

Posterior Fossa Arachnoid Cyst Associated with Chiari I and Syringomyelia

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The association of posterior fossa (PF) lesions with syringomyelia has been described occasionally both for solid and cystic abnormalities. Few cystic lesions of the PF have been found in association with syringomyelia, and even fewer are believed to cause cavitation of the spinal cord by descent and impaction of the cerebellar tonsils into the subarachnoid space at the level of the foramen magnum (FM). We describe a case of a PF arachnoid cyst associated with impairment of the cerebrospinal fluid (CSF) flow at the FM and syringomyelia.

CASE REPORT

This 61-year-old woman had delayed developmental milestones, and never used her right arm and hand to the full extent. She reported that at an early age 'electrical treatments' were given to improve the condition of her right arm. It was predicted that she would have great difficulty with her studies, yet she obtained a university level education. In 1994 she presented with her longstanding weakness of the right arm, as well as numbness of the right side of the face extending to the ipsilateral shoulder region. The patient was managed conservatively until early 2006. At this time she complained of disagreeable sensations of her right face and right upper limb, and was noted to have 4/5 weakness of the right shoulder abductors and elbow flexors.

When she came to surgical attention prior investigations had identified a 75 x 35 mm arachnoid cyst, extending from the right sigmoid sinus to the clivus, with compression and deformation of the cerebellum, IV ventricle and brain stem. There was no hydrocephalus. Crowding of the FM area was observed, but there was no overt descent of the tonsils below the FM. An 8 mm in diameter syrinx was seen from the cranio-cervical junction down to T10 (Figure 1).

Two weeks before her scheduled surgery she complained of double vision and extension of the numbness down the right upper and lower limb. She was taken to the operating room and an endoscopic fenestration of the cyst was performed February 2006. Her double vision disappeared and so did the leg symptoms, but her distressing paresthesias were still present. An MR CSF flow study revealed minimal CSF flow at the FM. In June 2006 the patient underwent a right PF craniotomy for a larger fenestration of the arachnoid cyst into the cisterns of the posterior fossa.

There was no significant change in her symptoms. The arachnoid cyst decreased minimally in size. Later, she reported swallowing and breathing difficulty, and it was hard for her to walk and talk simultaneously. In October 2006 a FM decompression with duroplasty was performed. Extensive

adhesions were found between the tonsils, medulla, upper spinal cord and arachnoid/dura.

In January 2009 the patient was stable from a clinical standpoint. The syrinx cavity now had a transverse diameter of 4 to 5 mm (Figure 2). A telephone interview June 2009, found the patient very satisfied with her progress: face and right arm sensory complaints were tolerable, the voice strong, and she was able to function and mobilize with no difficulty. The left arm and hand continue to be dominant.

DISCUSSION

In 2007 Martinez-Lage et al identified 15 published cases of syringomyelia accompanying diverse cystic processes of the PF. In 14 of those the cyst itself, or part of the cyst, had entered the FM and blocked the CSF flow in the subarachnoid space. In only two cases, including one of theirs, was the obstruction of the CSF flow at the level of the FM caused by impaction of the cerebellar tonsils, as in Chiari I¹. The Table lists these two cases, a third published in 1995, and the present case.

Either of the two mechanisms could lead to cavitation of the spinal cord and syrinx formation¹⁻³. In the present case the objectively demonstrated impairment of CSF pulsation across the FM was caused by impaction of the cerebellar tonsils into the FM subarachnoid spaces, worsened by extensive subarachnoid adhesions. We believe it is justified to describe this tonsillar impaction as a Chiari I anomaly as crowding of the CSF spaces with no descent of the tonsils has been observed in 9% of symptomatic Chiari I anomalies⁴.

It is conceivable that, in our case, the cyst pushed the tonsils down, creating a Chiari I condition which then gave rise to the syrinx. Other factors, for example an hemorrhage, possibly related to the arachnoid cyst, might have been responsible for the adhesions observed at the tonsillar level. And, if so, we do not need to postulate the presence of a Chiari I anomaly. At any rate the main causative mechanism of spinal cord cavitation is impairment of the CSF flow in the subarachnoid space at the level of the FM, and this was demonstrated in our case¹⁻⁴.

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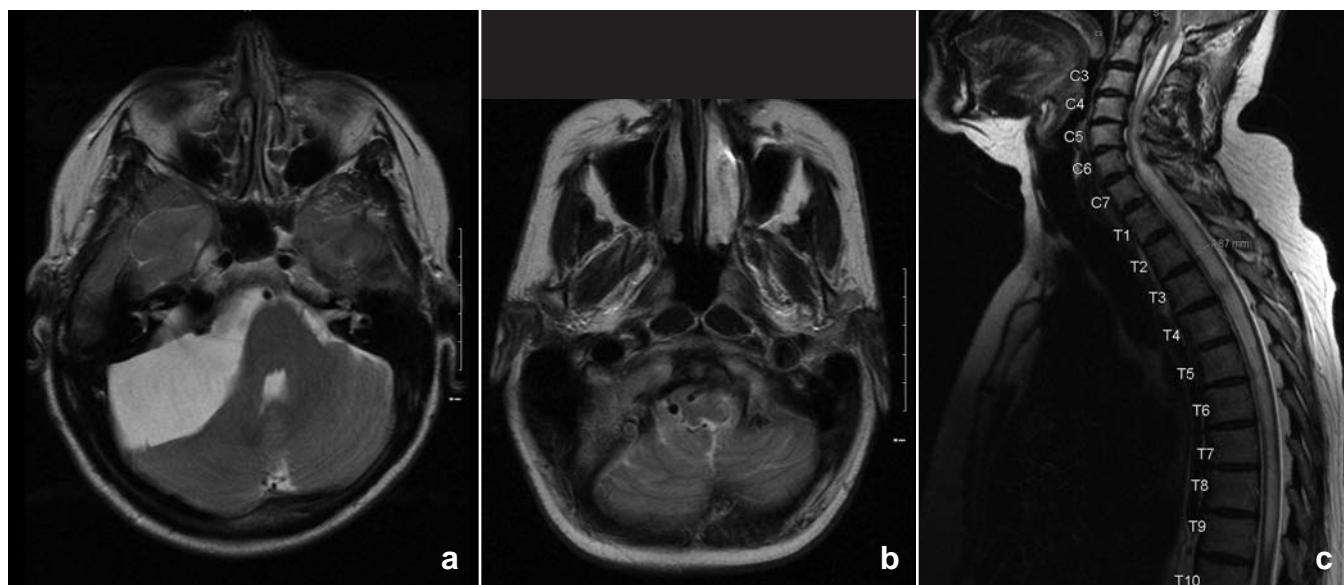


Figure 1: a) T2 weighted axial view of the Posterior Fossa Arachnoid Cyst with compression of the cerebellum, brain stem and IV ventricle. b) Same sequence at the level of the FM, showing crowding of the CSF spaces. c) Syrinx extending from C1 to T10 on Sagittal T2 weighted image.

Our patient came late to neurosurgical attention. The primary cause of the patient's problem was thought to be the arachnoid cyst, and treatment was directed to the cyst. The two cyst fenestrations were unsuccessful in decreasing the progression of the symptoms, but did resolve her double vision, which was probably secondary to stretching of the right VI cranial nerve. The fenestrations did not change the dimensions of the syrinx, likely because of the advanced CSF block at the subarachnoid spaces of the FM, with the tonsils fixed in place by adhesions to the meninges.

Only a FM decompression with extensive lysis of the subarachnoid adhesions followed by an augmentation duroplasty succeeded in stopping the patient's clinical progression, and in reducing the diameter of the syrinx. That there was subsequently a reduction in the dimensions of the syrinx indicates successful reestablishment of the FM CSF pathways. Our experience is in agreement with the literature: of the 12 cases of PF arachnoid cyst and syringomyelia compiled by Bauer et al, six underwent suboccipital craniotomy with fenestration of the cyst and decompression of the FM, with resolution of complaints

Table: Syringomyelia secondary to impairment of FM CSF subarachnoid flow caused by Posterior Fossa Arachnoid Cyst (AC) and Chiari I anomaly

| Year | Reference | Pathology | Tonsillar Descent | Treatment |
|------|----------------|---|-------------------|-------------------------------------|
| 1995 | 5 | Achondroplasia Supra and Retrocerebellar AC | Yes | ? |
| 2005 | 2 | Achondroplasia Retrocerebellar AC | Yes | FM Decompression AC Fenestration |
| 2007 | 1 | Hydrocephalus Retrocerebellar AC | Yes | ETV FM Decompression |
| 2009 | (Present case) | Anterocerebellar AC | No | AC Fenestration FM Decompression |

ETV = endoscopic third ventriculostomy

observed in five². It is probable that, in our case, performing the FM decompression first, would have improved the symptoms sooner, and would have obviated the need for other interventions.

We used the patient's response and MR imaging of the syrinx as our primary endpoints. Magnetic resonance CSF flow studies across the FM could have been performed before and after each intervention, and this would have strengthened our conclusions.

CONCLUSIONS

In lesions of the PF symptomatic syringomyelia may result from CSF blockage at the level of the FM subarachnoid spaces. In cystic PF lesions this may be done directly by the cyst, or indirectly by downward displacement of the cerebellar tonsils. In the present case there was no clear tonsillar herniation, but the tonsils were impacted at the level of the FM and fixed in place by adhesions. Although treatment should be individualized, the obliteration of the subarachnoid spaces at the level of the FM is the central mechanism of production of a syrinx, and FM decompression may prove the most successful therapeutic maneuver.

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Figure 2: Sagittal imaging of the spine following FM decompression, showing a decrease in the diameter of the syrinx.