theories implicate hypertension and steroid use as causative agents.

Key Words: Chiari I malformation, cerebellar tonsillar resection, posterior reversible encephalopathy syndrome, pressors, suboccipital craniectomy

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Quick Response Code:**INTRODUCTION**

Improved technology in magnetic resonance imaging (MRI) has facilitated the identification of Chiari I malformations at an increasing rate.^[2,25] Patients typically present with headaches (81%), pseudotumor-like episodes (78%), Meniere's disease-like syndrome (74%), lower cranial nerve signs (52%), and spinal cord disturbances without syringomyelia (66%).^[25] Common problems associated with Chiari I malformations include syringomyelia (65%), scoliosis (42%), and basilar

invaginations (12%).^[25] Several surgical treatment options are effective, the posterior fossa decompression method, including a sub-occipital decompression, which may also be done in conjunction with durotomy, duraplasty, cerebellar tonsillar resection, or cervical laminectomy.^[14,23,35,43]

Some of the most frequent postoperative symptoms include pain, weakness, numbness, and unsteadiness; while the most common postoperative complication is respiratory depression (14%).^[29] Additionally, after decompression surgery for Chiari I malformations, one

may expect issues leading to cranial nerve neuropathies; pseudomeningocele; cerebral spinal fluid (CSF) leaks; infection; aseptic meningitis; hydrocephalus; inadequate decompression, causing reobstruction of flow; and cerebellar ptosis.^[28,29] Another serious but rare potential complication of posterior fossa decompression is posterior fossa syndrome. This entity is more commonly described in tumor resection but remains a possibility when cerebellar manipulation, in particular manipulation of or injury to the vermis takes place, and has been described in cases of infection, trauma and hemorrhage.^[1,3,8,10,11,18,24,34,39,42] While patients with this postoperative complication characteristically present with cerebellar mutism, often this is accompanied by ataxia, hypotonia, cranial nerve palsies, emotional lability, dysphagia, decreased motor movement, or impaired eye opening.^[13,31]

In this case report, the authors present a patient who underwent surgical decompression of a Chiari I malformation with subsequent evolution of cerebellar vermian infarcts. Upon initiating hypertensive therapy for ischemia, the patient developed PRES.

CASE REPORT

History and examination

A 25-year-old right handed female initially presented with complaints of left upper and lower extremity paresthesias and gait disturbance as well as nausea and vomiting. Her symptoms had progressed for a few months prior to admission. Her initial workup included an MRI of the brain and cervical spine, which showed a Chiari I malformation with tonsillar descent beyond the level of the C1 lamina and syringomyelia and syringobulbia with dilation throughout the cervical spinal cord and no evidence of hydrocephalus [Figure 1]. On physical exam she was noted to be weak in bilateral upper extremities with 4+/5 motor strength in the right upper extremity and 4-/5 motor strength in the left upper extremity. Bilateral lower extremities were 5/5 strength with the exception of bilateral extensor hallucis longus, which were 4/5. She had brisk patellar reflexes bilaterally and impaired tandem gait.

Operation and pathological findings

She underwent a suboccipital craniectomy, C1 laminectomy, cerebellar tonsillar cauterization using bipolar cautery, and duraplasty with Gore-Tex dural substitute. Intraoperative somatosensory evoked potentials (SSEP) and motor evoked potentials (MEP) monitoring demonstrated impaired amplitude in the right lower extremity, with no change following positioning or throughout the operation other than gradual improvement in the right lower extremity MEP during the surgery. Postoperatively she improved to full strength in the right upper extremity and 4+/5 throughout the left upper extremity. She was started on a dexamethasone taper postoperatively as prophylaxis



Figure 1: A preoperative T1 sagittal spin echo MRI of the brain shows a Chiari I malformation with tonsillar descent beyond the level of the C1 lamina and syringomyelia and syringobulbia with dilation throughout the cervical spinal cord and no evidence of hydrocephalus

against an inflammatory response given the cerebellar manipulation. Postoperative MRI revealed a suggestion of an acute infarct of the right inferior vermis and otherwise normal postoperative changes [Figure 2]. The MRI findings were attributed to the bipolar cautery of the cerebellar tonsils. She remained in the hospital for 4 days postoperatively for pain control and monitoring of slight postoperative dysphagia and was subsequently discharged to a rehabilitation facility.

Postoperative course

She was readmitted to the hospital from rehab 6 days after discharge with complaints of worsening of her swallowing. She also noted left sided pain and worsening weakness. At this point her exam had declined from her postoperative exam and motor exam now revealed 3/5 strength in left dorsiflexion and extensor hallucis longus. At the time of readmission, she was normotensive with a blood pressure of 130/70. A swallow evaluation revealed significant aspiration. She underwent an MRI that showed bilateral acute infarcts of the cerebellar vermis [Figure 3]. We did not have a clear explanation for this increase from her postoperative scan. Given the evolution of the infarct she was started on pressors to increase her systolic parameters to 160-180 mmHg and optimize perfusion in the setting of ischemia. Two days after initiating pressors, the patient had a generalized tonic-clonic seizure. A CT of the head at that time demonstrated diffuse loss of sulci with effacement of the suprasellar and quadrigeminal cisterns. She was subsequently intubated for worsening mental status and lethargy. A follow-up MRI revealed diffuse bifrontal and parietal gray matter hyperintensity on FLAIR without changes on diffusion-weighted imaging consistent with PRES and suggestive of edema from hypoxic ischemic changes secondary to prior seizure [Figure 4]. An electroencephalography (EEG) showed no further

seizure activity and CSF cultures were negative. She underwent an angiogram to evaluate for vasospasm as a possible etiology of her evolving infarcts, which showed no evidence of vasospasm or vasculitis. Following those results, the pressors were weaned off. A repeat CT of the head was obtained, which showed increasing cerebral edema. As a result of the increasing edema and her compromised neurologic exam, an intracranial pressure (ICP) monitor was placed and maintained for 2 days, which showed low opening pressure, and ICPs ranging from 1 to 7. Follow-up MRIs demonstrated progressive resolution of the cerebral edema and FLAIR signal [Figures 5] with complete resolution at 2 years postoperative [Figure 6].

She had progressive improvement in her mental status and was extubated. She continued to have swallowing difficulties and underwent serial swallow evaluations, ultimately requiring placement of a percutaneous endogastric tube (PEG). She ultimately improved and had only residual left hand weakness with motor strength 4/5. She was ultimately transferred to rehab. She continued to have significant left upper extremity dysesthesia. At 9-month follow-up, she was noted to have worsening of her syrinx and further descent of her cerebellar tonsils and so she underwent placement of a ventriculoperitoneal shunt.

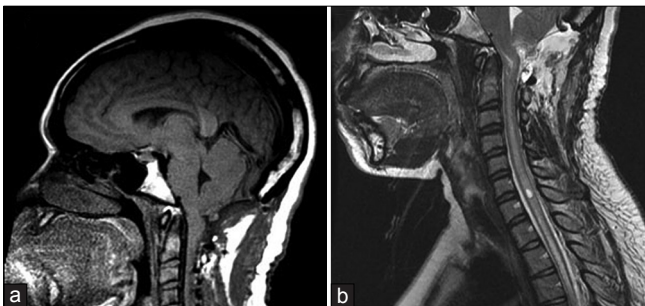


Figure 2: T1 sagittal MRI of the brain (a) and T2 sagittal MRI of the cervical spine (b) obtained on postoperative day one, which shows cerebellar prolapse

At her latest follow up 4 years postoperatively, her motor exam was 5/5 throughout her right upper extremity, 5/5 in her left deltoid, biceps and triceps and 4/5 left opponens and 3/5 interossei. Both lower extremities were 5/5 strength throughout with a steady gait and impaired tandem gait, significantly improved from prior, though she has a stiff and wide-based spastic gait. She was no longer having any dysphagia, had no bulbar symptoms and her PEG had been removed. Her left upper extremity dysesthesia has significantly improved. Her 4 year follow-up MRI demonstrates resolution of her syringomyelia [Figure 7].

DISCUSSION

PRES, also known as reversible posterior leukoencephalopathy syndrome, was first described in 1996 by Hinchey *et al.*^[16] PRES typically presents with clinical seizures, headaches, altered mental status/function, seizures, loss of vision, and relatively symmetric edema in the subcortical white matter as well as less frequently in the cortices of the occipital and parietal lobes.^[12,15,33,40] Neuroimaging can also demonstrate brainstem/cerebellar involvement in up to 58% of cases.^[22] Some of the more common causes include hypertensive encephalopathy,^[7] preeclampsia/eclampsia,^[7] cyclosporine A neurotoxicity,^[36,37,41] infections,^[6] electrolyte imbalance,^[6] solid organ transplantation,^[21] autoimmune diseases,^[15] immunosuppressive medications,^[19] and uremic encephalopathies.^[7,32] Patients with PRES frequently have acute medical conditions, receive immunosuppressive drugs,^[9] and have high blood pressure,^[4,5,15] though this is not always the case.^[27]

Currently, the mechanism of PRES is unknown. However, two theories have been suggested. The older theory, proposes hypertension leading to cerebral autoregulatory vasoconstriction, ischemia, and cerebral edema. The newer theory proposes that the hypertension is too great for autoregulation, causing vasogenic edema.^[5] Hypertension is seen in 50-70% of patients with PRES

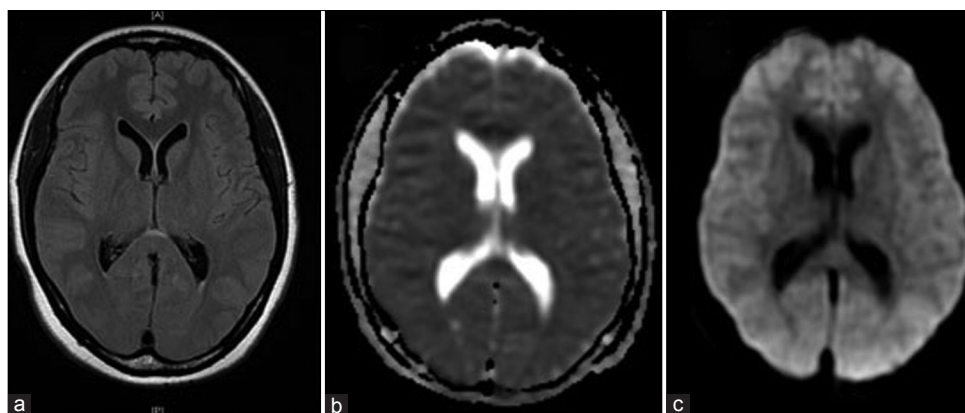


Figure 3: T2 axial FLAIR MRI (a) and apparent diffusion coefficient MRI (b) and diffusion MRI (c) at postoperative day 5, upon return with new onset left sided weakness, demonstrating no acute infarcts

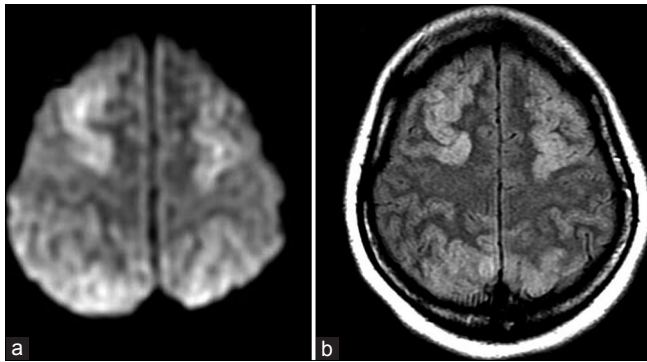


Figure 4: Diffusion Weighted Imaging (DWI) (a) and T2 axial FLAIR MRI (b) on postoperative day 9 showing increased signal in the bilateral frontal and parietal lobes, consistent with PRES

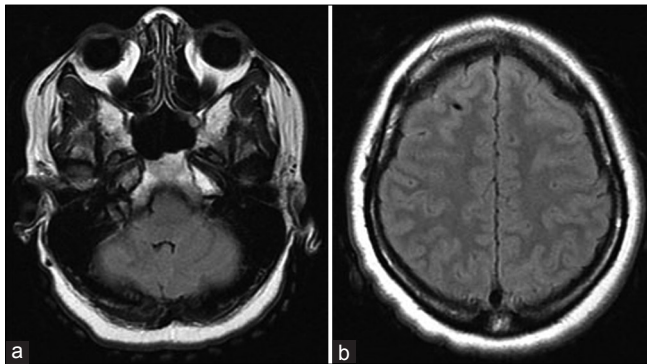


Figure 6: (a and b) T2 axial FLAIR MRI 2 years after surgery showing complete resolution of changes associated with PRES

and treatment of the hypertension is related to improved symptoms.^[5,17,38] A retrospective study of 36 patients presenting at the Mayo Clinic with PRES demonstrated hypertension with mean systolic blood pressure of 187 mmHg at presentation.^[22] Several medications including lenalidomide, dexamethasone, and cyclosporine have been associated with PRES.^[19,26,40] In particular, PRES was reported in a 60-year-old woman following treatment with high-dose steroids after an autologous peripheral blood stem cell transplantation for multiple myeloma.^[40] Steroid treatment in others has been associated with PRES, including a patient with bronchial asthma^[20] and Henoch–Schonlein purpura.^[30]

In this case report, PRES syndrome was not suspected until late in the patient's postoperative course, as her initial symptoms were suspected to be postoperative sequelae of her surgery. Following surgery, the patient progressed well despite some pain and slight dysphagia. During her admission to a rehabilitation facility, the patient was on the following postoperative medications: Pregabalin, dexamethasone, pantoprazole, docusate, milk of magnesia, morphine, oxycodone with acetaminophen, ondansetron, and senna. Issues leading to the diagnosis of PRES began 2 days into rehabilitation therapy when the patient presented with worsening dysphagia and new onset left lower extremity pain and weakness. Given the concern for evolving infarcts on MRI and stroke-like

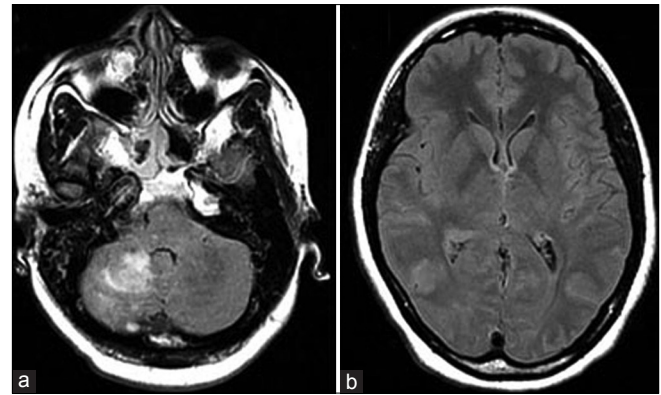


Figure 5: T2 axial FLAIR MRI demonstrating continuing hyperintensity of the right cerebellar vermis (a) and resolution of the right parietal increased signal one week after cessation of hypertensive therapy (b) (postoperative day 14)

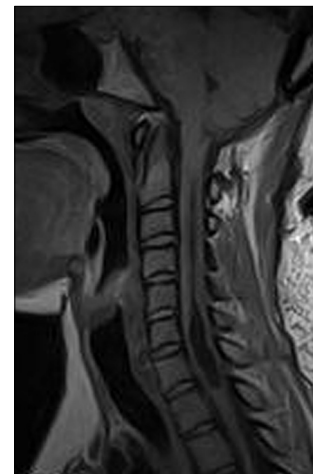


Figure 7: Sagittal T1 image 4 years postoperatively showing marked reduction in cervical syrinx and cerebellar prolapse

symptoms, hypertensive therapy was initiated to optimize perfusion. Shortly after initiating this therapy, our patient experienced a seizure followed by precipitous neurological decline. Eventually, MRI revealed typical findings of PRES including symmetrical edema of the bilateral frontal and parietal gray matter. With this new concern for PRES, hypertensive therapy was discontinued with subsequent improvement in her neurologic status and resolution of symptoms. Our patient likely developed PRES, in part, from the dexamethasone therapy and was accelerated by the use of pressors. Given that her initial MRI findings upon readmission did not fully explain the new left lower extremity weakness, it is also possible that the left lower extremity symptoms were an initial manifestation of PRES, which may have been a result of her steroid therapy. This was subsequently exacerbated by hypertensive therapy. Fortunately, the PRES was identified and the pressors were discontinued, allowing for normalization of the patient's blood pressure and leading to improvement of symptoms.

CONCLUSION

Here, we present an interesting case of a 25-year-old female who developed PRES several days postoperative from a Chiari decompression who further deteriorated following the administration of pressors. While the mechanism of PRES remains controversial given its diverse clinical presentation, some of the leading theories suggest steroids and hypertension as the causative agents. However, only 70% of PRES patients have hypertension, thus indicating additional unknown factors. Our patient did not have hypertension prior to the administration of pressors, leading us to the conclusion that steroid treatment may have been the inciting factor. Given this potential correlation, PRES should be considered in any patient presenting with a decline in neurological status after initiation of steroids and hypertensive therapy.

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