Resolution of Tonsillar Herniation and Cervical Syringomyelia Following Resection of a Large Petrous Meningioma: Case Report and Review of Literature

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ABSTRACT

We describe a case of a large, petrous meningioma associated with tonsillar herniation and cervical syringomyelia. The patient, a 53-year-old woman, had a 6-month history of a dull, aching pain in the occipital region associated with numbness in the right C2 dermatome and left gaze-evoked nystagmus. Magnetic resonance imaging (MRI) revealed a large tumor in the right posterior fossa associated with moderate supratentorial hydrocephalus. Secondary tonsillar herniation and cervical syringomyelia extending from C2 to C6 were also identified. The tumor, later confirmed to be a meningioma originating from the petrous region, was resected completely via a retrosigmoid approach. Postoperative MRI demonstrated total resolution of the tonsillar herniation and cervical syringomyelia. The radiological features, potential pathophysiological mechanisms, and treatment strategies are discussed in relation to the recent literature.

KEYWORDS: Acquired tonsillar herniation, craniocervical region, posterior fossa, herniation, petrous meningioma, syringomyelia

Acquired cerebellar tonsillar herniation has often been described in association with hydrocephalus and excessive drainage of cerebrospinal fluid (CSF). Syringomyelia has also frequently been re-

ported in patients with congenital craniovertebral anomalies such as basilar invagination and Chiari I malformation. The presenting triad of cerebellar tonsillar herniation and syringomyelia related to

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posterior fossa tumors, however, is uncommon.¹ Earlier literature on this association was based on postmortem reports²⁻⁷ or its suggested presence in myelographic studies.⁸⁻¹⁰ With magnetic resonance imaging (MRI), this association of pathologies has been reported in patients with posterior fossa epidermoid tumors,^{11,12} medulloblastomas,^{13,14} gliomas,¹³ metastatic renal cell carcinomas,¹³ synovialomas,¹⁴ posterior fossa cysts,¹⁵⁻²¹ dysplastic gangliocytomas,²² dysembryoplastic neuroepithelial tumors,²³ and meningiomas (Table 1).^{13,14,24-28} We describe an unusual case of a meningioma originating from the cerebellar surface of the petrous bone associated with both tonsillar herniation and syringomyelia.

CASE REPORT

A 53-year-old woman had a 6-month history of progressively worsening occipital pain, which was exacerbated by coughing and bending forward. On examination, she had a horizontal left gaze-evoked nystagmus and hypesthesia in the right C2 dermatome. MRI showed moderate supratentorial hydrocephalus and a large (4.0 × 4.5 cm), extra-axial, homogeneously enhancing tumor that appeared to originate from the right petrous dura compressing and displacing the lateral recess of the fourth ventricle to the left (Fig. 1). The cerebellar tonsils were herniated down to the C2 level, and a syrinx was centrally located from C2 to C6.

The patient underwent a right suboccipital craniectomy followed by further decompression of the rim of the foramen magnum and a C1-C2 laminectomy (Fig. 2). The tumor, which appeared gray, firm, and vascular, originated along the petrous pyramid. The tumor was resected completely using a retrosigmoid approach, and a lax duroplasty using pericranium was performed. The postoperative period was uneventful.

Histological examination was consistent with the diagnosis of a meningioma. On the second postoperative day, MRI showed tonsillar ascent

with improvement in the size of the syrinx. On discharge from the hospital, the patient reported no occipital pain and the extent of numbness over the C2 dermatome was remarkably reduced. One year after surgery, MRI showed complete resolution of the previously herniated tonsils and syrinx cavity (Fig. 3). The patient's neurological examination was normal.

DISCUSSION

The association of tonsillar herniation and syringomyelia with posterior fossa tumors is the exception rather than the rule. Large posterior fossa tumors often manifest with elevated intracranial pressure, but most have no associated tonsillar herniation or syringomyelia. In a multicenter retrospective analysis, Tachibana and colleagues¹³ reviewed MRI data from 164 patients with posterior fossa tumors. Of these 164 patients, 24 also had tonsillar herniation. Of these 24 patients, only 5 patients presented with the triad of a posterior fossa tumor, tonsillar herniation, and syringomyelia. Of these 5 patients, 2 had meningiomas.

During a 16-year period, Klekamp and associates¹⁴ treated only 3 patients who had both posterior fossa tumors and an associated syringomyelia. Only 1 of these 3 patients had a meningioma. In another review of 54 cases of syringomyelia,¹ fewer than 1% of the patients presented with secondary hindbrain herniation related to brain or meningeal tumors of the posterior fossa. In the same report, the author noted a different retrospective analysis of 545 cases of syringomyelia of which only 4 patients presented with both syringomyelia and hindbrain herniation associated with an intracranial tumor. However, this report did not specify whether the intracranial tumors were supratentorial or infratentorial.

The advent of MRI has been immensely helpful in the diagnosis of this phenomenon and has the potential to improve the understanding of the pathophysiological events that cause tonsillar

Table 1 Posterior Fossa Meningiomas with Associated Tonsillar Herniation and Syringomyelia

Authors	Age/ Sex	Tumor Location	Presenting Symptoms Related to:		Duration of Symptoms	MR-Syrinx / Hydrocephalus	Surgery	Follow-up
			Tumor	Syrinx				
Fukui et al ²⁴	66/F	Left CPA	+	-	3 years	C2-C4/NA	Retromastoid craniectomy, complete resection	Reduction in syrinx at 2 months
Klekamp et al ¹⁴	17/M	Posterior fossa	NA	NA	NA	Cervical*/NA	VP shunt, complete tumor resection, removal of arachnoid adhesions	Reduction in syrinx at 2 weeks
Tachibana et al ¹³	53/F	Tentorial	+	-	3 years	Cervical*/+	Complete tumor resection	Reduction in syrinx unknown time frame
Anegawa et al ²⁵	50/F	Petrous pyramid	+	-	3 months	Cervical*/+	Complete tumor resection	Reduction in syrinx at 3 months
	36/F	Posterolateral pyramid	+	-		Cervical*/NA	Complete tumor resection	
Jaiswal, Chandra ²⁶	30/F	Right CPA	+	+	5 years	Cervical*/+	Retromastoid craniectomy, complete tumor resection	NA
Karttunen et al ²⁷	34/F	Tentorial	+	-	NA	C2-T2/-	Suboccipital craniectomy, complete tumor resection	Reduction in syrinx at 22 months
Bhat ²⁸	38/F	Right tentorial	+	-	6 months	Cervical*/+	Retromastoid craniectomy, complete tumor resection	Resolution of syrinx at 3 months
Present case	53/F	Petrous pyramid	+	-	6 months	C2-C6/+	Retromastoid craniectomy, complete tumor resection	Resolution of syrinx at 1 year

CPA, cerebellopontine angle; NA, information not available; *, extent of syrinx not reported; VP, ventriculoperitoneal

Figure 1 High-resolution preoperative MRI shows a hyperintense posterior fossa mass (meningioma) in right petrous region and an associated central cervical syrinx (top row) and tonsillar herniation extending to about C2 (bottom right); the syrinx extends centrally from C2 to C6 (bottom left).

herniation and syringomyelia.^{11,26,29} Serial pre- and postoperative images also help formulate the surgical plan and follow-up management strategies, respectively, thus avoiding unnecessary or additional surgical procedures for the treatment of tonsillar herniation and syringomyelia.

The clinical symptomatology of patients with posterior fossa meningiomas associated with tonsil-

lar herniation and syringomyelia is interesting. Most of these patients develop symptoms related to the tumor rather than to the syrinx, which was asymptomatic in all but one of our cases (Table 1). Syrinx formation appears to be inversely related to the development of symptoms. Syrinx formation is thought to occur relatively late during tumor progression. More rapidly dividing tumors have a

Figure 2 Intraoperative photograph shows surgical exposure after suboccipital craniotomy and C1-C2 laminectomies. Note herniated cerebellar tonsil (star) below the level of the foramen magnum (rectangles).

greater and earlier impact on the adjoining brainstem and cerebellum than slower-growing tumors, causing tumor-related symptoms before a syrinx forms. In contrast, slower-growing tumors permit more compensation than fast-growing tumors, resulting in the late appearance of symptoms. Furthermore, when symptoms attributable to syringomyelia occur, they are most often observed in more indolent tumors such as meningiomas and posterior fossa epidermoid tumors.^{11,13} Tumor size did not appear to correlate directly with syrinx size, and syrinx size did not correlate directly with the manifestation of symptoms related to the syrinx.

The proximity of the tumor in relation to the CSF pathways also affects the clinical profile. Intrinsic posterior fossa tumors such as astrocytomas

and medulloblastomas with tonsillar herniation tend to become symptomatic after a relatively short duration. They are seldom associated with syringomyelia, possibly because of their proximity to the fourth ventricle. Obstructive supratentorial hydrocephalus may further accentuate the impaction of the tonsils in the region of the foramen magnum. The size of the extrinsic tumor in our patient gradually increased and produced a subacute to chronic tonsillar herniation associated with cervical syringomyelia. Therefore, the presence of tonsillar herniation and syringomyelia in tandem or in isolation may have a direct temporal relationship to the location and nature of the tumor.

While it is fairly clear that syringomyelia formation is a late or chronic event when associated with posterior fossa tumors, the underlying pathophysiological mechanism remains uncertain. The congenital and acquired craniovertebral anomalies associated with syringomyelia have been a principal substrate for elucidating the pathophysiology of syrinx formation and CSF circulation around the foramen magnum and spinal subarachnoid space (SAS). Furthermore, it has been speculated that the mechanism underlying the development of syringomyelia in brain tumors associated with tonsillar herniation is analogous to that underlying congenital Chiari I malformations.

In both cases, the tumor per se is not the direct causative factor of syrinx formation. As the size of the tumor increases, it displaces the adjacent brain and compresses CSF pathways and vasculature within the intracranial compartment. This primary mass effect results in the secondary extrusion of the cerebellar tonsils downward into the foramen magnum and upper cervical canal. The process can be accentuated by progressive hydrocephalus or a small posterior fossa, which may be a primary developmental anomaly.^{7,30-34} Although the volume of the posterior cranial fossa was relatively normal in our patient, the presence of a large extrinsic tumor, adjoining cerebellar edema, progressive supratentorial hydrocephalus, and the downward displacement of the tentorium all combined to reduce the volume of the posterior fossa

Figure 3 High-resolution postoperative MRIs at 1-year follow-up show complete tumor resection (left) and resolution of previous cerebellar tonsil herniation and syrinx cavity (right).

and may have contributed to the occurrence of this phenomenon.

Various theories on the pathogenesis and progression of syringomyelia have been proposed. The prominent early theories were the “hydrodynamic” theory proposed by Gardner and colleagues and the “craniospinal dissociation” theory proposed by Williams and colleagues. The former initially implicated a congenital defect in the fourth ventricle while the latter pointed to pressure differentials above and below the foramen magnum. Although the theories differ, both ultimately assume that syrinx formation is the result of a diversionary flow of CSF through a patent obex and into the central canal of the spinal cord.^{35–37}

Among the previously reported cases of posterior fossa meningiomas associated with tonsillar herniation and syringomyelia (Table 1), Fukui and associates,²⁴ Tachibana and colleagues,¹³ and Karttunen and coworkers²⁷ concluded that syringomyelia formation in their cases was the result of a dissociation of craniospinal pressure, diverting intracranial CSF through a patent communication into the central canal. However, Tachibana et al¹³

found no such communication on radiographic studies. Likewise, Aneawa and associates²⁵ and Klekamp and colleagues¹⁴ found no such communication on MRI studies of their patients. Recent studies have confirmed the lack of a patent communication between the syrinx and the central canal in adults.^{6,7,14}

Lhermitte and Boveri³⁸ postulated that the tumor interfered with the blood supply of the upper cervical cord, causing softening and cavitation in the spinal cord. Tauber and Langworthy³⁹ suggested that the decrease in the lumen of the anterior spinal artery with subsequent ischemia was responsible for the origin of syringomyelia. Others have postulated that increased CSF pressure waves caused by arterial or venous pulsations decreased reabsorption of CSF in the spinal SAS, or that a piston-like action of the cerebellar tonsils during systole below the level of obstruction has the potential to increase the flow of extracellular fluid or cause it to dissect the central cavity along the perivascular spaces of Virchow-Robin or dorsal roots.^{33,40,41}

The incidence of syringomyelia associated with tumor-induced tonsillar herniation is about

21%.^{13,14} This relatively high incidence suggests that a pre-existent Chiari I malformation in these patients is unlikely.³³ The genesis and progression of syringomyelia in association with posterior fossa tumors are thought to be tertiary phenomena that follow blockage of CSF flow at the foramen magnum and alterations in the passage of extracellular fluid in the spinal cord related to occlusion of the SAS at the foramen magnum.^{7,13,14,31,33,42}

In patients with posterior fossa tumors and tonsillar herniation, the tonsils occlude the SAS around the area of the foramen magnum. In this location, the tonsils move downward with each systolic increase in pressure. The movement produces pressure waves along the surface of the spinal cord. The pressure waves compress the spinal cord and have the potential to propel the syrinx fluid downward with each pulsating beat, thereby increasing the flow between the spinal CSF and the extracellular fluid of the spinal cord.³³

This mechanism also may be responsible for the maintenance of syringomyelia by the pulsatile pressure waves forcing CSF into the spinal cord through perivascular and interstitial spaces.^{40,41} The compression of the spinal cord exerted by spinal CSF causes rostral and caudal movement of the fluid within the syrinx and could extend the length of the syrinx via a sloshing mechanism.³³ The maximum pulsatile pressure waves are known to occur in the upper cervical canal and dissipate with increasing distances down the canal.¹

Anegawa and coworkers²⁵ proposed that tonsillar herniation in their patient with a posterior fossa meningioma resulted in a pressure disturbance that forced CSF across the medulla into the central canal creating a syrinx. Subsequently, the obstructed central canal would prevent CSF from escaping the syrinx. In contrast, Klekamp and associates¹⁴ suggested that alterations in CSF flow in their patient with a posterior fossa meningioma increased the flow of extracellular fluid, which accumulated and formed a syrinx.

There are problems with all of the proposed theories, but most authorities would agree that some synthesis of these theories plays a role in the

development of a syrinx in patients with posterior fossa tumors. Almost all of these theories require spinal SAS pressure to be greater than syrinx pressure, but direct measurements have reflected the opposite gradient in the cases studied.⁴³⁻⁴⁶

Recently, a new theory of syrinx formation has been proposed. In this theory, Levine⁴⁶ noted that obstruction of the SAS at the foramen magnum creates a pressure differential above and below the obstruction. This differential favors dilation of vessels below and collapse of the vessels above the obstruction. He further explained that the spatial changes in the caliber of vessels produces chronic mechanical stress on the spinal cord, particularly below the obstruction, which also would be affected by the dynamic physiological pressure changes in the body (e.g., Valsalva). Over time these stresses disrupt the blood-spinal cord barrier and result in the release of fluid (ultrafiltrate) and the formation of a syrinx. Some components of this theory also might contribute to syrinx formation in patients with tumors of the posterior fossa. However, the exact mechanism still remains elusive.

What do these pathological mechanisms and the reported operative experiences reveal about treatment options for patients with this triad? In our case, complete resection of the posterior fossa meningioma resulted in the gradual ascent of the cerebellar tonsils as well as the reduction and disappearance of the syrinx as demonstrated by the patient's satisfactory clinical improvement and serial MRIs. In the other reported cases (Table 1), complete resection of posterior fossa tumors alone promoted resolution and improved the herniation, size of the syrinx, and symptoms.

The extirpation of the posterior fossa tumor eliminates the source of mass effect in the posterior fossa. This, in turn, increases the relative size of the posterior fossa and decreases the downward force placed on the cerebellar tonsils. The amount of impaction at the foramen magnum is diminished, which allows them to return to their original position within the posterior fossa. Elimination of the SAS obstruction and craniospinal pressure dissociation also permits CSF flow through the

subarachnoid space and basal cisterns to return to normal.

To further insure that CSF flow remained unobstructed, we, as did Klekamp and colleagues,¹⁴ decided to enlarge the area around the foramen magnum further using a dural graft. A C1-C2 laminectomy aided in overall decompression. Unfortunately, in most of the reported cases, the surgical procedures are not described with sufficient detail to compare or contrast operative methods. However, in other studies using cardiac-gated CSF flow imaging studies, resolution of CSF flow dynamics has been associated with improvements in tonsillar herniation and syrinx size.⁴⁷⁻⁵⁰

Although the exact mechanism of syrinx formation is still unknown, removal of the tumor and insuring that the CSF pathways are clear for SAS flow undeniably returns CSF flow dynamics to their normal state. This alone is sufficient to promote resolution of the syrinx, as seen at 1 year in our case. Furthermore, after complete removal of a posterior fossa tumor, there has been no report of an additional surgical procedure needed to treat a residual syrinx. However, in an isolated case of a right parietal meningioma, decompressive surgery for a Chiari I malformation was performed to treat persistent upper cervical cord symptoms.⁵¹

CONCLUSION

The association of petrous meningioma with tonsillar herniation and syringomyelia is uncommon. Although the exact mechanism of syrinx formation is still unknown, the blockage of normal CSF circulation pathway at the foramen magnum is the crucial factor. Surgical extirpation of the tumor restores normal CSF circulation at the foramen magnum and produces an excellent outcome. In most cases, the need for an additional surgical procedure to treat associated tonsillar herniation and syringomyelia can be avoided completely.

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Commentary

In this interesting case report, a patient with a large posterior fossa meningioma presented with a Chiari I malformation and cervical syringomyelia. The Chiari malformation and syrinx resolved after the tumor was removed and a decompressive craniectomy, C1-C2 laminectomy, and duraplasty were performed.

As the authors noted, this triad is rare, despite the incidence of posterior fossa meningiomas. Consequently, this patient's posterior fossa may already have been small. The authors treated both pathologies within the same surgical setting. Not surprisingly, these abnormalities resolved completely.

Chandranath Sen, M.D.¹

Commentary

Mr. Fox and colleagues present a well-written and well-illustrated case report along with a thorough review of the literature highlighting the potential for tumor-associated hindbrain herniation to lead to secondary spinal cord syrinx formation. While a rare association, it is one that should be recognized by skull base surgeons, particularly to avoid the mistake of initiating direct treatments for the syrinx, which almost always resolves over time

once the hindbrain herniation has been reduced and normal flow of cerebrospinal fluid (CSF) and pressure equilibration dynamics have been restored.

Indeed, we are increasingly recognizing that spinal cord syringes are almost always secondary phenomena. They usually improve or resolve after successful treatment of their primary cause. This holds true for lysing spinal canal arachnoid adhesions for post-traumatic or postmeningitic syringes; releasing caudal cord tension for tethered cord syringes (e.g., thickened filum, lipomenigocele, or scarred myelomeningocele repair); removing the spinal cord tumor for tumor-associated syringes; posterior fossa decompression of Chiari malformation-induced syrinx; anterior decompression for ventral compression or instability-induced syrinx (e.g., platybasia, C1-C2 rheumatoid arthritis); removing the shunt for lumboperitoneal shunt-induced syringes; and shunting the ventricles for hydrocephalus-induced hindbrain herniation leading to spinal cord syrinx. Removing a mass lesion (e.g., tumor) leading to hindbrain herniation should now be added to that list. Restoring normal flow of CSF and pressure equilibration dynamics throughout the craniospinal axis should be the goal. Primarily shunting or fenestrating a spinal cord syrinx should become a rare procedure, reserved for the small subset of cases where chronicity of the condition does not allow sufficient spontaneous resolution.

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