Arachnoid cyst resulting in tonsillar herniation and syringomyelia in a patient with achondroplasia

Case report

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Achondroplasia has been associated with varying degrees of cervicomedullary and spinal compression, although usually in the pediatric population. Large arachnoid cysts have also been found to result in tonsillar herniation and syringomyelia. The authors present the case of a patient with achondroplasia who presented with symptoms of foramen magnum compression and syringomyelia, and who was subsequently found to have a large posterior fossa arachnoid cyst.

This 38-year-old woman with achondroplasia presented with an 8-month history of headache and numbness of the hands and fingers. Admission magnetic resonance (MR) imaging of the head and spine revealed a large arachnoid cyst in the posterior cranial fossa, a 6-mm tonsillar herniation consistent with an acquired Chiari malformation, and a large cervicothoracic syrinx. The patient was treated using suboccipital craniectomy, C-1 laminectomy, fenestration of the arachnoid cyst, and decompression of the acquired Chiari malformation with duraplasty.

Surgical decompression resulted in improvement of the presenting symptoms, adequate decompression of crowding at the foramen magnum, and resolution of the syrinx. Although there was only partial reduction in the retrocerebellar cisternal space on follow-up MR imaging, no residual symptoms were related to this.

KEY WORDS • Chiari malformation • syringomyelia • arachnoid cyst • achondroplasia

Achondroplasia, the most common form of bone dysplasia and the best-known form of congenital dwarfism, is caused by a disturbance of endochondral bone formation.^{10,16,19,20,24} Neurological symptoms are identified in as many as 47% of patients with achondroplasia. Symptoms in children include psychomotor delay, hypotonia, feeding and sleep disorders, apnea, macrocrania with or without hydrocephalus, and compressive spinal syndromes.¹⁹ Cervicomedullary compression is also common in the pediatric population^{20,24} and is almost always diagnosed and treated before adulthood. In the adult population of patients with achondroplasia, spinal stenosis with compression of the spinal cord results in many radicular pain syndromes.^{2,15,19} The neurological manifestations can be severe, with extreme cases resulting in death due to hydrocephalus or compression of the brainstem. Patients in all age groups with achondroplasia generally have a small cranial base, whereas the rest of the skull is normal in size. This size discrepancy may predispose patients to anomalous formation of the posterior fossa and foramen magnum, thus creating a smaller than normal posterior fossa.

Syringomyelia has been associated with many intracra-

nial and spinal anomalies. One theory is that syringomyelia results from obstruction of CSF flow at the level of the foramen magnum and the outlet of the fourth ventricle.¹ Obstruction of CSF flow may lead to an increased pulsatile pressure within the spinal canal and may force CSF into the spinal cord.¹⁸ This obstruction is most commonly caused by the Chiari malformation and crowding of the posterior fossa. The Chiari malformation has been traditionally defined as downward herniation of the cerebellar tonsils through the foramen magnum.¹⁴ The average delay in diagnosis of a Chiari malformation is 5 years after the onset of symptoms, leaving ample time for the development of other complications such as syrinx.⁵

Intracranial and posterior fossa mass lesions such as tumors or arachnoid cysts may also result in tonsillar herniation. Arachnoid cysts are benign collections of CSF within an anomalous arachnoid enclosure that account for approximately 1% of intracranial mass lesions.⁶ There have been several reports of large arachnoid cysts in the posterior fossa associated with syrinx.^{4,9,21,23} We describe the case of a patient with achondroplasia who presented with an 8-month history of headache and neurological symptoms. Brain MR imaging revealed a large arachnoid cyst in the posterior fossa that was causing a 6-mm tonsillar herniation and a cervicothoracic syrinx.

Abbreviations used in this paper: CSF = cerebrospinal fluid; MR = magnetic resonance.

CASE REPORT

History. This 38-year-old woman with achondroplasia presented with an 8-month history of occipital shock-like headache that radiated to the temporal regions. The pain was not aggravated by Valsalva maneuvers. Concurrently, she noticed bilateral tone deafness, bilateral tingling of the fingers, numbness in her lower lip, and numbness in the back of her head, neck, and perineal region. She also reported a sharp pain radiating from the left scapular region anteriorly to the chest. Other complaints included dizziness, fatigue, depression, and impaired memory.

Examination. Physical examination revealed short stature with short arms and legs characteristic of achondroplasia. The initial neurological examination revealed diminished gag reflex bilaterally, and decreased sensation to light touch in the V1 and V2 distributions on the right side of the face and diffusely in the right arm and leg. Patellar reflexes were brisk. Results of the remainder of the neurological examination were normal.

A noncontrasted MR image of the brain revealed a large arachnoid cyst in the posterior fossa resulting in herniation of the tonsils 6 mm below the foramen magnum (Fig. 1). The posterior fossa measurements showed a long, thin supraocciput measuring 68 mm (normal 41.8 ± 5.2 mm), the clivus measured 37 mm (normal 40.4 ± 5.1 mm), and a reduced tentorial angle of 70° (normal $82.5 \pm 7.2^\circ$) was observed. Sagittal MR imaging of the cervical spine revealed a large syrinx spanning the C6–T1 vertebral bodies (Fig. 2).

Operation. After review of the neuroimaging studies and discussion of treatment options with the patient, she elected to proceed with surgery. In the operating room, after in-

duction of general anesthesia, the patient was placed prone with her head supported in a Mayfield headrest. Hair covering an 8-cm patch in the midline of the occiput centered on the inion was shaved, and an incision was created from just above the inion to C-2. After the posterior fossa and lamina of C-1 were exposed, a craniotomy was performed extending from just above the inion to the foramen magnum. The C-1 lamina was removed. Opening the posterior fossa and upper cervical dura mater revealed a large arachnoid cyst filled with CSF and encompassing the upper portion of the posterior fossa. The cyst did not extend to the foramen magnum, where the tonsils were impacted and herniated to the level of C-1. The cerebellar hemispheres appeared to be compressed anteriorly.

The posterior wall of the cyst was opened and a portion of the anterior wall was fenestrated into the quadrigeminal cistern on the right (prominent veins on the left prohibited fenestration). Cyst wall excision and fenestration did not change the impacted position of the tonsils, so their inferomedial aspects were gently shrunk using bipolar cautery. A 3×4 -cm triangular pericranial graft was harvested from under the scalp above the inion and incorporated into the dural closure at the foramen. The bone flap was then trimmed to provide a 2×3 -cm craniectomy at the foramen magnum and was reattached with microplates. The wound was closed in layers and the patient was extubated and transferred to the postanesthesia care unit, where she had an uneventful recovery course. She continued to convalesce well and was discharged on the 3rd postoperative day.

Postoperative Course. At her 3-month follow-up review, the patient reported resolution of her occipital and temporal pain, dizziness, and lip numbness, and improvement in

Fig. 1. Preoperative sagittal (left) and axial (right) MR views of the brain revealing a large posterior fossa arachnoid cyst and caudal displacement of the cerebellar tonsils.

Fig. 2. Preoperative sagittal view of the cervical spine demonstrating syringomyelia.

memory and fatigue. She noted persistent pain in the scapular region and persistent numbness in the upper extremities and hands. A postoperative MR image of the brain, although demonstrating only a small decrease in the size of

the posterior fossa fluid collection, revealed less deviation of the brainstem anteriorly and adequate decompression at the foramen magnum (Fig. 3 *left* and *center*). Follow-up cervical MR imaging, which was performed at 6 months, revealed complete resolution of the syrinx (Fig. 3 *right*). The patient's only complaint at the 6-month follow-up review was intermittent intrascapular pain that subsequently improved with physical therapy.

DISCUSSION

There has been one previous report of a patient with achondroplasia, posterior fossa cyst, syringomyelia, and cerebellar tonsillar ectopy,¹⁶ although treatment was not addressed in that paper. Evidence supports the hypothesis that the fundamental problem in the Chiari Type I malformation is a volumetrically small posterior cranial fossa.¹⁴ Indeed, this origin is highly probable in a patient with achondroplasia. The posterior fossa is formed by the endochondral ossification of the cartilaginous frame that forms the cranial base, as well as the first four embryonic somites that form the occipital bone.¹³ In a patient with achondroplasia, these structures would be expected to develop anomalously and to be smaller than normal.

The most common findings are a narrow, anteriorly displaced foramen magnum in a small, shallow posterior cranial fossa, and a small cisterna magna.¹⁶ The size of the foramen magnum in an adult patient with achondroplasia is comparable to its size at birth in an individual without this disorder.⁸ Interestingly, these abnormalities often result in upward displacement of the brainstem, sometimes in conjunction with angulation of the pons and medulla oblongata,^{16,22} possibly explaining the fact that Chiari Type I malformation is somewhat rare in the population with achondroplasia. In our patient, we hypothesize that the large arachnoid cyst in the already small posterior fossa resulted in a mass effect with caudal herniation of the cerebellar tonsils. This then led to obstruction of CSF flow at the foramen magnum and caused the formation of a large

Fig. 3. Postoperative sagittal (*left*) and axial (*center*) brain and sagittal cervical spine (*right*) MR images demonstrating resolution of the syrinx.

TABLE 1
*Literature review of syringomyelia in patients with posterior fossa arachnoid cysts**

Authors & Year	Overview of Study	Patient Age (yrs), Sex	Presentation	Neuroimaging Evaluation	Treatment	Outcome
Banna, 1988	three patients w/ syringomyelia, PF cyst, & hydrocephalus	6, F	presented at age 2 mos w/ weakness of UEs; during treatment for preexisting scoliosis, syrinx & cyst were found	CT myelogram: expansion of spinal cord above T-9, large midline PF arachnoid cyst herniating through foramen magnum & compressing the cord at C-1 & C-2; MRI: holocord syringomyelia	PF craniotomy w/ excision of cyst	improved motor function in both arms; repeated MRI at 1 yr revealed collapsed cord, which did not require further treatment
		14, M	suffered from long-term cerebral palsy; presented w/ progressive lt-sided weakness in UE more than LE (history of traumatic delivery w/ skull fracture); mild lt facial weakness sparing forehead; uvula deviated to rt, tongue deviated to lt; hyperreflexia w/ extensor plantar reflex; dysmetria & past pointing prominent on lt	CT: obstructive hydrocephalus w/ large PF cyst compressing 4th ventricle; no communication btwn cyst & ventricular system; marked dilation of cervical & thoracic spinal cord	PF craniotomy w/ removal of cyst wall & dorsal myelotomy of cervical spinal cord	symptoms stabilized w/ some suggestion of improvement in lt arm weakness; no further progression of patient's condition over 6-yr FU
		2, M	increasing head circumference; Dandy-Walker syndrome diagnosed & VP shunt placed; on evaluation for scoliosis, improvement of hydrocephalus but no resolution of cyst	CT: large midline PF cyst, which did not communicate w/ ventricular system on contrast myelography; myelogram: extensive cervical syringomyelia	cystoperitoneal CSF shunting	CT: full collapse of cyst & syrinx; FU findings demonstrated small 4th ventricle, which suggested that the original diagnosis of Dandy-Walker syndrome was incorrect
Zager, et al., 1990	patient w/ PF arachnoid cyst & syringomyelia, & 2 patients w/ syringomyelia due to Chiari I malformation	9, F	large arachnoid cyst & obstructive hydrocephalus at age 7 wks; after treatment, she presented at age 9 yrs w/ HA & midthoracic back pain; she also had bilat leg weakness & spasticity, & urinary urgency	obstructive hydrocephalus & large midline arachnoid cyst of posterior cranial fossa; open exploration demonstrated syringomyelia from T-3 to T-6	treated at age 7 wks w/ VP shunt, which became disconnected; at age 9 yrs, spinal canal was opened & syrinx drained by needle aspiration; another catheter was inserted into arachnoid cyst & subsequently connected to existing VP shunt, which was functional	papilledema rapidly improved; LE function gradually improved, & at 6 mos patient showed only slightly increased ankle reflexes
Nakai, et al., 1995	patient w/ achondroplasia, PF cyst, syringomyelia, & tonsillar herniation	19, F	HA & numbness of limbs	MRI: displacement of medulla & cerebellar tonsils downward into spinal canal; dilated lat & third ventricles; cerebellum displaced anteriorly by arachnoid cyst; cervical syringomyelia	not reported	not reported
Wakamoto & Kobayashi, 1996	patient w/ PF arachnoid cyst w/ syringomyelia	47, F	history of skull fracture at birth; presented w/ occipitalgia, hydrocephalus, pain in rt shoulder & hand, coughing, & hiccuping	large midline PF cyst w/ syrinx extending from C-1 to T-11	cystoperitoneal CSF shunting	resolution of symptoms & syrinx at 2-mo FU

(cont.)

Arachnoid cyst with tonsillar herniation and syringomyelia

TABLE 1 (continued)
Literature review of syringomyelia in patients with posterior fossa arachnoid cysts*

Authors & Year	Overview of Study	Patient Age (yrs), Sex	Presentation	Neuroimaging Evaluation	Treatment	Outcome
Matsumoto, et al., 1997	patient w/ syringomyelia secondary to cerebral abscess & PF arachnoid cyst	18, M	history of meningitis & brain abscess; presented w/ pain & sensory disturbance of lt shoulder	syrinx from upper cervical to lower thoracic spinal cord	PF decompression w/ opening of foramen of Magendie	symptoms & syrinx improved w/ in 1 mo
Arun Kumar, et al., 1998	two patients w/ PF arachnoid cysts & syringomyelia	15, F	diplopia; HA; nasal regurgitation of fluid; hoarseness of voice; incoordination of lt UE; bilat papilledema & nystagmus; 5th, 6th, & 7th CN paresis on lt side; bilat spasticity of LEs, & increased LE reflexes	MRI: cervicothoracic syringomyelia & large arachnoid cyst in PF w/ anterior displacement & compression of 4th ventricle w/ hydrocephalus; CSF flow study: no flow in the aqueduct or foramen magnum	PF craniectomy, foramen magnum decompression, & marsupialization of cyst	6-mo FU: patient had only mild residual 6th CN paresis; MRI revealed decrease in cyst size & marked decrease in syrinx size
		39, M	bilat LE weakness, sensory loss from C-2 to L-1 on rt, & dorsal column sensory impairment in all extremities; deep tendon reflexes absent in UEs & exaggerated in LEs	MRI: cavitation of spinal cord from C-1 to conus, large arachnoid cyst in PF, hydrocephalus; CSF flow study: no flow across foramen magnum	PF craniectomy, foramen magnum decompression, & marsupialization of cyst	MRI on postop Day 10 revealed restoration of CSF flow across foramen magnum into upper cervical subarachnoid space; syrinx collapsed & symptoms improved
Shinoda, et al., 1998	patient w/ retrocerebellar arachnoid cyst w/ syringomyelia	22, M	drop attacks & hydrocephalus	obstructive ventricular hydrocephalus, huge PF arachnoid cyst, & syringomyelia on MRI	cystoperitoneal CSF shunting	resolution of symptoms; drop attacks ceased; reduction in size of ventricles, cyst, & syrinx
Jain, et al., 2000	patient w/ retrocerebellar arachnoid cyst w/ syringomyelia	14, F	severe HA, vomiting, & loss of consciousness; bilat papilledema; hydrocephalus	large midline retrocerebellar arachnoid cyst w/ compression of 4th ventricle; syrinx extended from C-2 to T-1	suboccipital craniectomy w/ total excision of cyst wall	marked improvement in symptoms; gross reduction in size of retrocerebellar cisternal space; minimal reduction in size of syrinx, but no pre- or postop symptoms related to this
Nomura, et al., 2002	patient w/ PF arachnoid cyst manifesting as cervical syringomyelic myelopathy	32, M	history of cystoperitoneal CSF shunting at 3 yrs of age; presented at age 32 yrs w/ hypesthesia of both hands, finger movement clumsiness, & gait disturbance; reflexes exaggerated in both LEs	MRI at age 3 yrs: large arachnoid cyst occupying most of PF w/ severe compression of cerebellum; MRI at age 32: presyrinx state extending from C-4 to T-1; cyst wall impeded flow of CSF at foramen magnum; shunt functional	neuroendoscopic fenestration of cyst	improvement of CSF flow & resolution of presyrinx state (flow studies demonstrated CSF flow btwn cyst & major cistern, relieving pressure); hypesthesia & gait disturbance subsequently resolved
present study	patient w/ achondroplasia & PF arachnoid cyst w/ syringomyelia & herniation of cerebellar tonsils	38, F	occipital HA; bilat tone deafness; bilat numbness & tingling of fingers; numbness of occipital region & perineum; gag reflex diminished; decreased sensory function on rt in V1, V2, UE, & LE; bilat hyperreflexia	large PF arachnoid cyst w/ herniation of cerebellar tonsils to 6 mm below foramen magnum; syrinx extending from cervical spinal cord to midthoracic cord	PF craniotomy, fenestration of cyst, & duraplasty	3 mos: improvement in all symptoms; 6 mos: complete resolution of syrinx w/ little change in cyst cavity dimensions on MRI

* Btwn = between; CN = cranial nerve; CT = computerized tomography; FU = follow up; HA = headache; LE = lower extremity; PF = posterior fossa; UE = upper extremity; VP = ventriculoperitoneal.

cervicothoracic syrinx.

The pathophysiological origin of syringomyelia has been the focus of several theories. The hydrodynamic theory proposed by Gardner, et al.,⁷ in 1957 hypothesized that syrinx formation is the direct result of blockade of the outlets of the fourth ventricle. This leads to CSF being forced caudally into the spinal cord, with each arterial pulsation caus-

ing increased pressure within the canal and dilation of the cavity, thus forming a syrinx. Another theory, set forth by Williams,²⁵ holds that Valsalva maneuvers lead to increased intraabdominal and thoracic pressure, which is transmitted to the venous system of the Batson plexus and into the spinal subarachnoid space. This causes a net outflow of CSF from the spinal to the intracranial subarachnoid space.

The posterior fossa cerebellar tonsillar herniation acts as a ball valve, restricting the passage of CSF back from the intracranial to the spinal subarachnoid space and creating a pressure differential between the subarachnoid space in the spinal canal and the cranium. The decreased pressure in the spinal subarachnoid space acts as a vacuum, pulling CSF down into the central spinal cord.

Ball and Dayan³ postulated that the pressure in the spinal subarachnoid space may increase with arterial pulsations and with blockage at the foramen magnum, and that CSF is thus forced into the central canal or into a cystic formation through the Virchow–Robin spaces of the spinal cord parenchyma. The Ball and Dayan hypothesis combined with elements of the others seems most appropriate in cases of a posterior fossa arachnoid cyst, and the theory has been substantiated using dynamic CSF flow (or cine) MR imaging studies.¹ In our patient, the cyst and cerebellar tonsils were forced downward with CSF pressure at each systolic beat. The arachnoid cyst resulted in caudal displacement of the cerebellar tonsils, which act as a ball valve at the rostral end of the spinal column, blocking CSF flow in or out of the spinal subarachnoid space. When pressure is increased within the spinal subarachnoid space by Valsalva maneuvers or normal systolic pulsations, CSF is forced into the spinal cord parenchyma and collects to form a syrinx. In this model, the syrinx does not have to be in communication with the central canal and does not necessarily collapse immediately when CSF flow dynamics are restored.

According to recent literature, the most effective treatment of syringomyelia is deactivation of the filling mechanism.¹¹ In our patient the filling and maintenance pressures for the syrinx were maintained by blockage at the foramen magnum. Thus, the focus of the surgical intervention in this case was fenestration of the cyst and expansion of the foramen magnum. Although it is possible that the tonsils would have ascended with cyst fenestration alone, the presence of the syrinx led the treating surgeon to expand the foramen as well. Nomura, et al.,¹⁷ advocate the use of endoscopic fenestration of the lesion in treatment of arachnoid cyst and syringomyelia. Nevertheless, in light of the already deformed posterior fossa secondary to achondroplasia, a full suboccipital craniectomy with C-1 laminectomy, shrinkage of the tonsils, fenestration of the cyst, and duraplasty was performed. This allowed for good relaxation in the region of the foramen magnum and reestablishment of an adequate cisterna magna. With improved CSF flow dynamics, the syrinx began to shrink in size and at 6 months neuroimaging studies revealed that it had completely resolved. The symptoms related to the syrinx also resolved over this time.

A review of the English-language literature located case reports of 12 patients with posterior fossa arachnoid cyst and syringomyelia (Table 1). Of these, six underwent suboccipital craniotomy with fenestration of the cyst and decompression of the foramen magnum.^{1,4,9,12} Symptoms resolved in all cases but one,⁴ in which the patient's symptoms remained unchanged. The syrinx also resolved on follow-up neuroimaging in all patients but one,⁹ in whom there was minimal resolution of the syrinx but improvement symptomatically. Two patients were treated with cystoperitoneal shunt placement, which resulted in decreases in syrinx size and resolution of symptoms in both cases.^{21,23} Two patients were treated with insertion of both ventriculoperitoneal and cystoperitoneal shunts,^{4,26} and symptoms

cleared in both; however, the reports did not mention resolution of the syrinx on neuroimaging. One case was treated with endoscopic fenestration as mentioned earlier,¹⁷ and resulted in resolution of symptoms and the syrinx. These findings lead us to suggest that all of the aforementioned treatments are effective in the management of syringomyelia caused by a posterior fossa arachnoid cyst. Suboccipital craniotomy with open cyst removal offers the benefit of allowing the relaxation of the entire foramen magnum and the opportunity to explore the area to relieve any arachnoid adhesions that may further restrict flow.

CONCLUSIONS

We present a case report summarizing treatment in a patient with the endochondral bone formation defect of achondroplasia, in whom a large arachnoid cyst in the posterior fossa resulted in tonsillar herniation and syringomyelia. The defect in cartilage formation resulted in the development of a long, thin supraocciput, and the cyst caused anterior deviation of the cerebellar hemispheres and brainstem. Herniation of the cerebellar tonsils resulted in obstruction of CSF flow and resultant syringomyelia. Surgical treatment with cyst fenestration, tonsillar shrinkage, and duraplasty resulted in significant clinical improvement and resolution of the syrinx.

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