

Case Report

De Novo Chiari I Malformation and Syringomyelia Arising from Enlarging Posterior Fossae Meningioma: Case Report and Discussion

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Received: June 14, 2021; Accepted: July 13, 2021;

Published: July 20, 2021

Abstract

The authors believe this is the first described case completely illustrating the de novo development of Chiari I Malformation and associated syringomyelia in a patient with posterior fossae meningioma. The case demonstrates and supports current theories regarding Chiari I malformations and the pathophysiology of syringomyelia.

Keywords: Chiari I malformation; Pathophysiology; Meningiomas

Introduction

Syringomyelia can be associated with pathology in the craniocervical junction; however, the scenario of a Chiari I Malformation and syringomyelia arising from a posterior fossae meningioma is infrequent. We describe discuss a case of an enlarging infratentorial meningioma with radiographic progression showing de novo development of a Chiari I Malformation and syringomyelia.

Case Presentation

This patient is a 64-year-old woman with a history of breast carcinoma who was found to have two posterior fossae meningiomas during workup for a syncopal episode in 2007. A leS-sided cerebellar meningioma near the tentorium measured approximately 3cm and, on the right, there was a smaller cerebellar meningioma measuring approximately 0.5cm. There was no associated edema or mass effect; there was no evidence of cerebellar tonsillar herniation or syrinx (Figure 1 and 2). She was asymptomatic and was administered serial scans.

Serial studies did show a progressive increase in the size of both meningiomas, but she remained asymptomatic until 2012. That year she noted occasional frontal headaches when she coughed; however, she denied any other symptoms. Her January 2012 MRI revealed a slight increase in the leS-sided meningioma measuring 3.6 x 3.1 x 3.8 cm. There was some mild mass effect without any evidence of vasogenic edema. The right-sided meningioma measured 1.7 x 1.4 x 1.8 cm, also slightly increased. Resection of the leS-sided lesion was recommended, but she declined. A year later, a February 2013 MRI showed minimal to no increase in size. She was relatively asymptomatic and opted for close follow up.

In February 2014, there were no major changes in her condition. She denied persistent headaches; however, she did again have a component of tussive headaches, which was apparent with coughing and sneezing. In addition, Valsalva maneuvers would also reproduce her symptoms, which was not apparent before. She experienced pressure on the back of the head and frontally. She denied neck pain, paresthesias, dysesthesias, or sensory changes in the extremities. She denied balance, fine motor, or gross motor changes or deficits. The

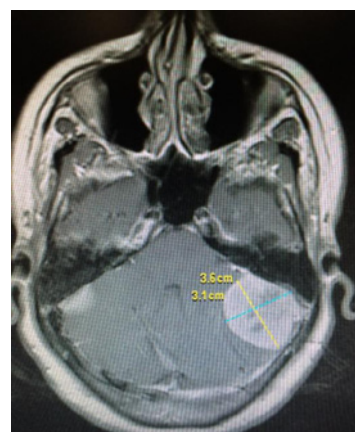


Figure 1:



Figure 2:

exam was normal.

MRI of the brain with and without gadolinium obtained just prior to her office visit in February 2014 suggested that the two meningiomas had enlarged. The leS-sided mass measured 4.4 x



Figure 3:

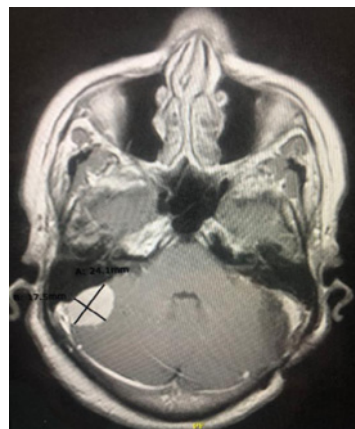


Figure 6:



Figure 4:

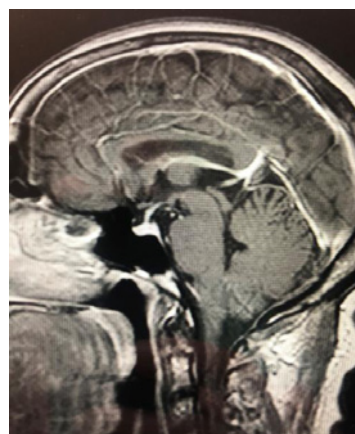


Figure 7:



Figure 5:



Figure 8:

3 x 4 cm; the right-sided mass measured 2.3 x 1.6 x 2.2 cm. There was interval development of moderate ventricular dilatation with transependymal edema and tonsillar herniation consistent with a new Chiari I malformation (Figure 3 and 4). There was a suggestion of a new cervical syrinx.

A dedicated MRI of the cervical spine with and without

gadolinium was obtained. This study showed mild underlying cervical spondylosis and, most impressively, a non-enhancing syringomyelia extending from C2 down to T3 (Figure 5).

A suboccipital craniectomy for resection of the leS meningioma was recommended and performed on March 19, 2014. She did well, and her preoperative symptoms completely resolved. Postoperative

MRIs showed complete resection of the leS cerebellar meningioma and subsequent resolution of her Chiari and syrinx (Figure 6-8).

Discussion

To our knowledge, this is the first case report with documented sequential enlargement of a posterior lesion resulting in Chiari I malformation and syrinx with the resolution of both pathologies post-resection. There are other reports of infratentorial lesions causing both Chiari I and syringomyelia with similar postoperative courses. Posterior fossae tumors associated with syringomyelia or syringobulbia include meningiomas [1-8], Cerebellar astrocytoma [9], brainstem glioma [10], fourth ventricular epidermoid tumor [11], and cerebellar hemangioblastoma [12]. However, patients in these specific reports present with the pathologies; there are no images prior to the appearance of the tonsillar herniation or syrinx.

This report is unique in the radiographic evolution demonstrating the de novo development of Chiari I and syrinx with the corresponding progression of tumor size.

There are multiple theories that attempt to explain the pathophysiology of syringomyelia. Broadly, there are three types of spinal cord cavities/syrinxes showing three types of communication with the fourth ventricle and central canal: communicating and non-communicating central canal syrinxes, and extracanalicular syrinx [13-15]. An extracanalicular syrinx is associated with spinal cord trauma and injury [16]; however, intramedullary spinal tumors with associated syrinx must always be ruled out.

A communicating syrinx or hydromyelia is predominantly found in children with hydrocephalus [15]. Syringomyelia or a non-communicating central canal syrinx is a cavity consisting of a focal dilation of the central canal, separated from the fourth ventricle by a syrinx-free segment of the spinal cord. This cyst occurs primarily in adult patients with disruption of CSF flow near the cervicomedullary junction. Pathophysiology of a syringomyelia has been historically debated.

Initial theories assumed a communication between the fourth ventricle and the central canal. For instance, Gardner's Theory [17] suggests that the flow of cerebrospinal fluid is disturbed by a congenital imperforation of Magendie's foramen. A water-hammer effect is then produced due to the Cerebrospinal fluid movement (CSF) out of the cisterna magna, leading to augmentation of the central canal from the fourth ventricle, producing a syringomyelia. Williams and Timperley [10] reported three cases of syringomyelia with brainstem gliomas and postulated that an obstruction of the subarachnoid space at the foramen magnum could act as a check valve, allowing CSF to cross above the foramen magnum more readily than below, resulting in a craniospinal pressure dissociation. Activities, such as coughing, exacerbate this pressure gradient resulting in spinal fluid flowing from the fourth ventricle into the central canal [18-21], resulting in syringomyelia.

These hydrodynamic hypotheses are based on the theoretical presumption that there must be a direct flow of CSF from the ventricle to the central canal. Neuropathological investigations [13,22] and radiographic studies [15] show that a communication between the syrinx and the fourth ventricle does not exist in syringomyelia, only in the hydromyelia. One explanation is that syringomyelia fluid arises

from the CSF in the spinal subarachnoid space [23]. Ball and Dayan [24] emphasize the importance of activities that increase thoracic or abdominal pressure. The subsequent increase in epidural venous pressure and spinal CSF pressure causes the CSF to seep along Virchow-Robin spaces into the spinal cord to form a syrinx. Aubin et al. [25] documented the transneuronal movement of cerebrospinal fluid into the spinal cord by comparing the CT density of the spinal cord, subarachnoid space, and syringomyelic cavity.

Foramen magnum tumors can divert the spinal CSF away from the intracranial subarachnoid space and into the spinal cord parenchyma along the Virchow-Robin space [23]. Phase-contrast/cine MRIs indicate that an irregular flow of cerebrospinal fluid in the subarachnoid space of the spinal cord may force fluid into the central canal through the interstitial space of the spinal cord [14].

A similar phenomenon occurs in Chiari I malformations when cerebellar tonsils obstruct the foramen magnum. Heiss et al., Levy et al., and Oldfield et al. [26-28] suggest that the cardiac cycle creates pressure fluctuations and pulsations that drive spinal subarachnoid CSF into the cord parenchyma. In Chiari I, the amount of CSF crossing the cranial-cervical junction decreases. The tonsils move inferiorly during systole and superiorly during diastole driving the spinal subarachnoid CSF like a piston. The resultant high-amplitude pulsations drive CSF into the cord along perivascular spaces.

Levine [23] proposes an alternative explanation for the formation of syringomyelia associated with a lesion at the foramen magnum. The differentially higher CSF pressure above the block at the foramen magnum results in changes in the transmural venous pressure. Erect posture, coughing or straining, and pulsatile fluctuations of CSF pressure during the cardiac cycle will transiently widen the differential. This results in capillary constriction and vasodilation above and below the block at the foramen magnum respectively, eventually causing partial breakdown of the blood-spinal cord barrier, allowing ultrafiltrate of crystalloids and low protein to accumulate within the spinal cord, forming a syrinx.

Likewise, Koyanagi [29] believes that in the setting of a Chiari I malformation and tonsillar herniation, there is reduced compliance of the posterior spinal veins and the spinal subarachnoid space resulting in diminished absorption of extracellular fluid through the intramedullary venous channels causing formation of syringomyelia [30-35].

Surgical resection of tumor re-establishes CSF flow with resolution of both Chiari I and syrinx [2,6].

Conclusion

This case report definitively illustrates how an enlarging posterior fossae meningioma can result in a de novo development of a Chiari I malformation and associated syringomyelia. Resection of the tumor resolved the tonsillar herniation and re-established CSF flow resulting in the disappearance of the syrinx. Although generally accepted as true, this case report further supports the pathophysiology of the spinal cord syrinx development as crowding and subsequent disruption of CSF flow at the craniocervical junction.

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