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Case Report

Acquired Chiari malformation Type I and holocord syringomyelia associated with a high-flow supratentorial fistulous arteriovenous malformations: A case report and literature review

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ABSTRACT

Background: Chiari malformation Type I (CMI) is generally considered a congenital lesion and typically associated with syringomyelia. Acquired CMI or adult Chiari malformation caused by intracranial mass is extremely rare. Brain arteriovenous malformations (AVMs) are characteristically symptomatic due to seizure, intracranial hemorrhage, or neurological deficit. We report an extremely rare case of an acquired CMI and extensive syringomyelia associated with a large supratentorial AVM.

Case Description: A 35-year-old woman was referred to our institute after a diagnosis of CMI and extensive syringomyelia from whole-spine magnetic resonance imaging (MRI) due to complaining of low back pain radiating to the right leg for the past 1 month. She had intermittent headache for 2 years. The patient underwent suboccipital decompression and C1 laminectomy followed by duraplasty. Two months later, she developed severe right-sided sciatic pain and complete right foot drop. Follow-up MRI revealed progressive enlargement of a syrinx cavity at the lower spinal cord and a large right parieto-occipital AVM with markedly dilated cortical draining veins and diffuse engorgement of dural venous sinuses was detected. This AVM supplied mainly by enlarged cortical branches of the right middle cerebral artery and posterior cerebral artery with multiple dural supplies. Endovascular treatment of a high-flow fistulous AVM was successfully performed with N-butyl cyanoacrylate (NBCA) through the hypertrophic branches of the right middle cerebral artery. Four months after embolization, the patient had recovered completely from the right foot drop. Further staged embolization was planned to reduce the size and flow of the AVM before stereotactic radiosurgery. However, the patient was lost to follow-up due to financial reason. One year later, she developed sudden severe headache followed by alteration of conscious due to intraventricular hemorrhage from the AVM, leading to obstructive hydrocephalus requiring cerebrospinal fluid diversion. During a period of 2 years, the patient underwent several staged embolization with NBCA and Onyx. Final cerebral angiography after embolization demonstrated a significant reduction in size and flow of the brain AVM. A control whole-spine MRI revealed a significant reduction in syrinx size. At the end of embolization, the patient had no neurological deficit. However, she had suffered from persistent central neuropathic pain at the right lower extremity. The AVM remnant was further treated by stereotactic radiosurgery.

Conclusion: Increased cerebral venous hypertension secondary to a high-flow supratentorial AVM leading to posterior fossa venous hypertension may play a major role in the pathogenesis of CMI, induced the formation

of syringomyelia. Endovascular treatment of brain AVM, the underlying cause of CMI, resulted in a significant reduction of the size of the syrinx. The need for cranial imaging in initial evaluation of cases with adult Chiari malformation is important.

Keywords: Brain arteriovenous malformations, Chiari malformation type I, Foot drop, Holocord syringomyelia, Neuropathic pain

INTRODUCTION

Chiari malformation Type I (CMI) is the most common type of CM and characterized by caudal descent of the cerebellar tonsils exceeding 5 mm below the foramen magnum with no involvement of the brainstem. The onset of symptoms usually occurs during adolescence or adulthood. The predominant clinical symptoms are recurrent headache and cervical pain. CMI is in general a congenital origin related to posterior fossa anomalies and typically associated with syringomyelia, but rarely associated with hydrocephalus. Symptoms of syringomyelia are usually related to spinal cord dysfunction and neuropathic pain.[4,6,26] Acquired CMI also known as adult CM is often described as a heterogeneous group of conditions wherein disruption of normal cerebrospinal fluid (CSF) flow through the foramen magnum is the underlying commonality. Acquired or secondary CMI caused by intracranial mass is extremely rare.[25] The clinical presentation of acquired CMI can be similar to that of congenital CMI, and syrinx may develop as a result of tonsillar descent. Identification of the exact cause of acquired CMI before treatment is extremely important because the underlying cause should be treated directly.[6]

Holocord syringomyelia is syrinx involving most or all levels of the spinal cord and patients with extensive syrinx have the potential for more extensive and severe neurological sequelae than those with small syrinxes.[20]

The authors described an extremely rare case of a large high-flow supratentorial arteriovenous malformation (AVM) causing an acquired CMI with holocord syringomyelia. In our case, CMI was initially misdiagnosed as congenital origin and the patient underwent bony decompression with duraplasty followed by deterioration of the clinical symptoms. Subsequently, a pre-existing supratentorial AVM was discovered accidentally during work up of CMI lesion. The authors also reviewed the patients with brain AVM associated with CMI in the literature.

CASE DESCRIPTION

A 35-year-old woman with a medical history of β-thalassemia trait visited the local hospital and complained of intermittent headache for 2 years. She experienced low back pain radiating to the right leg for 1 month. Magnetic resonance imaging (MRI) of the whole spine was obtained and revealed the caudal descent of the cerebellar tonsils through the foramen magnum and a syringomyelic cavity extending from the cervicomedullary junction to the conus medullaris. At the lower spinal cord, syringomyelia affected predominantly on the right side [Figure 1]. These findings were consistent with CMI with holocord syringomyelia. The patient was transferred to our institute and underwent suboccipital decompression and C1 laminectomy followed by duraplasty. Postoperative course was uneventful and she was discharged home 1 week after the surgery.

Two months later, she developed severe right-sided sciatic pain and complete right foot drop. She walked with a compensatory high steppage gait and required a cane during walking. Medical research council grading for the power strength of the right lower extremity revealed the following: iliopsoas 5/5, hip adductors 4/5, gluteus medius 4/5, quadriceps 4/5, hamstrings 4/5, tibialis anterior 0/5, extensor hallucis longus 0/5, gastrocnemius 4/5, and toes flexors 3/5. Examination of her both upper extremities and left lower extremity was normal. Follow-up full spinal MRI showed the progression of syringomyelia at the lower spinal cord and edema in the conus medullaris [Figure 2]. During performing MRI, the neuroradiologist suspected abnormal

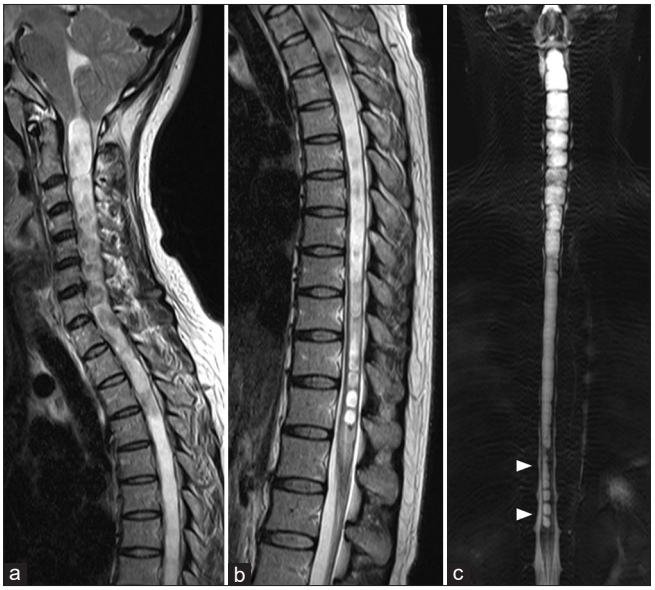

Figure 1: Sagittal T2-weighted magnetic resonance images of (a) cervicothoracic and (b) thoracolumbar spines reveal cerebellar tonsillar herniation below the foramen magnum and holocord syringomyelia. (c) Coronal view of three-dimensional magnetic resonance myelography of the whole spine clearly demonstrates the septations in the syrinx. At the lower spinal cord, syringomyelia (arrowheads) affected predominantly on the right side.

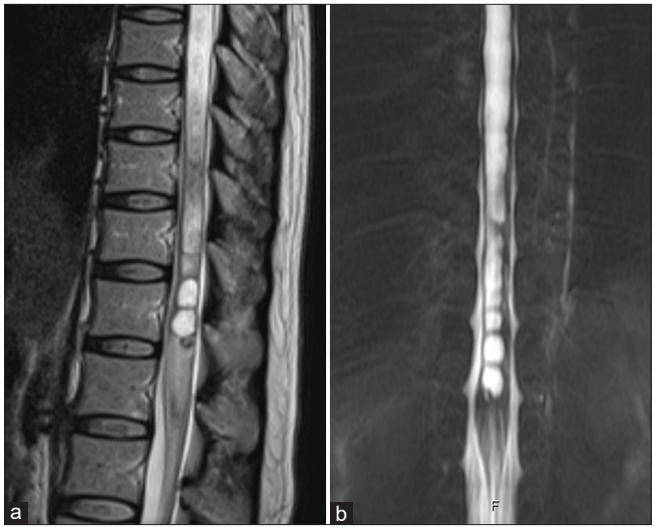

Figure 2: (a) Sagittal T2-weighted magnetic resonance image and (b) coronal view of three-dimensional magnetic resonance myelography of the thoracolumbar spine demonstrate the progression of syringomyelia at the lower spinal cord and edema in the conus medullaris.

flow voids in the brain. Therefore, cranial MRI and MRA were obtained and demonstrated a large AVM involving the right parietal and occipital lobes with markedly dilated cortical draining veins and diffuse engorgement of dural venous sinuses, including the posterior two-third portion of the superior sagittal sinus (SSS), bilateral transverse sinuses, torcula, straight sinus, and the vein of Galen. There was a large flow-related venous varix in the anterior portion of the nidus, probably protruded into the posterior horn of the right lateral ventricle. In addition, dilated bilateral basal veins of Rosenthal, left lateral mesencephalic vein, and left vein of cerebellopontine fissure were observed. No hydrocephalus was seen [Figure 3]. We recognized that CMI in this case was initially misdiagnosed as congenital origin. In addition, cranial imaging in initial evaluation of our case with adult CM should be performed before bony decompression and duraplasty.

Subsequently, cerebral angiography was performed and revealed a large right parieto-occipital AVM supplied mainly by enlarged cortical branches of the right middle cerebral artery (posterior parietal and angular branches) and posterior cerebral artery (parieto-occipital and calcarine branches) with multiple dural supply. Dural supply to the AVM included the meningohypophyseal and inferolateral branches of the right cavernous ICA, bilateral middle meningeal arteries, and right occipital artery. In addition, there was indirect supply through leptomeningeal anastomosis from the cortical branches of the right anterior cerebral artery. The AVM drained predominantly toward the SSS through enlarged parietal and occipital cortical veins [Figure 4]. Another venous drainage drained into the right transverse sinus through the dilated occipital basal veins. In addition, venous draining of the inferior compartment of the AVM drained into the straight sinus and the vein of Galen with retrograde venous reflux into bilateral enlarged basal veins of Rosenthal and dilated vein running along left-sided brainstem corresponding with dilated posterior fossa veins on MRI [Figure 5].

The patient underwent transarterial embolization with N-butyl cyanoacrylate (NBCA) through the hypertrophic branches of the right middle cerebral artery. Superselective catheterization of the middle cerebral branches demonstrated the fistulous type of AVM [Figure 6]. Four months after embolization, the patient recovered completely from the right foot drop. Further staged embolization was planned to reduce the size and flow of the AVM before stereotactic radiosurgery. However, the patient was lost to follow-up due to financial reason.

One year later, the patient developed sudden severe headache followed by alteration of conscious. She was admitted at the local hospital. Cranial computed tomography (CT) scan showed intraventricular hemorrhage in the right lateral and fourth ventricles with ventricular enlargement [Figure 7]. She underwent left frontal ventriculostomy followed by the left occipital ventriculoperitoneal shunt. After clinical stabilization, the patient was referred to our institute. During a period of 2 years, the patient underwent several staged embolization with NBCA and onyx through the branches of the right posterior cerebral artery, bilateral middle meningeal arteries, and right occipital artery [Figure 8]. Final cerebral angiography after embolization demonstrated a significant reduction in size and flow of the brain AVM [Figure 9]. Follow-up cranial CT scan showed no hydrocephalus [Figure 10]. A control whole-spine MRI revealed significant reduction in syrinx size [Figure 11]. The AVM remnant was further treated by stereotactic radiosurgery. At the end of embolization, the patient had no neurological deficit. However, she had suffered from persistent central neuropathic pain at the right lower extremity. The pain was controlled in acceptable level with tramadol at the dosage of 50–100 mg/day and gabapentin 1800 mg/day.

DISCUSSION

Acquired CMI with syringomyelia

Acquired or secondary CMI and associated syrinx have been previously reported following lumbar puncture, lumboperitoneal shunting, and spontaneous spinal CSF leakage.[1,18,21] In addition, space-occupying lesions, such as meningioma, colloid cyst, craniopharyngioma, posterior fossa arachnoid cyst, epidermoid cyst, and medulloblastoma, are rare causes of CMI with syringomyelia.[24]

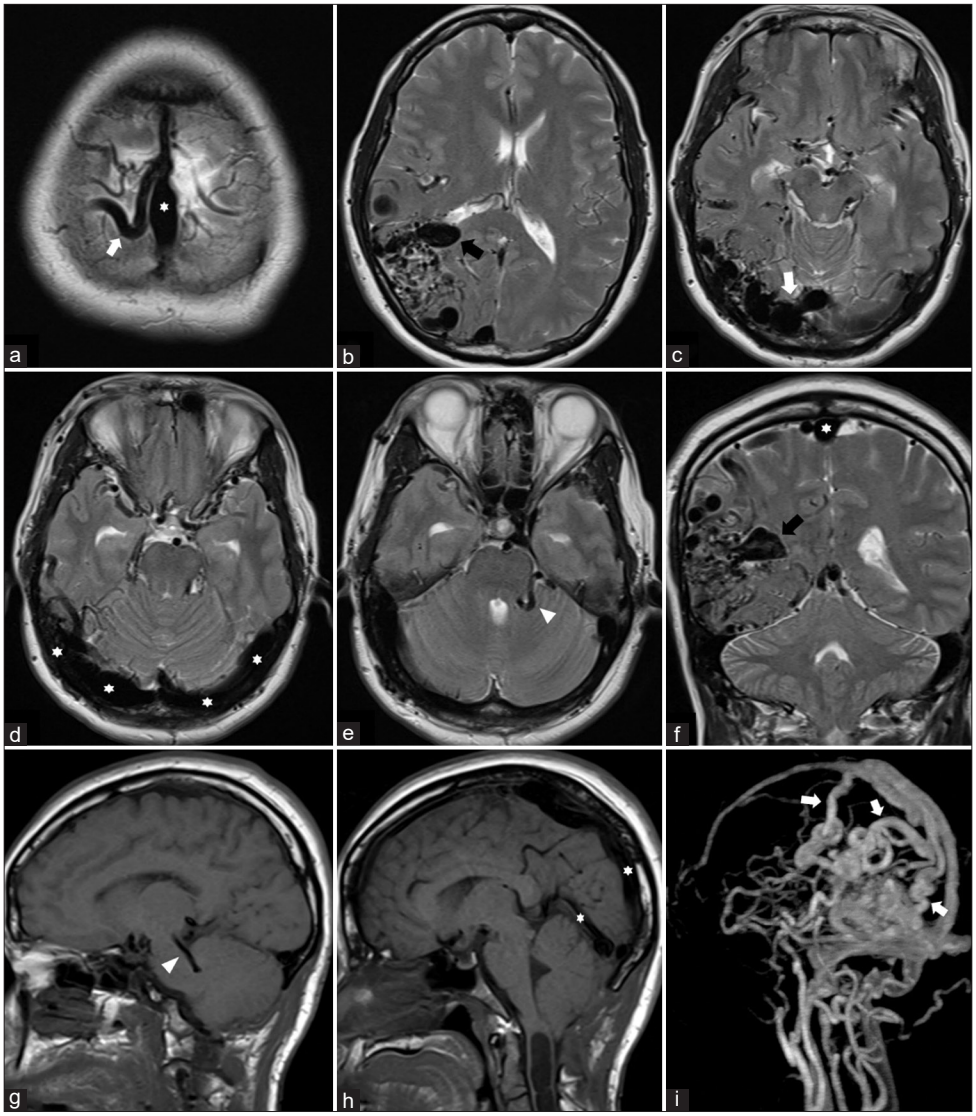

Figure 3: (a-e) Sequential axial, (f) coronal T2-weighted, (g and h) sagittal T1-weighted magnetic resonance images, and (i) contrast-enhanced magnetic resonance angiography reveal a large brain arteriovenous malformation involving the right parietal and occipital lobes with markedly dilated cortical draining veins (arrows) and diffuse engorgement of dural venous sinuses (asterisks), including the posterior two-third portion of the superior sagittal sinus, bilateral transverse sinuses, torcula, and straight sinus. There is a large venous varix (black arrows) in the anterior portion of the nidus, probably protruded into the posterior horn of the right lateral ventricle. In addition, dilated posterior fossa veins (arrowheads) are noted.

According to the systemic review of acquired CMI and associated syrinx secondary to space-occupying lesions by Wang *et al*.,[25] they found that acquired CMI appeared to have a higher rate of associated syringomyelia compared with congenital origin. Meningioma and arachnoid cyst were the lesions most associated with acquired CMI. Most space-occupying lesions causing acquired CMI were infratentorial and large. Syrinx resolved or improved following resection of the underlying lesion in nearly all cases. Another literature review of CMI with syringomyelia in patients with posterior fossa arachnoid cysts by Martínez-Lage *et al*.,[11] they found that the obstruction to CSF circulation through the foramen magnum was because of blockage by the walls of the cysts itself in most cases. Rarely, acquired CMI originated from the cerebellar displacement exerted by the push of the arachnoid cyst.

Acquired CMI associated syringomyelia caused by supratentorial mass is extremely rare.[9,10,16,23] CMI with syringomyelia may produce clinical symptoms earlier than primary lesion with secondary CMI.[24] Interestingly, Morioka *et al*.[13] reported a case of acquired CMI associated with bilateral chronic subdural hematomas in young woman. They speculated that bilateral chronic subdural hematomas

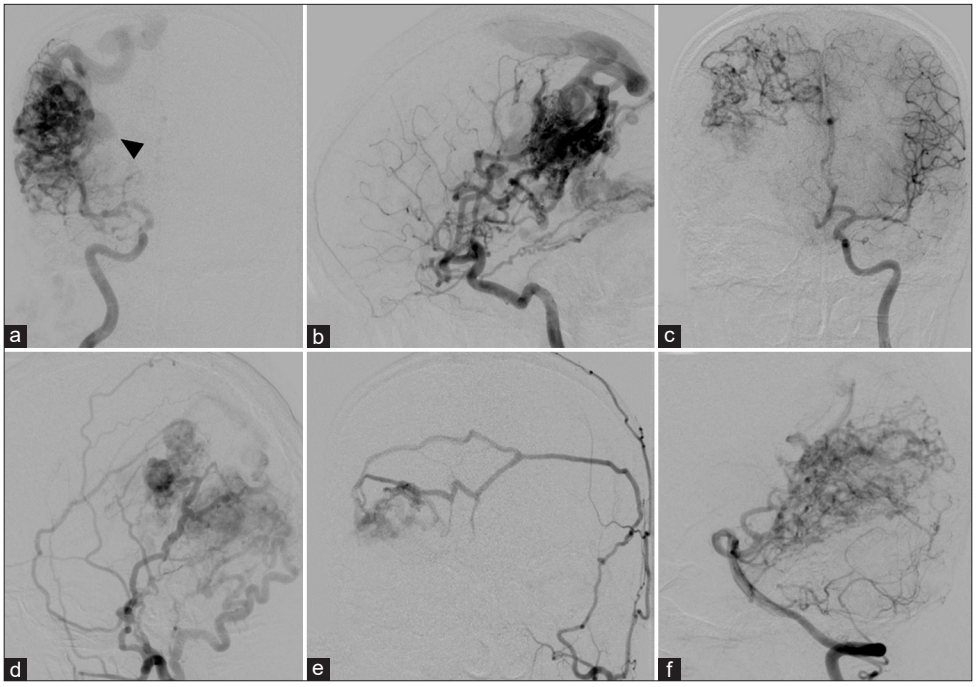

Figure 4: (a) Anteroposterior (AP) and (b) lateral views of the right internal carotid artery (ICA), (c) AP view of the left ICA, (d) lateral view of the right external carotid artery, (e) AP view of the left external carotid artery, and (f) lateral view of the left vertebral artery injections show a large right parieto-occipital arteriovenous malformation (AVM) supplied by enlarged cortical branches of the right middle cerebral artery, and posterior cerebral artery with multiple dural supply. Dural supply to the AVM includes the meningohypophyseal and inferolateral branches of the right cavernous ICA, bilateral middle meningeal arteries, and right occipital artery. In addition, there is indirect supply through leptomeningeal anastomosis from the cortical branches of the right anterior cerebral artery. The AVM drains toward the superior sagittal sinus through enlarged cortical veins. A large venous varix (arrowhead) is noted.

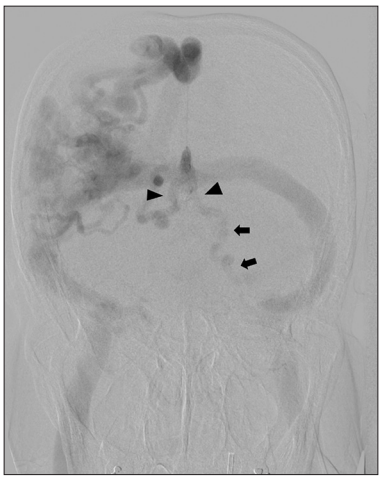

Figure 5: Anteroposterior view of the left vertebral artery injection illustrates retrograde venous reflux into bilateral enlarged basal veins of Rosenthal (arrowheads) and dilated vein running along left-sided brainstem (arrows) corresponding with dilated posterior fossa veins on magnetic resonance images.

may cause the central herniation and downward impaction of the cerebellar tonsils into the foramen magnum which was sufficient to block egress of CSF from the fourth ventricle, leading to expansion of the central canal. Spontaneous resolution of the hematomas resulted in clinical and radiological improvement. They also suggested that the mechanism for the development of CMI and associated syringomyelia from supratentorial lesions is analogous to that from posterior fossa lesions.

The pathogenesis of syringomyelia associated with acquired CMI

Based on anatomical and dynamic MRI, and intraoperative ultrasonography before, during, and after surgical decompression of the foramen magnum, Heiss *et al*.[5] studied the pathophysiology of syringomyelia progression in patients with congenital CMI. They found that the progression of syrinx was associated with obstruction of the subarachnoid space at the foramen magnum. They explained that the brain expansion occurs during systole leading to CSF movement from the basal cisterns to the upper portion of the spinal canal. The CSF returns to the cranial compartment during diastole. With obstruction to rapid movement of CSF at the foramen magnum, the cerebellar tonsils move downward with each systolic pulse, functioning as a prison on the partially isolated spinal CSF and producing a systolic

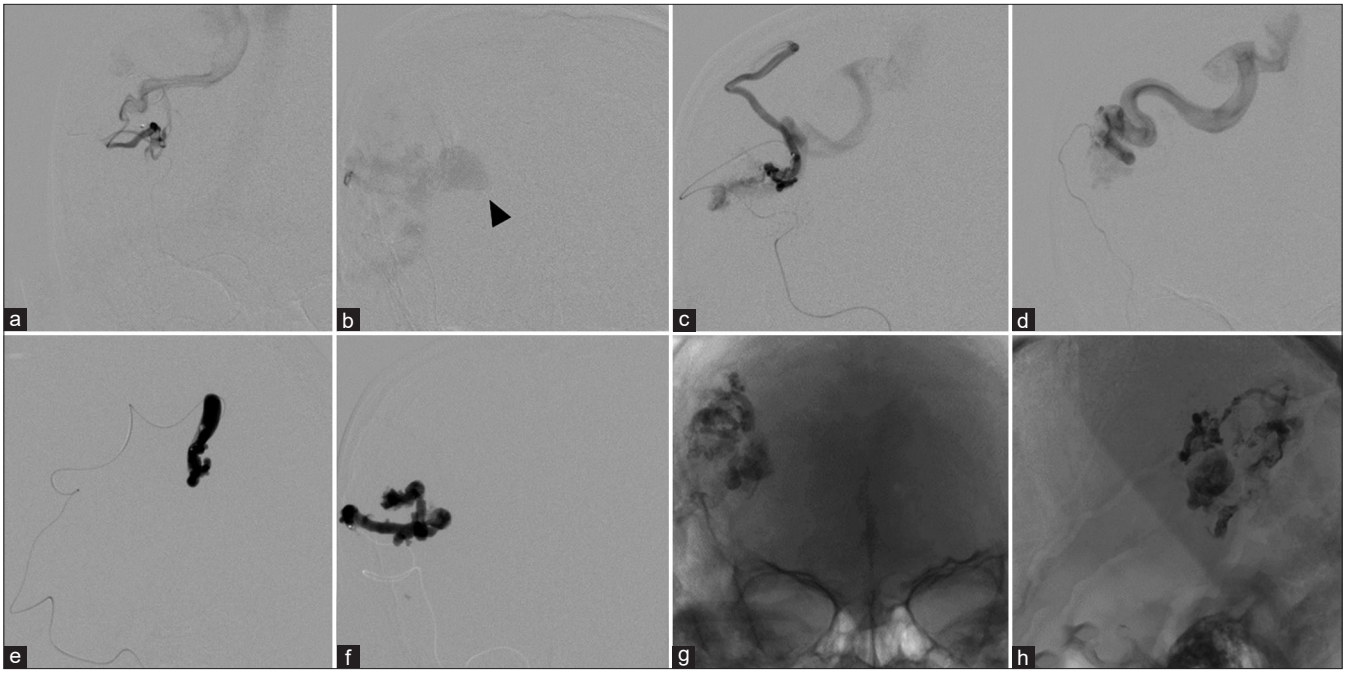

Figure 6: (a-d) Anteroposterior views of different injections from the branches of the right middle cerebral artery through the microcatheters reveals the fistulous sites of arteriovenous malformation with markedly dilated cortical draining veins. A large venous varix (arrowhead) is noted. (e and f) During embolization with N-butyl cyanoacrylate. (g) Anteroposterior and (h) lateral views of unsubtracted image demonstrate the glue cast.

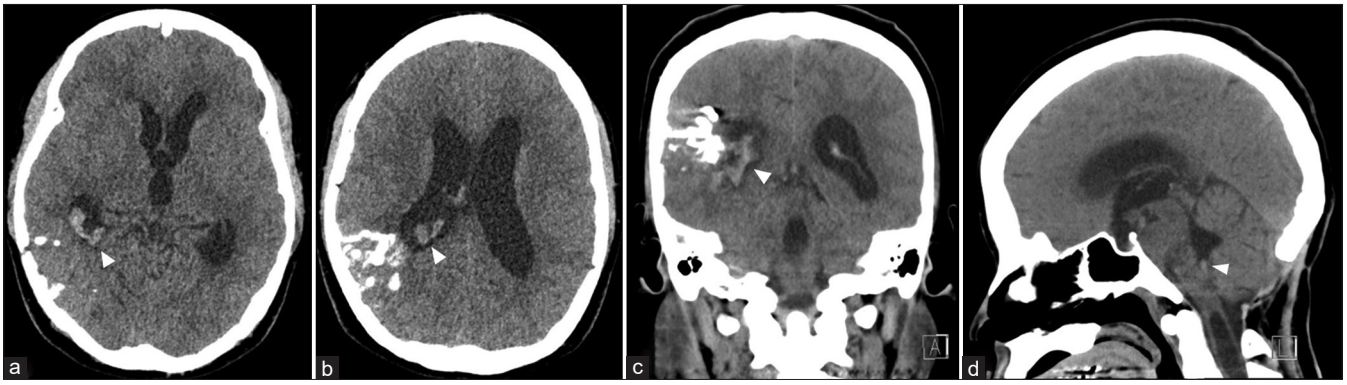

Figure 7: (a and b) Axial, (c) coronal, and (d) sagittal views of cranial computed tomography scan reveal intraventricular hemorrhage (arrowheads) in the right lateral and fourth ventricles with ventricular enlargement.

pressure wave in the spinal CSF acting on the surface of the spinal cord. The progression of syrinx results from abruptly compressing the spinal cord and propelling the fluid in the syrinx longitudinally with each pulse and may be force spinal CSF driven into the spinal cord through the perivascular and interstitial spaces.

The pathogenesis of syringomyelia in patients with acquired CMI may be analogous to the development of syrinx with congenital CMI.[13] There is functional occlusion at the level of the foramen magnum, leading to alteration of CSF velocity and pressure. However, the pathogenesis of syrinx formation in acquired CMI remains unclear because why some lesions in posterior fossa cause an acquired CMI whereas others do not.[25]

Literature review of brain AVM associated with Chiari Type I malformation

We reviewed the literature of patients with cerebral AVMs associated with CMI and found five cases, including our one case [Table 1].[2,14,15,19] All patients were female with a median age of 19 years (range 1–35 years).

First, O'Shaughnessy *et al*.[14] reported the first case of acquired CMI associated with a supratentorial AVM. A 19-year-old

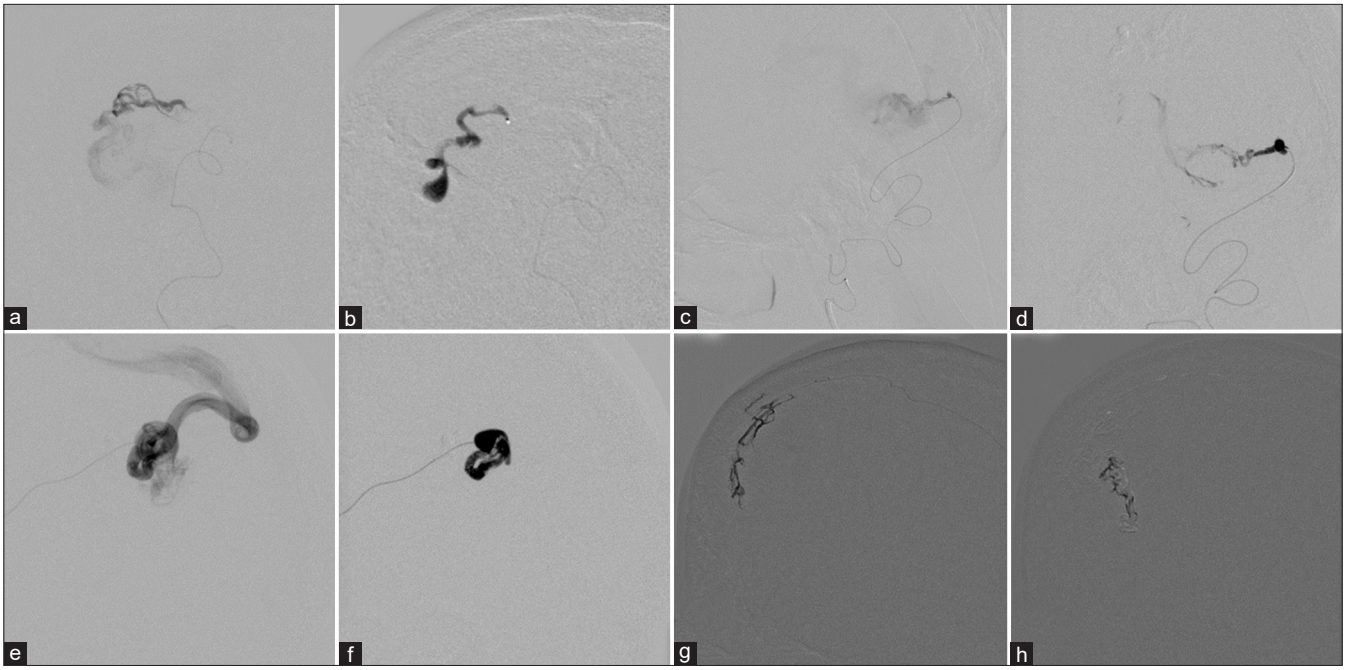

Figure 8: (a) Anteroposterior view of the right posterior cerebral artery, lateral view of (c) the right occipital artery, and (e) the right middle meningeal artery injections the fistulous sites of arteriovenous malformation with markedly dilated cortical draining veins. (b, d, and f) During glue injections with good penetration into the proximal draining veins. (g and h) During Onyx injections through the contralateral middle meningeal branch in anteroposterior view.

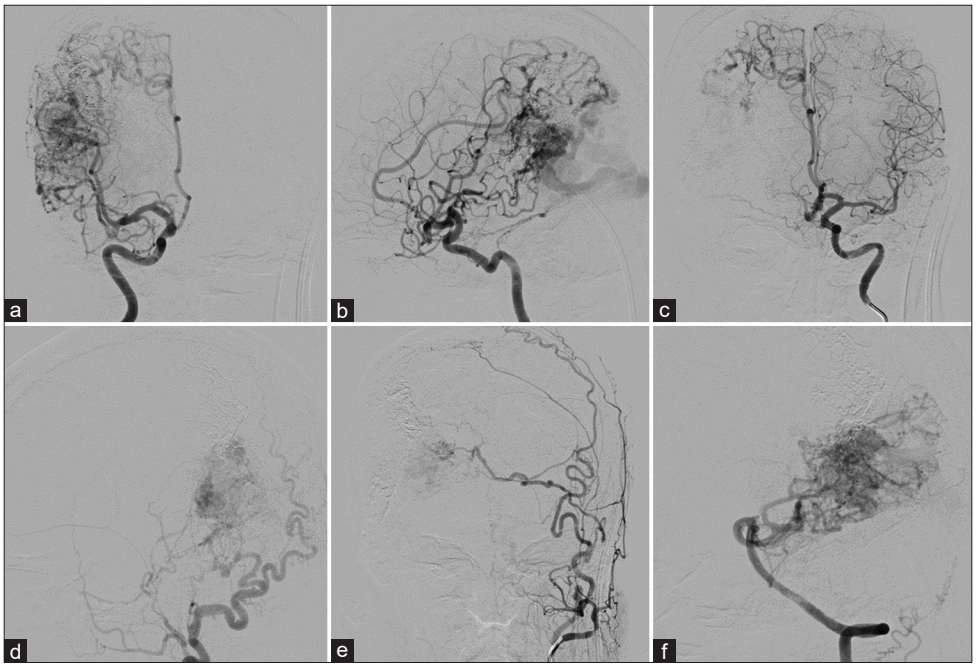

Figure 9: Cerebral angiography obtained before radiosurgery. (a) Anteroposterior (AP), (b) lateral views of the right internal carotid artery (ICA), (c) AP view of lateral view of the left ICA, (d) lateral view of the right external carotid artery, (e) AP view of the left ECA, and (f) lateral view of the left vertebral artery injections demonstrate significant reduction in size and flow of the brain arteriovenous malformation.

woman was diagnosed a large AVM in the right temporal, parietal, and occipital lobes 3 years earlier due to headache. CMI was seen to originate and worsen in concert with progression of the AVM. They speculated that the acquired CMI resulted from a high-flow venopathy of transverse and sigmoid sinuses causing occlusion of the right sigmoid

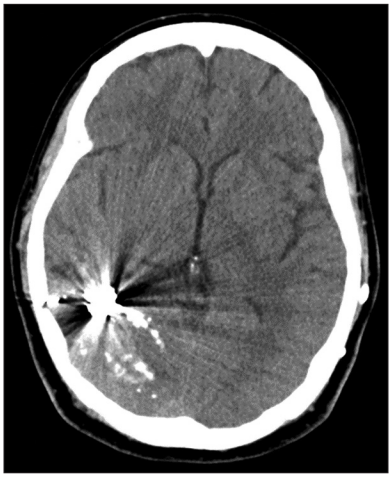

Figure 10: Cranial computed tomography scan obtained following several staged embolization shows no ventricular enlargement and metal artifact from embolic material.

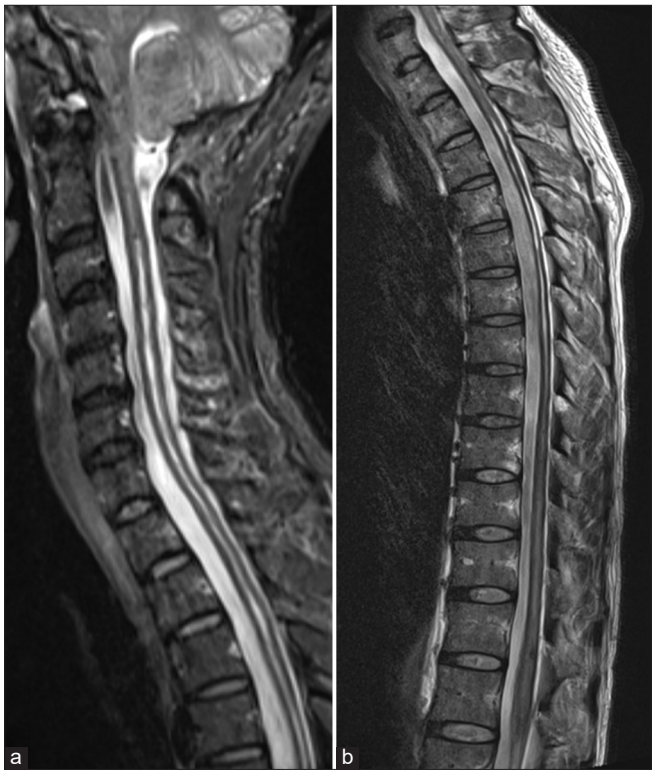

Figure 11: Sagittal T2-weighted magnetic resonance images of (a) cervicothoracic and (b) thoracolumbar spines reveal significant resolution of the syrinx cavity as compared to the previous images.

sinus, severe stenosis of the left sigmoid sinus, and extensive recruitment of venous drainage in posterior fossa. Treatment of the AVM by several staged embolization and angioplasty and stenting of the left sigmoid sinus followed by surgery resulted in improvement of the tonsillar herniation. Second, Rodesch *et al*.[19] reported the second case of 21-year-old woman with a large tectal AVM associated with acquired CMI, cervical syringomyelia, and hydrocephalus. They speculated that hydrovenous disorder associated with the AVM created CMI that secondarily resulted in syringomyelia. In addition, CMI may cause by secondary congestion of the posterior fossa caused by infratentorial venous drainage of the AVM without severe restriction of the sinusal venous out flow. All cerebellar tonsillar herniation, syringomyelia, and hydrocephalus resolved spontaneously after endovascular treatment of the vascular malformation. Third, Chen *et al*.[2] described another case of 1-year-old girl with acquired CMI associated with a frontal fistulous AVM, causing high-flow venopathy over the SSS, transverse sinus, and posterior fossa draining veins. The overflowing of the AV shunting was further worsened by the occlusion of the left sigmoid sinus. They speculated that a high-flow fistulous AVM exerted significant hemodynamic stress and may cause significant venous hypertension of posterior fossa, leading to tonsillar herniation. Both tonsillar herniation and hydrocephalus resolved after excision of the AVM. Fourth, Ogul and Kantarci[15] reported MRI finding of a 3-year-old girl with coincidence of a large left-sided supratentorial AVM and CMI with cervical syringomyelia. There were extremely dilated venous sinuses in this case.

Regarding other type of cerebral vascular malformation associated with acquired CMI and syringomyelia, Cognard *et al*.[3] described two patients with Type II dural arteriovenous fistulas (DAVFs) of the SSS associated with tonsillar herniation and syringomyelia. In each of these patients, acquired CMI was found after a follow-up period of 15–16 years, respectively, representing long-standing course. They speculated that moderate-flow DAVFs may produce insufficiency of the cerebral venous drainage, leading to intracranial hypertension, in cases with impairment of the cerebral venous outflow. On the other hand, very high-flow DAVFs may create similar insufficiency of the venous drainage despite a normal or even enlarged sinus. Tonsillar herniation in these two patients improved after arterial embolization which decreased the flow rate inside the shunt, leading to reducing the pressure inside the sinus.

The proposed pathophysiology of adult Chiari malformation coexisting with brain AVM in our case

A high-flow supratentorial fistulous AVM may cause hypertensive venopathy, representing by diffuse engorgement of sinuses and markedly dilated superficial cortical veins. The venous hypertension of posterior fossa was evident by the dilated posterior fossa veins. Subsequently, the venous hypertension of the posterior fossa caused pressure gradient over the craniospinal junction and resulted in CMI. Therefore, treating the CMI primarily resulted in deterioration of clinical symptoms later due to the existence

Table 1: Literature review of patients with brain arteriovenous malformations associated with CMI.

Authors	Gender/ Age	Symptoms and signs	Location	Syringomyelia/ Hydrocephalus	Treatment	Neurological outcome
O'Shaughnessy *et al*. (2006)[14]	F/19	A 3-year history of Rt. frontal headache, incomplete hemisensory deficit in the Lt. face, arm, and leg/visual deterioration over a 2-month period.	Rt. Supratentorial AVM	−/−	• Several staged embolization. • Angioplasty and stenting of the Lt. sigmoid sinus. • Surgery	Stable
Rodesch *et al*. (2007)[19]	F/21	Headache, frequent sensations of loss of equilibrium, and intermittent strabismus.	Infratentorial AVM (tectum)	+/+	• Four sessions of embolization • Radiosurgery for AVM remnant.	GR
Chen *et al*. (2015)[2]	G/1	Incidental findings on CT scan after sustained head injury. Three months later, she developed left upper limb weakness, vomiting, and decreased activity.	Rt. Supratentorial AVM	−/+	• Embolization • Surgery	N/A
Ogul and Kantarci (2017)[15]	G/3	Severe vomiting	Lt. Supratentorial AVM	+/N/A	N/A	N/A
Present case	F/35	A 2-year history of intermittent headache. She experienced low back pain radiating to the right leg for 1 month. She developed severe right-sided sciatic pain and complete right foot drop after surgery for CMI 2 months later.	Rt. Supratentorial AVM	+/+	• SOC and duraplasty • Several staged embolization • VP shunt • Radiosurgery	Persistent neuropathic pain

AVM: Arteriovenous malformations, F: Female, G: Girl, GR: Good recovery, Lt: Left, N/A: Data not available, Rt: Right, SOC: suboccipital craniectomy, VP: ventriculoperitoneal, CMI: Chiari malformation type I

of the primary cause, a large high-flow AVM. After significant reduction of the flow of the fistulous AVM, the resolution of extensive syringomyelia was seen on follow-up imaging.

CMI and associated Hydrocephalus

Hydrocephalus occurs in 3–10% of patients with CMI, particularly in children with long-standing course.[6,16] Interestingly, hydrocephalus was present in 64% of patients harboring acquired CMI secondary to space-occupying lesions and appeared to be obstructive cause by lesions in posterior fossa.[25] In our case, there was no hydrocephalus in the initial course. However, associated hydrocephalus developed due to fourth ventricular outflow obstruction following intraventricular hemorrhage from partially treated brain AVM. In addition, the venous hypertension would also result in impaired CSF resorption.

The management of acquired CMI with syringomyelia

The goals of CMI surgery are restoring the normal flow of CSF across the foramen magnum and reduction the size of syringomyelia.[6] According to a systemic review of the treatments of CMI by Zhao *et al*.,[26] they found that bony decompression plus duraplasty showed the most favorable outcomes, resulting in reduction in the syrinx cavity.

Resection of cerebellar tonsils was not recommended due to high rate of complications. Shunting of the syrinx may worsen clinical signs and symptoms and enlarge the size of associated syrinx.

Congenital CMs should be treated with surgical decompression and duraplasty, whereas acquired CMI may be managed by treating the primary disease first.[2,14] Identification of the exact cause of acquired CMI is the most pivotal component of the management. Acquired CMI treatment should be directed at the underlying cause.[6] CMI caused by cerebral AVM is reversible, if adequate venous drainage can be reconstituted following proper endovascular treatment of the AVM.[19] A thorough understanding of the pathogenesis and natural history of the coexistence of CMI and intracranial AVM are imperative in guiding the proper management of this condition.

In the present study, CMI was misdiagnosed as congenital origin and unusual progressive enlargement of a syrinx cavity occurred following bony decompression and duraplasty. Inadequate decompression of the craniocervical junction should be considered initially. Fortunately, after detecting acquired CMI caused by supratentorial AVM, we treated the primary cause and achieved significant reduction in the syrinx cavity. The goal of endovascular treatment was aiming at reducing the flow rate of the fistulous AVM.

Holocord syringomyelia presenting as foot drop

Foot drop causes predominantly from the peripheral nervous system. CMI and associated holocord syrinx are an extremely rare central cause of foot drop and <10 cases are found in the literature, usually occurring in childhood or adolescence. Most patients complained abrupt onset or rapidly progressive foot drop. Early diagnosis of CMI and holocord syrinx is pivotal and prompt surgical decompression may lead to an excellent prognosis.[8,12,17] In our case in adulthood, acquired CMI was misdiagnosed as congenial origin and subsequent surgical decompression for CMI was performed with resulting in rapidly progressive foot drop later. However, treating the primary cause, that is, brain AVM, of CMI associated holocord syrinx by endovascular treatment can facilitate motor function recovery.

Syringomyelia and neuropathic pain

Persistent central neuropathic pain may cause by intramedullary lesion such as intramedullary hemorrhage.[7] Neuropathic pain may occur in patients harboring CMI associated with syringomyelia and often a devastating residual even after successful surgery.[4] Neuropathic pain is frequently associated with syrinx and may pose treatment challenges.[6] Interestingly, Seki *et al*.[22] investigated the neuropathic pain caused by syringomyelia associated with CMI. They classified the morphology of the syrinx into deviated, enlarged, central, and bulkhead types based on axial MR images from the level corresponding to the dermatomal distribution of pain. They found that the deviated type was associated with a significant neuropathic pain in both preoperative and postoperative states. Pain related to syringomyelia may not respond to treatment, even though the size of the syrinx was reduced.[6] In our case, the syrinx deviated to the right side at the lower spinal cord, probably corresponding with the persistent central neuropathic pain of the right lower extremity.

CONCLUSION

We reported the fifth case of acquired CMI with associated syringomyelia secondary to brain AVM. The development of acquired CMI and holocord syringomyelia may cause by intracranial venous hypertension resulting from supratentorial AVM-induced venopathy. Complication of intraventricular hemorrhage from the AVM may precipitate the occurrence of hydrocephalus requiring CSF diversion. Endovascular treatment of brain AVM achieved successful management of holocord syringomyelia, worsening after suboccipital craniectomy and duraplasty. A thorough understanding of the pathogenesis and natural history of the coexistence of CMI with syringomyelia and intracranial AVM are imperative in guiding the proper management of this entity. The need for cranial imaging in initial evaluation of cases with adult Chiari malformation is important.

Declaration of patient consent

Institutional Review Board (IRB) permission obtained for the study.

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Conflicts of interest

There are no conflicts of interest.

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