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M Banna

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Syringomyelia in Association with Posterior Fossa Cysts

Mohamed Banna¹

This paper presents three patients with a triad of syringomyelia, midline posterior fossa cysts, and hydrocephalus. In the first patient, the clinical presentation was related to spinal cord cavitation, and the cranial anomalies were unexpected. In cases 2 and 3, the brain anomalies dominated the clinical picture, and syringomyelia was unexpected. These cases show that an examination of the whole neuraxis is as important in patients with midline posterior fossa cysts as it is in patients with developmental syringomyelia or Chiari I malformation.

The causes of syringomyelia are well documented [1]. They include ectopia of the cerebellar tonsils, arachnoidal adhesions at the foramen of Magendie, intramedullary tumors, spinal trauma, and adhesive arachnoiditis. Few cases are idiopathic, and a case has been reported in association with nontraumatic spinal intradural arachnoid cyst [2]. The purpose of this article is to describe three cases with a triad of obstructive hydrocephalus, midline posterior fossa arachnoid cysts, and syringomyelia. In case 1, the initial symptoms were related to the spinal lesion; the hydrocephalus and the posterior fossa cyst were unexpected findings. In cases 2 and 3, the patients had a long history of cerebral pathology and they later presented with symptoms that implicated the spinal cord.

Case Reports

Case 1

A 6-year-old girl was first seen at the age of 2 months because of weakness of the upper extremities. This was diagnosed as Erb's palsy, and was attributed to a laborious delivery. At the age of 2 years she was found to have bilateral congenital hip dislocation. This was treated surgically with excellent results. Treatment was then directed toward correcting a preexisting scoliosis. A myelogram showed expansion of the spinal cord above T9. Postmyelography CT revealed, unexpectedly, a large midline cyst in the posterior fossa. The cyst herniated through the foramen magnum compressing the spinal cord at C1 and C2. A moderate degree of obstructive hydrocephalus was present. There was no opacification of the cyst or a syrinx on the delayed CT images (Figs. 1A and 1B). However, because of the strong possibility of hydromyelia, MR was performed, which demonstrated the cyst and revealed a syrinx that extended almost the entire length of the cord (Fig. 1C).

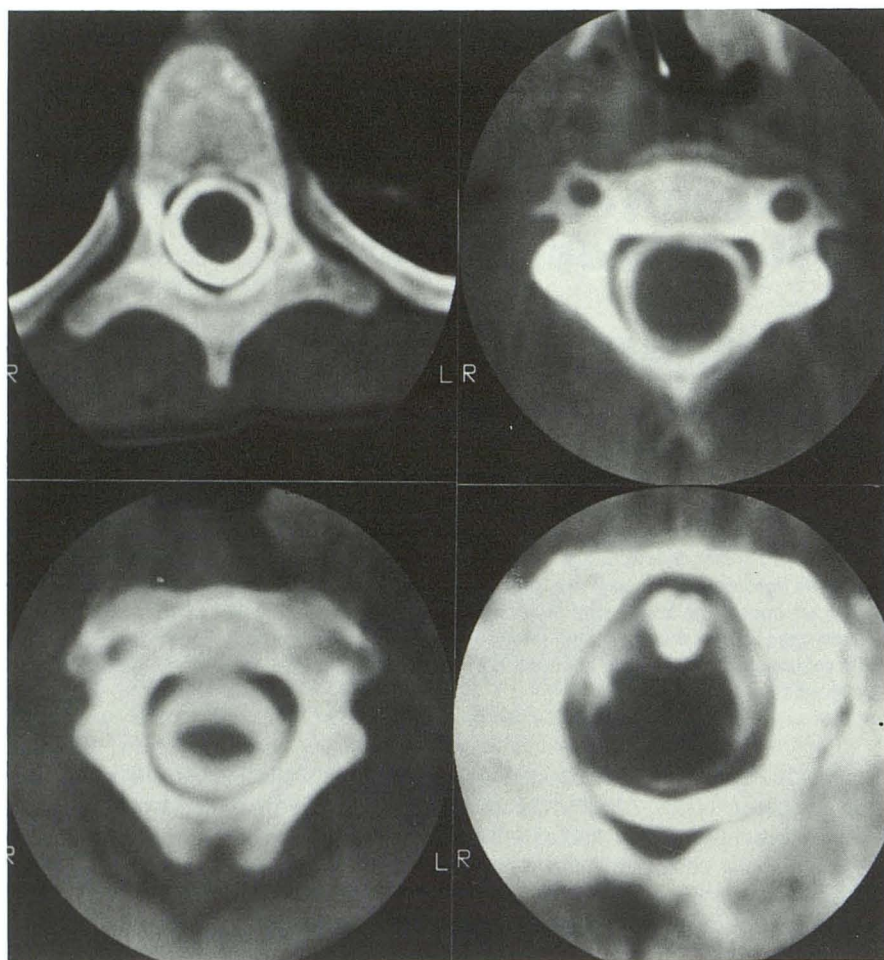
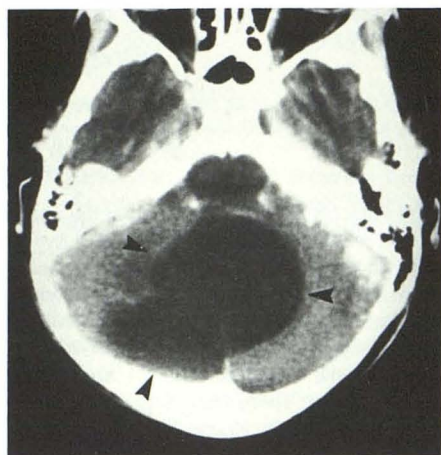
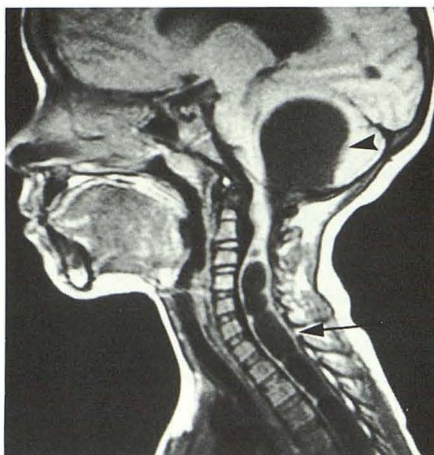
At surgery the posterior rim of the foramen magnum and the posterior arch of the atlas and the laminae of the axis were excised. A cyst displacing the cord anteriorly was found and there were severe arachnoidal adhesions at its lower part. When the cyst was opened, clear fluid came out under pressure. Thin nerve filaments were also seen in the wall of the cyst. The posterior wall of the cyst was removed and when the anterior wall was dissected from the spinal cord, a gush of CSF was expelled from the central canal and the cord collapsed. During the follow-up period there was some improvement in the motor power in both arms, and a repeat MR 1 year later showed a collapsed cord (Fig. 1D). Spinal myelotomy to drain the syrinx was not required.

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¹Department of Diagnostic Radiology, King Faisal Specialist Hospital and Research Centre, P.O. Box 3354, Riyadh, 11211, Saudi Arabia. Address reprint requests to M. Banna.

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**A****B****C****D****Fig. 1.—Case 1.**

A, CT myelogram shows expansion of spinal cord in thoracic (top left) and cervical (top right) regions below C2. The cord was atrophied at C2 (bottom left) and anteriorly displaced by an intradural extramedullary cyst at C1 (bottom right).

B, Postmyelography CT shows unopacified large posterior fossa cyst (arrowheads).

C, Sagittal T1-weighted MR image shows cyst (arrowhead) and syrinx (arrow). Note compression and forward displacement of fourth ventricle as well as herniation of cyst into foramen magnum.

D, One year after surgery, MR image shows collapse of syrinx.

Case 2

A 14-year-old boy suffering from cerebral palsy was examined because of progressive left-sided weakness, affecting the arm more than the leg. His medical history showed a very difficult labor related to cephalopelvic disproportion. His birth had been traumatic, he had a skull fracture and several rib fractures, and had to be ventilated for 15 min after delivery. In his early psychomotor development he was

significantly delayed and there was a poorly documented finding that his head had grown more rapidly than normal in the first 7–9 months of life. However, he suddenly started to improve, and by 3 or 4 years of age was able to manage well at a level with his peers, except for his motor clumsiness.

On examination, there was mild left facial weakness that did not involve the forehead. There was slight weakness of the palate, with the uvula deviating toward the right on elevation and his tongue

protruding slightly to the left. Deep tendon reflexes were hyperactive, and both plantar responses were extensor. Dysmetria and past pointing were more prominent on the left side.

Cranial CT showed obstructive hydrocephalus associated with a large posterior fossa cyst compressing the fourth ventricle (Fig. 2A). To ascertain whether the cyst communicated with the ventricular system, a positive-contrast CT-ventriculogram was obtained. At the time of ventriculography, it was noted that the intracranial pressure was normal. The ventriculogram demonstrated communication between the third ventricle and the quadrigeminal cistern, and between the third ventricle, the displaced fourth ventricle, and a dilated central canal in the cervical and thoracic spinal cord (Fig. 2B). There was no communication with the cyst occupying the left cerebellar hemisphere (Fig. 2C).

A posterior fossa craniotomy revealed a huge left cerebellar cyst. It appeared to consist of filmy arachnoid, but its anterior extension seemed to blend with a gliotic cerebellar hemisphere. The outlets of the fourth ventricle were scarred and there were dense adhesions around the foramen of Magendie. The cyst wall for the most part was removed and the fourth ventricle opened to allow free communication of CSF with remnants of the cisterna magna. Dorsal myelotomy of the cervical region was also performed.

The patient's convalescence was unremarkable, and the operative procedures appeared to stabilize his clinical course. There was some suggestion of improvement of the left arm weakness. There has been no further progression of his condition for the last 6 years.

Histologic examination of the cyst wall showed that it consisted of thickened arachnoid with no evidence of inflammation. The thickened arachnoid was attached to pieces of atrophic cerebellar tissue in which the molecular and granular cell layers were thin and no Purkinje cells were present. No ependymal or glial tissue was seen in the wall of the cyst (Fig. 2D).

Case 3

A 2-year-old boy was investigated at another hospital for increasing head circumference. A CT scan showed dilatation of the lateral ventricles and a midline posterior fossa cyst. A diagnosis of Dandy-Walker syndrome was made and a ventriculoperitoneal shunt was inserted. One year later, the child was evaluated for the treatment of progressive scoliosis. A repeat CT scan showed marked reduction in the size of the lateral ventricles since his initial presentation, but there was no change in the size of the posterior fossa cyst. Accordingly, a positive-contrast CT-ventriculogram was obtained, which revealed no communication between the cyst and the ventricular system (Fig. 3A). A myelogram done a few days later revealed extensive hydromyelia of the spinal cord and unexpected opacification of the cyst (Figs. 3B and 3C). Accordingly, a second shunt was inserted connecting the posterior fossa cyst to the peritoneal cavity. This resulted in collapse of the posterior fossa cyst, as well as of the hydromyelic cavity (Figs. 3D and 3E). The cranial CT scan done after the collapse

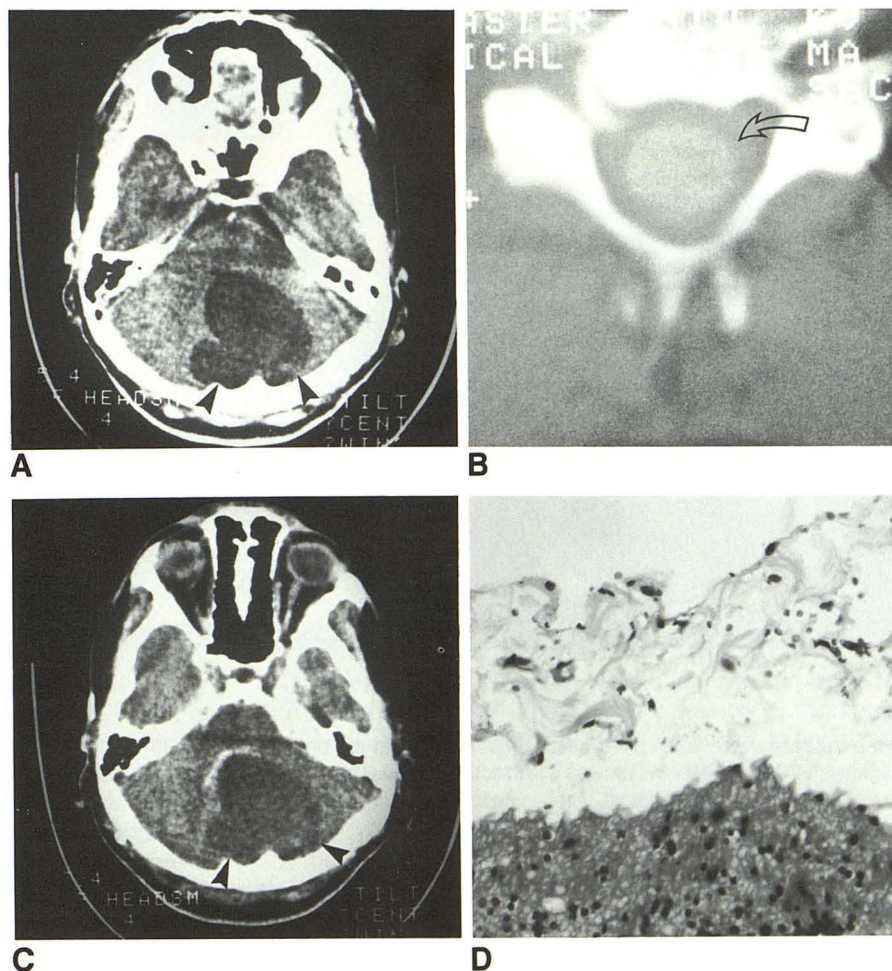


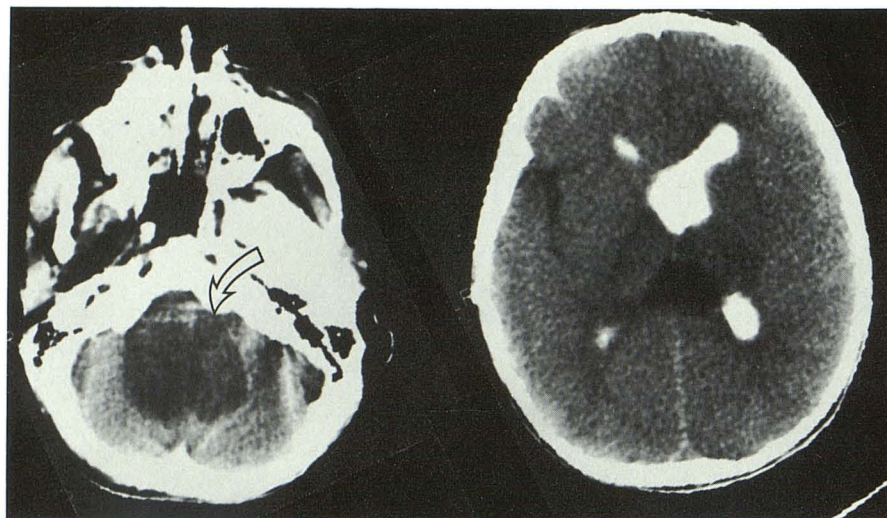
Fig. 2.—Case 2.

A, CT after IV contrast injection shows unenhanced well-defined cyst (arrowheads).

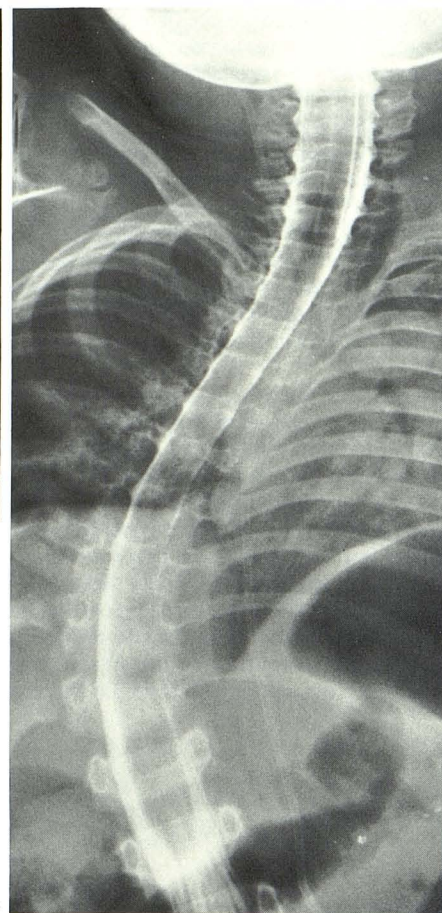
B, CT through upper cervical spine shows large opacified syrinx (arrow) that connected with fourth ventricle.

C, CT-ventriculogram shows no communication between cyst (arrowheads) and opacified, anteriorly displaced fourth ventricle.

D, Photomicrograph of cyst wall shows thickened arachnoid wall (upper half of illustration) and atrophied cerebellar tissue (lower half of illustration).



A



B

Fig. 3.—Case 3.

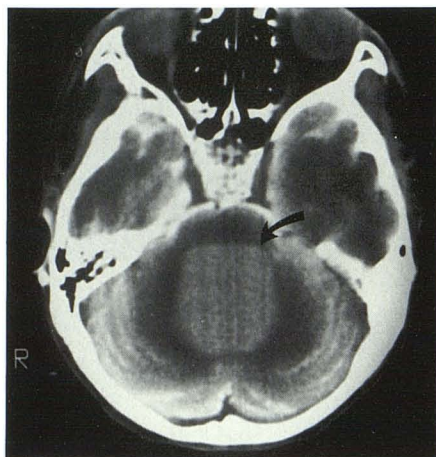
A, CT-ventriculogram shows opacification of lateral ventricles with no opacification of posterior fossa cyst (arrow).

B, Lumbar route myelogram shows generalized enlargement of spinal cord associated with scoliosis.

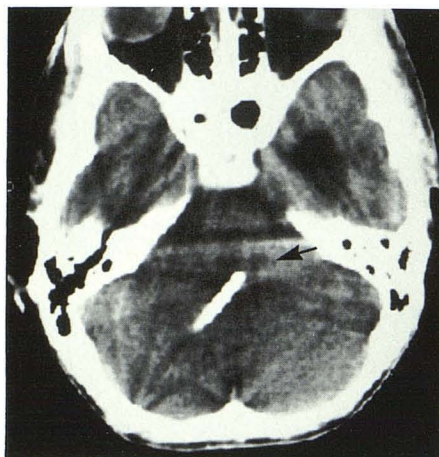
C, Postmyelography CT shows opacification of posterior fossa cyst (arrow).

D, CT after insertion of a cystoperitoneal shunt shows collapse of the cyst. Fourth ventricle is visualized with difficulty (arrow) because of motion artifacts. The vermis is normally developed and there is no evidence of Dandy-Walker syndrome.

E, (on opposite page.) Spinal CT after cystoperitoneal shunt confirms collapse of hydromyelic cavity that is still recognizable within the density of small, atrophied cord (arrowheads).



C



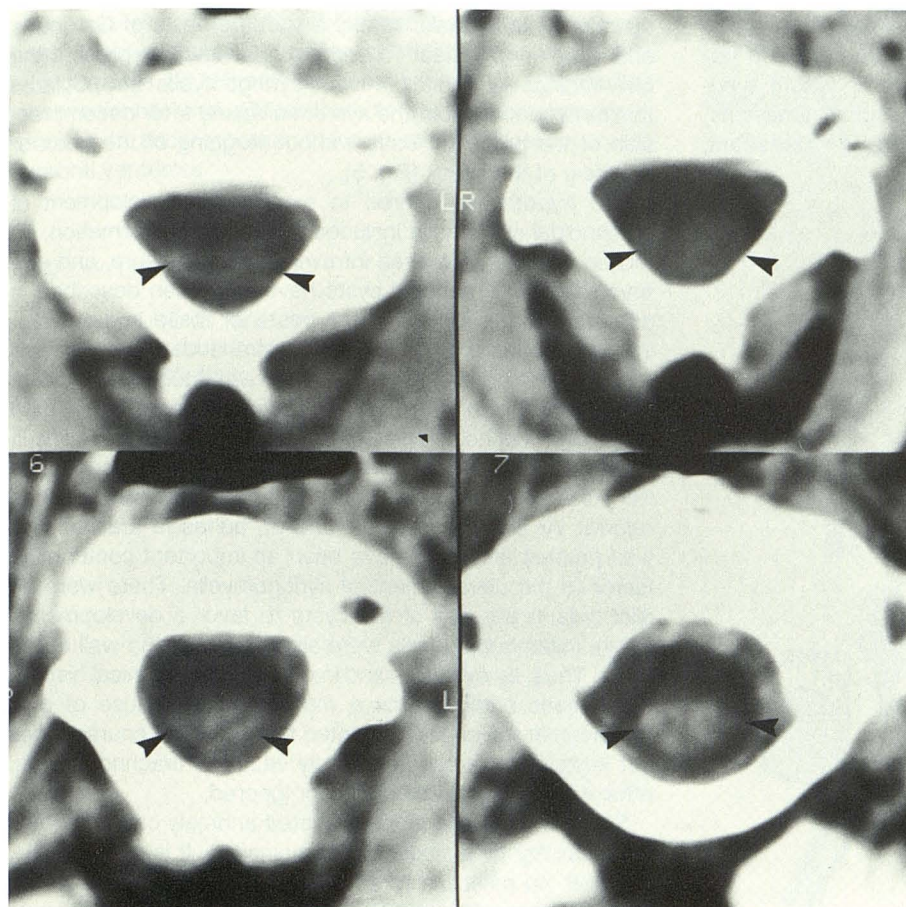
D

of the cyst showed a small fourth ventricle, implying that the original diagnosis of Dandy-Walker cyst was incorrect. What was thought to be a distended fourth ventricle was, in fact, a large midline cyst compressing and obscuring the fourth ventricle.

Discussion

Posterior fossa arachnoid cysts are uncommon and often present in the pediatric age group. They have been classified

according to their topographic location into supracerebellar, infracerebellar, retrocerebellar, laterocerebellar (pontocerebellar), and clival cysts [3]. Arachnoid cysts may communicate with either the subarachnoid space or the ventricular system, or they may be isolated. In this paper we describe infracerebellar midline cysts. Patients with this type of cyst may present with hydrocephalus, manifestations of raised intracranial pressure, truncal ataxia, pyramidal signs, or compression of the lower cranial nerves. The severity of symptoms may



E

be episodic and may be influenced by posture [4]. As evident from the cases described here, some patients may have a syringomyelic syndrome that dominates the clinical picture, some may have cerebellar symptoms that obscure the cyst, or, in some patients, the cyst may only be discovered at the investigation of "idiopathic" scoliosis.

The mechanism of spinal cord cavitation in association with posterior fossa cysts is analogous to the development of syringomyelia with Chiari I malformation or adhesions at the foramen of Magendie. Gardner et al. [5] believed that arachnoid cysts in the region of the foramina of Luschka and Magendie, Arnold-Chiari malformation, Dandy-Walker syndrome, and thick arachnoidal bands around the cerebellar tonsils and medulla can all be associated with syringomyelia. These investigators believed that these lesions are merely different expressions of the same process; namely, failure of the outlets of the fourth ventricle to develop normally in the rhombencephalic roof. These four hindbrain expressions of atresia occur in adults in varying degrees and combinations, and are frequently accompanied by congenital scoliosis, basilar impression, and syringomyelia [5]. Other investigators [4] expressed the view that in their experience there has been little evidence to suggest an association between arachnoid cysts of the posterior fossa and the Arnold-Chiari malformation, Dandy-Walker syndrome, or thickened arachnoidal

bands around the cerebellar tonsils and medulla. None of their patients, these authors continued, had an associated congenital scoliosis or basilar invagination, but one had syringomyelia. Regardless of the interrelation between the aforementioned anomalies, we wish to draw attention to two important practical facts. First, any of these anomalies could be associated with syringomyelia, some more frequently than others (Fig. 4). Second, syringohydromyelia is a well-documented, but frequently overlooked, rare cause of scoliosis in the pediatric age group [6].

The pathogenesis of syringomyelia in association with Chiari I malformation and other related anomalies has been discussed by several authors [1]. Most agree that syringomyelia in association with these anomalies is the result of intermittent obstruction of CSF at the foramen of Magendie. The CSF is driven into the central canal possibly by the arterial pulsations (Gardner's theory) or in response to increased intracranial venous pressure that is brought about by straining, coughing, sneezing, and Valsalva maneuvers (Williams' theory) [1]. A third theory, advanced by Ball and Dayan, advocated that raised intraspinal pressure forces the CSF from the spinal subarachnoid space through the perivascular spaces into the central canal, where it may flow cranially, caudally, or paracentrally through minute pouches or diverticulae [1]. We believe that the last hypothesis offers the most

reasonable explanation for contrast opacification of spinal cord cavities on CT-cisternography in patients with no obstructive lesion at the foramen of Magendie, in whom syringomyelia might be due to trauma, tumors, spinal adhesions, and so forth. If the foramen of Magendie is obstructed,

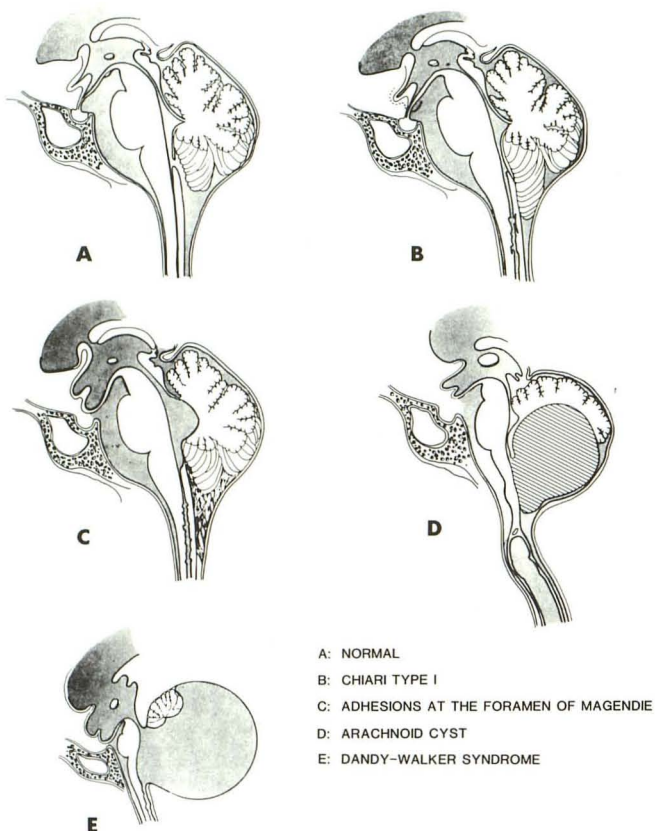


Fig. 4.—Posterior fossa lesions that might be associated with syringomyelia. E is redrawn, with some modification, from Figure 10 in [5].

syringomyelia is easier to explain on the basis of Gardner's and Williams' theories. To support this view, we present the MR findings of a patient with syringomyelia and Chiari I malformation in whom the syrinx collapsed after decompression of the posterior fossa without plugging of the obex or shunting of the syrinx (Fig. 5).

The hypotheses offered to explain the development of arachnoid cysts have included congenital malformation, infection, trauma, increased intraventricular pressure, and embryonic rests. Arachnoid cysts have also been described in association with tumors in the posterior fossa and are presumably produced by some form of transudate [3]. In cases 1 and 2 in the present study, there was history of significant birth injury, which would favor a traumatic origin. In the third case, hydrocephalus was present at birth and it was difficult to rule out birth trauma, even though it could not be confirmed from the information available at the time of the patient's referral. At surgery (cases 1 and 2), adhesive arachnoiditis was present and must have been an important contributing factor to the development of syringomyelia. There were no glial cells in the wall of the cysts to favor a developmental origin. Inflammatory cells were also absent in the wall of the cysts. Thus, by exclusion and in the light of the clinical history, a traumatic origin seems a more probable cause of cyst formation in the cases presented here. This, of course, does not imply that trauma is the only cause of arachnoid cysts; other causes cannot be denied or ignored.

Syringomyelia and any associated anomaly can be shown in its entirety by a single MR examination. It is not possible, however, to establish whether or not a cyst communicates with the ventricular system on MR. On CT, an arachnoid cyst appears as a clearly defined hypodense area that is equal in density to the CSF. The lesion is extraneuronal, it is not accompanied by brain edema, and does not enhance after IV contrast injection. The fourth ventricle is displaced and compressed, and may not be clearly visualized on the routine axial slices except after drainage or excision of the cyst. Positive-contrast CT-cisternography or ventriculography may be required to show the extent of the syrinx and verify any communication with the CSF spaces [5].

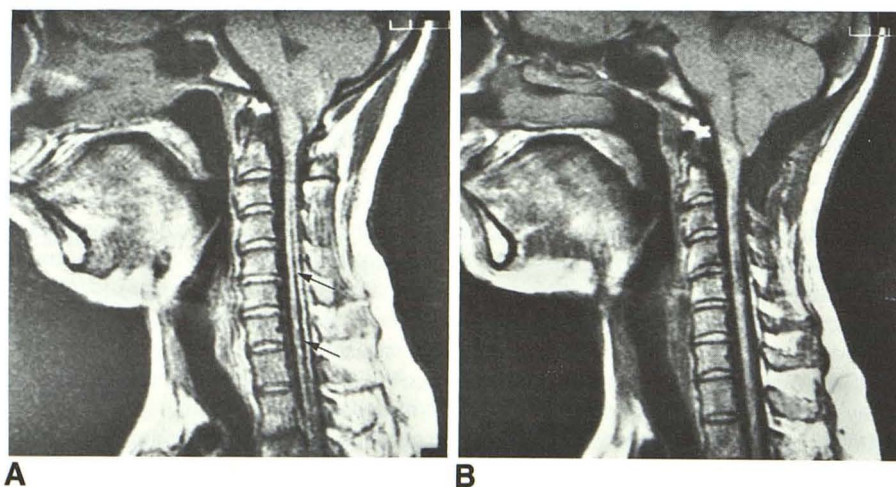


Fig. 5.—Collapse of syrinx after posterior fossa decompression only in a patient with Chiari I malformation.

A, Sagittal T1-weighted MR image before surgery shows herniation of cerebellar tonsils and a narrow syrinx (arrows).

B, MR image 2 months after surgical decompression of posterior fossa illustrates collapse of syrinx. Surgical decompression was accomplished by removing posterior rim of foramen magnum, posterior arch of atlas, and laminae of C2. The dura was incised and the defect so created was closed using lyophilized dura mater to provide a slack covering to the herniated tonsils. (Courtesy of Dr. E. Siqueira, King Faisal Specialist Hospital.)

In conclusion, the pertinent findings are that (1) there is a possible coexistence of midline posterior fossa cysts with syringomyelia, (2) MR provides most information with minimal inconvenience, and (3) positive-contrast cisternography is required to show whether the cyst communicates with the cerebral ventricles.

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