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This information is current as of April 19, 2024.

AJNR Am J Neuroradiol 1987, 8 (6) 973-978
<http://www.ajnr.org/content/8/6/973>

Syringomyelia as a Consequence of Compressive Extramedullary Lesions: Postoperative Clinical and Radiological Manifestations

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MR imaging was performed to determine the cause of the onset of new neurologic symptoms in five patients who had previously undergone surgical excision of extramedullary masses. Syringomyelia and the absence of recurrent or residual lesions were documented in all cases. Three patients showed long cysts (multiseptated in two and smooth in one) with low signal intensity on both T1- and T2-weighted images. The flow-void phenomenon related to fluid motion in these three cysts, which were enlarging clinically, was responsible for the hypointensity on the T2-weighted images. In two patients the fluid within the lesions behaved similarly to normal nonpulsatile CSF and may have represented syrinx cavities in a state of "arrested growth." Three patients had surgical decompression under real-time intraoperative sonographic control, which showed the presence of intramedullary cyst-fluid pulsations in two cases and the absence of cyst-fluid pulsations in one case. These sonographic observations correlated with the MR findings.

We postulate that these syrinx cavities form as a result both of the effect that the original extramedullary lesion had upon the underlying spinal cord and the subsequent postoperative alterations in the CSF dynamics at the level of prior surgery. Syringomyelia should be considered in patients with recurrent or new symptoms who previously had surgery for extramedullary lesions.

Syringomyelia presenting synchronously with extramedullary tumors has been regarded as rare [1-3], and syringomyelia in patients with prior removal of extramedullary lesions has not been described. Five patients in whom extramedullary lesions had been previously removed were examined by MR imaging. The presence of long syrinx cavities and the absence of residual or recurrent lesions were documented in all cases. Because of this experience, we describe the development of syringomyelia after the removal of extramedullary lesions, and postulate a mechanism for their formation and enlargement.

This article appears in the November/December 1987 issue of *AJNR* and the February 1988 issue of *AJR*.

Received March 17, 1987; accepted after revision June 3, 1987.

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AJNR 8:973-978, November/December 1987
 0195-6108/87/0806-0973

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Subjects and Methods

Five patients ranging in age from 33-61 years (mean, 42 years) were studied. All patients had extramedullary lesions that had been removed 5-24 years previously (mean, 16 years), and all patients were examined on a 0.5- or 1.5-T superconducting MR imaging system. Thin sections (5 mm) in sagittal and axial projections were obtained through the regions of interest. T1-weighted images, TR 500-1000/TE 26-38, and T2-weighted images, TR 1500-2000/TE 80-120, were obtained. Three patients had metrizamide myelography (230 mg/ml, 10 ml) by lumbar puncture followed by 3-hr delayed CT (GE 9800) using 10-mm slice thickness. One patient had myelocystography (160 mg/ml, 3 ml). Three patients had surgery, which was monitored with real-time intraoperative sonography using a 7.5-MHz transducer.

Case Reports

Case 1

A 60-year-old woman presented with a 2-year history of bilateral lower extremity weakness and difficulty voiding. Twelve years earlier she had similar symptoms and a T7 meningioma was discovered and removed. Postoperatively, her weakness improved and her voiding difficulty disappeared. On her current examination, increased muscle tone and absent vibratory sense in both lower extremities and urinary incontinence were present. A T1-weighted MR image showed a low signal intensity within a dilated cord (Fig. 1A). This cyst extended from T7 to the conus medullaris and was noted to have irregular borders and internal septations. A T2-weighted image showed the lesion to have similar signal intensity to the adjacent CSF (Fig. 1B). No areas suspicious for tumor recurrence were seen. Metrizamide myelocystography (Fig. 1C) performed at the T12-L1 level clearly showed the multiseptated nature of the lesion (Fig. 1C). Intraoperative spinal sonography confirmed the presence of a large, septated, nonpulsatile cyst (Fig. 1D). The syrinx cavity was shunted into the subarachnoid space, and fluid obtained from the syrinx had an increased protein concentration (65 mg/dl). After surgery, the pa-

tient's lower extremity weakness improved; however, bladder control was not regained.

Case 2

Twenty years before her current evaluation, a 61-year-old woman experienced bilateral lower extremity weakness. A T4 meningioma was found and removed, and she recovered completely. Seventeen years later she presented with a new onset of left lower extremity weakness and stiffness of the left foot. Reexploration revealed recurrent meningioma, which was completely removed. The patient improved; however, 1 year later she noted lower extremity weakness, which progressed over the next 2 years. Physical examination showed markedly decreased muscle strength in her left lower extremity and decreased position and vibratory sensation in both lower extremities. Metrizamide myelography showed a block at T11 and delayed metrizamide CT revealed a cord cyst extending from T2 to T11. T1- and T2-weighted MR images showed a low-signal lesion centrally located within an enlarged cord (Fig. 2A). The lesion extended from T1 to T11 and demonstrated some internal areas of increased signal intensity suggesting intracystic septations. No re-

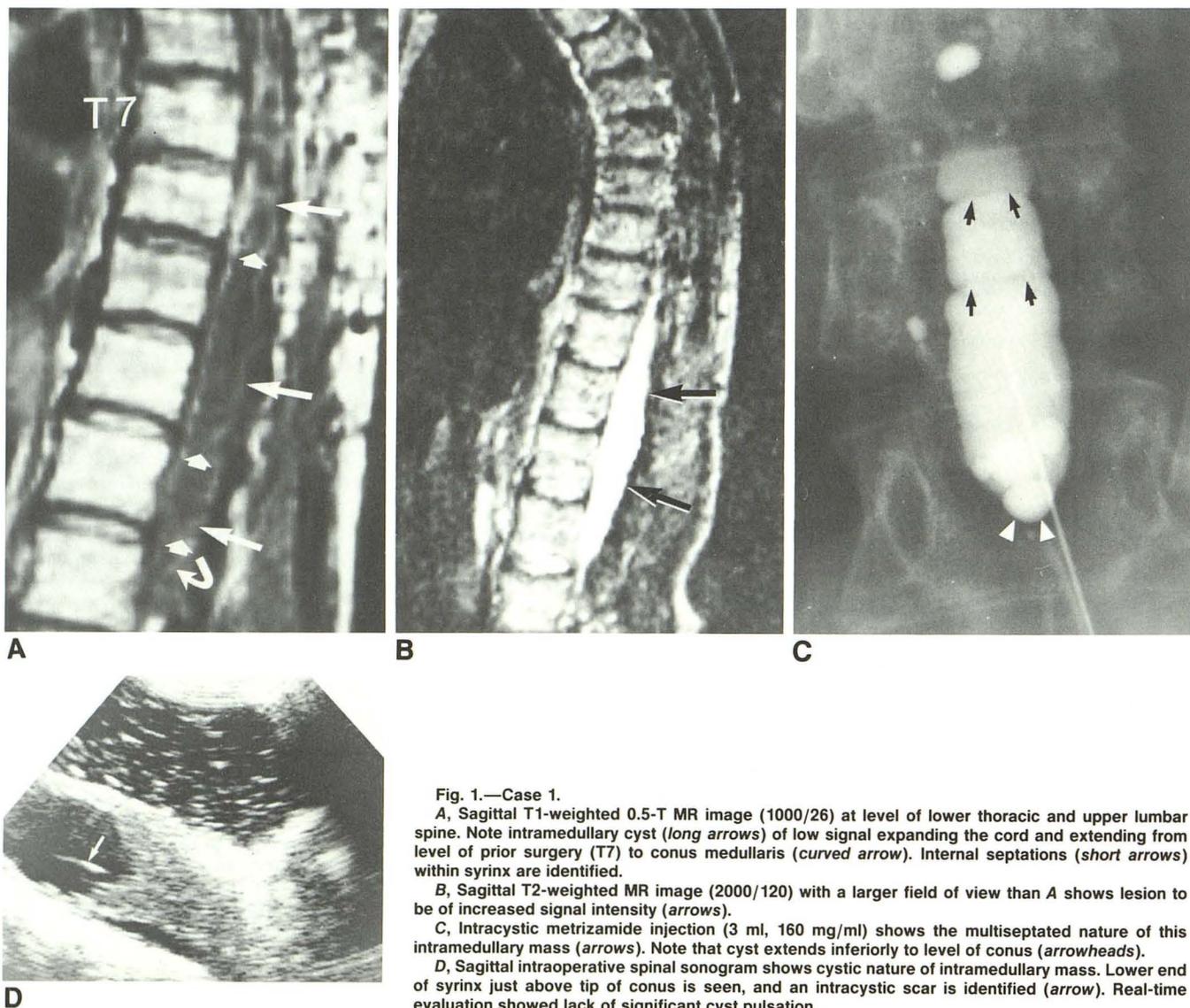


Fig. 1.—Case 1.

A, Sagittal T1-weighted 0.5-T MR image (1000/26) at level of lower thoracic and upper lumbar spine. Note intramedullary cyst (long arrows) of low signal expanding the cord and extending from level of prior surgery (T7) to conus medullaris (curved arrow). Internal septations (short arrows) within syrinx are identified.

B, Sagittal T2-weighted MR image (2000/120) with a larger field of view than A shows lesion to be of increased signal intensity (arrows).

C, Intracystic metrizamide injection (3 ml, 160 mg/ml) shows the multiseptated nature of this intramedullary mass (arrows). Note that cyst extends inferiorly to level of conus (arrowheads).

D, Sagittal intraoperative spinal sonogram shows cystic nature of intramedullary mass. Lower end of syrinx just above tip of conus is seen, and an intracystic scar is identified (arrow). Real-time evaluation showed lack of significant cyst pulsation.

Fig. 2.—Case 2.

A, Upper portion of syrinx cavity (arrows) is seen on this sagittal T2-weighted 0.5-T MR image (1500/80). The lesion is of low signal intensity and was of similar intensity to that found on T1-weighted image.

B, Transverse intraoperative spinal sonogram at T4 level. A linear area of increased echogenicity (arrow) represents intracystic scar. Real-time evaluation showed marked fluid pulsations within cyst.

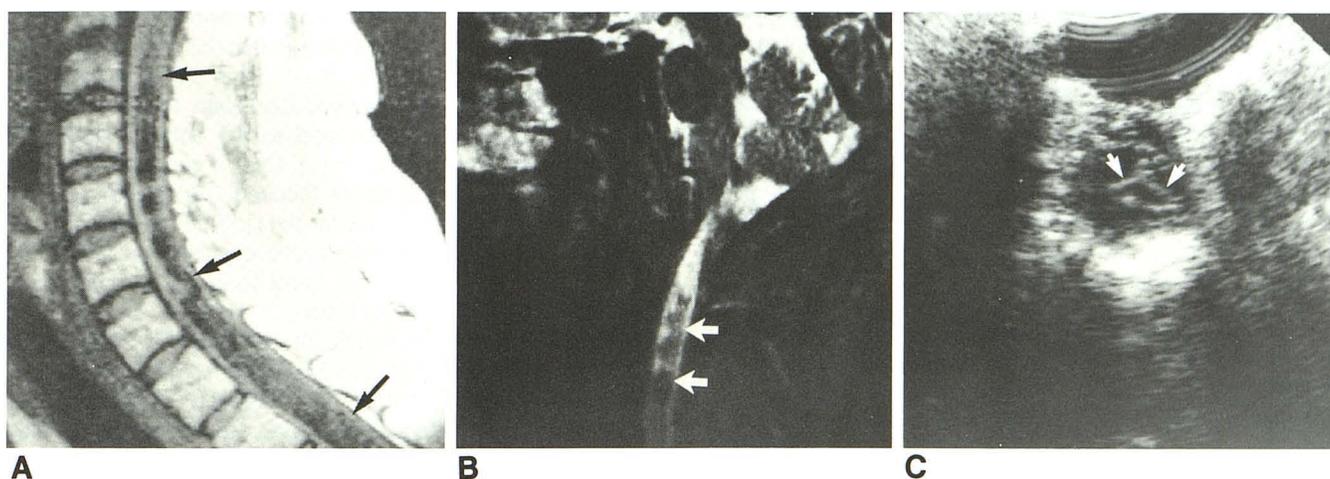
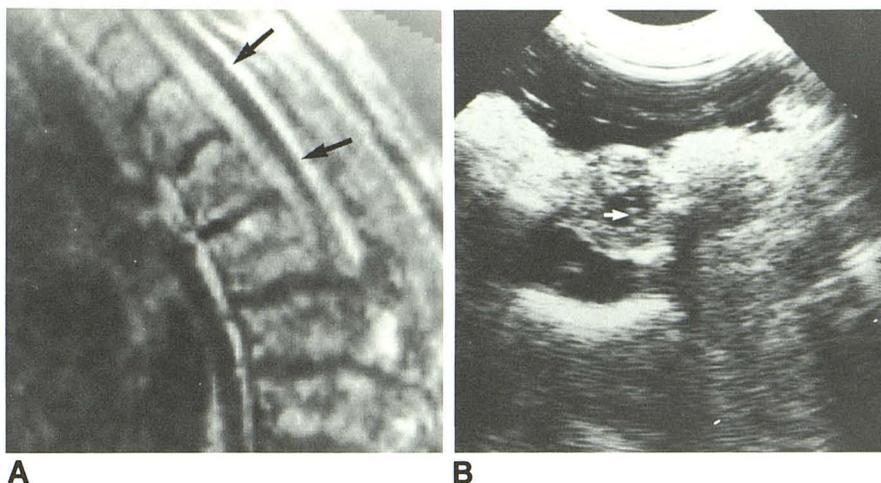


Fig. 3.—Case 3.

A, Sagittal T1-weighted 0.5-T MR image (500/38) shows multiseptated syrinx cavity of low signal intensity (arrows) extending from upper cervical region to T4.

B, Sagittal T2-weighted MR image (2000/60) of upper cervical spine and head shows low signal intensity of cyst cavity (arrows).

C, Transverse intraoperative spinal sonogram at level of C7 confirmed presence of syrinx cavity containing multiple scars (arrows). Real-time evaluation revealed cyst pulsations.

current tumor was seen. Intraoperative spinal sonography (Fig. 2B) confirmed the presence of a multiseptated pulsatile cyst cavity. A syrinx to subarachnoid space shunt was performed. The fluid obtained from the syrinx cavity had an elevated protein (161 mg/dl). Moderate improvement of strength and sensation was noted postoperatively.

Case 3

A 42-year-old man presented with a history of right upper extremity pain, right lower extremity pain and weakness, sphincter dysfunction, and impotence that had progressed over the last 4 years. Twenty-four years earlier he had progressive right-sided weakness and right lower extremity numbness and spasticity. A T4 meningioma was resected. Postoperatively, his symptoms improved. The present physical examination showed increased muscle tone in the right lower extremity, a right Babinski, and areflexia of both upper extremities.

Decreased anal sphincter tone and urinary incontinence were also noted. Metrizamide myelography showed diffuse widening of the

cervical spinal cord. On MR, the T1-weighted image showed a low-signal lesion extending from C2 to T4 with irregular borders and multiple areas of increased signal intensity within the lesion (Fig. 3A). The T2-weighted image showed the lesion to be of decreased signal intensity as compared with the adjacent CSF, and multiple septations were seen (Fig. 3B). No residual or recurrent tumor was noted. Intraoperative spinal sonography confirmed the presence of a pulsatile cystic lesion containing multiple scars (Fig. 3C). A syrinx to subarachnoid space shunt was placed. Analysis of the syrinx fluid revealed increased protein content (40 mg/dl). Postoperatively, the patient experienced relief of his symptoms in both upper extremities and regained some sphincter control.

Case 4

A 41-year-old man presented with a 2-year history of paresthesias and progressive weakness of both lower extremities, and wasting of the muscles of the right lower extremity. Twenty years previously he had presented with right-sided intercostal pain and weakness and

numbness of the right foot. A neurilemmoma at the T7–T8 level was resected. Postoperatively, the intercostal pain receded but the symptoms in the right foot persisted. The present physical examination revealed weakness of the entire right lower extremity. On MR, a long, narrow syrinx that was hypointense on T1- and T2-weighted images, and that extended from T1–T6, was identified. This cystic cavity had smooth borders and was centrally located within a nonexpanded spinal cord (Fig. 4). No tumor recurrence was noted. Because the cord was not expanded on MR, no surgery was performed and the patient is being followed clinically.

Case 5

A 33-year-old man was evaluated 5 years earlier for progressive pain and weakness in both upper extremities and the right lower extremity. Physical examination at that time revealed muscle atrophy of the right upper extremities and bilateral Babinski signs. Metrizamide myelography and delayed metrizamide CT showed compression of the spinal cord at C4–C5 by a calcified disk. There was no evidence of metrizamide accumulation within the cord. A discectomy, anterior decompression, and anterior cervical fusion from C3 to C5 were performed. Postoperatively, the patient experienced remission of his symptoms with the exception of mild pain in the right upper and lower extremities. At present evaluation, the patient complained of interscapular pain radiating to the right upper and lower extremities of 6 months' duration. Sagittal and axial MR was performed and showed a low-signal lesion on the T1-weighted image extending from the level of C1 to C6 (Fig. 5A). The lesion had smooth borders and was centrally located within the spinal cord. The T2-weighted image showed the lesion to be of increased signal intensity, similar to the surrounding CSF (Fig. 5B). Because of the MR findings and the clinically stable symptoms, the patient is being managed conservatively at this time.

Discussion

The most important clinical consideration in patients who have previously been operated on for extramedullary spinal lesions and who present with reappearance or worsening of symptoms is a recurrence of the original lesion. The possibility of secondary syringomyelia must, however, be considered in the differential diagnosis. Syringomyelia coexisting with extramedullary tumors has been reported in only 13 cases [1–3],

and to our knowledge, no cases have been described in which syringomyelia was found in patients who had prior removal of extramedullary lesions. We have recently encountered five such patients.

Several mechanisms can be postulated to explain the formation of syringomyelia under conditions that bear some similarities to our cases. Ischemia of the cord with secondary degeneration of the gray matter [4, 5] or filling of an obstructed central canal by CSF that is extrachoroidally produced [3] are two proposed mechanisms. A more recent hypothesis states that long-standing compression of the cord due to the presence of an extramedullary mass may lead to neuroglial damage, which is manifested by enlargement of the extracellular perivascular spaces. The removal of such an extramedullary lesion will not reverse the damage the spinal cord has already sustained nor will the microcystic areas within the cord disappear as a result of the surgery. The formation of multiple subarachnoid adhesions at the level of previous surgery may funnel CSF pulsations toward the area of the damaged cord where microcystic changes have occurred. This situation, in association with focally enlarged Virchow Robin spaces, can lead to the entry of abnormal amounts of CSF into the cord [1]. Over a period of time, the water-hammer effect of these focused CSF pulsations can cause these microcysts to enlarge [1]. This mechanism can explain the formation of long cystic cavities that are either caudal (case 1), rostral (cases 3 and 4), or caudal and rostral (cases 2 and 5) to the initial lesion.

The long symptom-free period experienced by our patients makes it unlikely that large and/or expanding syrinx cavities were present at the time of initial diagnosis. The exact time required for this type of syringomyelia to form and manifest itself is difficult to estimate, but given the proposed theory of formation and propagation, months to years are probably required, similar to that seen in the traumatically injured spinal cord [6]. The pathologic mechanisms that might lead to the formation of this type of syringomyelia are illustrated in Figure 6.

In our cases, MR was able to demonstrate the intramedullary cysts and to suggest their dynamic state. In three of our patients (cases 2, 3, and 4) the T2-weighted images demon-

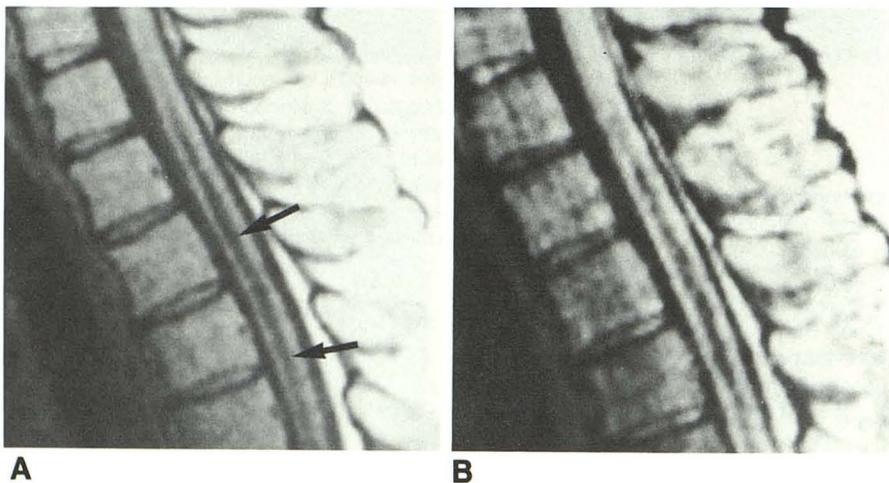
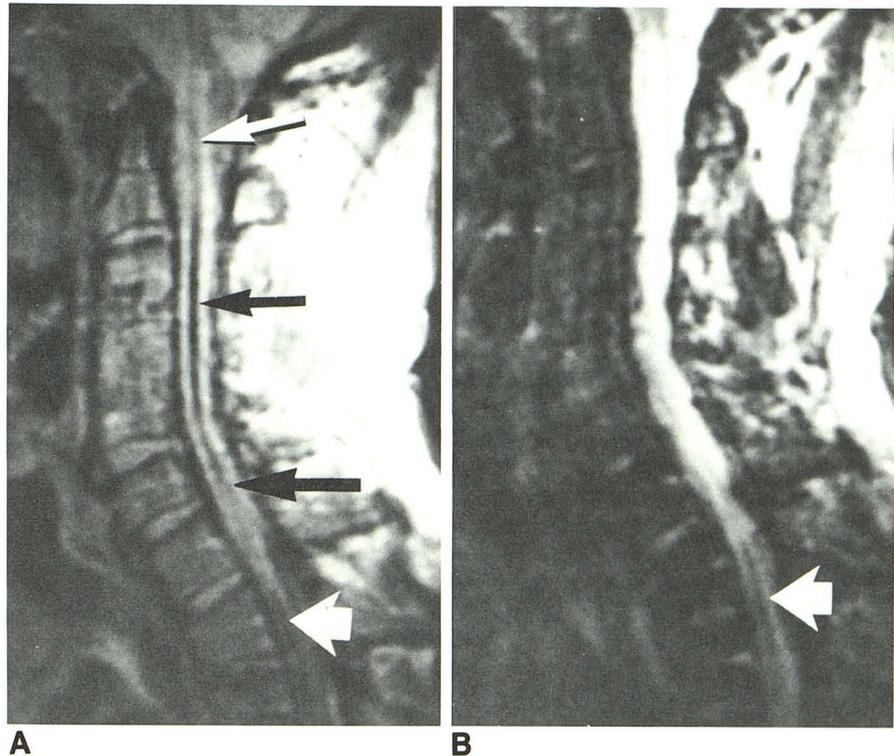


Fig. 4.—Case 4.
A, Sagittal T1-weighted 1.5-T MR image (1000/26) shows low-signal central syrinx cavity extending from level of T1 to T6 (arrows).
B, Sagittal T2-weighted MR image (2000/80) shows continued low signal intensity within syrinx cavity.

Fig. 5.—Case 5.

A, Sagittal T1-weighted 0.5-T MR image (1000/26) of cervical spine shows low signal syrinx cavity extending from C1 to C6 (*thin arrows*) and probably another short intramedullary cyst at the T1–T2 level (*thick arrows* in A and B). The cord is not dilated.

B, Sagittal T2-weighted MR image (2000/120) shows upper cystic lesion to be of increased signal intensity. Since neither cardiac gating nor extra gradients were used to suppress motion artifacts, we cannot be sure that the hypointensity at T1–T2 does not represent flow artifacts.



strated long, hypointense syrinx cavities, the appearance of which we believe were secondary to intracystic fluid motion and turbulence. A similar phenomenon has been demonstrated at narrow CSF pathways (foramina of Monro, aqueduct of Sylvius, and foramen of Magendie), where relatively fast pulsatile CSF motion occurs [7, 8]. This flow-void phenomenon is most likely caused by spin phase shifts and time-of-flight effects [9–11]. Although ours is the first study to report a correlation between the CSF motion as suggested by MR and subsequently shown with intraoperative spinal sonography, a recent presentation indicates that pulsatile fluid in cyst cavities can be identified and differentiated from myelomalacia by virtue of its signal augmentation when imaged by gradient echoes [12]. In the three patients who had surgery, fluid analysis from the syrinx cavities revealed increased protein content. Although such chemical changes can play a role in the MR signal characteristics [13], there was no correlation between the protein content and the MR signal in the three patients whose cyst fluid was analyzed. The explanation for this is that the MR signal of such fluid depends far more on its dynamic state (i.e., fluid pulsation and turbulence) than on the composition of the fluid.

The presence of pulsatile and nonpulsatile cystic cavities coexisting has been observed [14]. It may be that where there is an absence of the flow-void phenomenon, restriction of fluid motion by multiple intracystic septations is present. In three of our cases the multiseptated nature of the syrinx cavities was documented by intraoperative spinal sonography (Figs. 1D, 2B, 3C). These intracystic septations represent fibroglial remnants [12] of the spinal cord that probably are segments of neural tissue that were resistant to complete destruction caused by the enlarging syrinx. As these sepa-

rated compartments develop communications with each other, greater fluid motion may then be allowed, resulting in the flow-void phenomenon within the cyst cavities and promoting their enlargement. Since two of our patients (cases 2 and 3) with multiseptated lesions manifested intrasyrinx flow-void phenomenon, the size of the openings between these cavities may be an important factor in determining the presence or absence of this occurrence. We believe that both multiloculated and uniloculated lesions have a potential for growth that depends on the dynamic state of the fluid in the cyst and the size of the connections between loculations, when they exist. The preoperative identification of these fibroglial remnants is important, because it may indicate to the surgeon the need to lyse the septa in order to create a single cavity suitable for shunt catheter placement. With MR, it is possible that because of the thickness of each slice, septa that appear to be complete may actually be incomplete at surgery.

Given this theory of cord compression and subsequent postoperative syrinx formation, one might think that this situation would be seen more often in patients operated on for chronic compressive extradural lesions, such as a herniated disk. In our limited series, however, we report only one such patient (case 5), leading us to speculate that in extradural lesions, the presence of an intact dura may offer some protection to the spinal cord, which is not present in cases of intradural masses (cases 1–4).

As MR becomes more widespread and is used for the evaluation of patients previously operated on for extramedullary masses, the presence of secondary syringomyelia will become increasingly recognized. In these situations, we suggest that syrinx cavities that are hypointense on T2-weighted

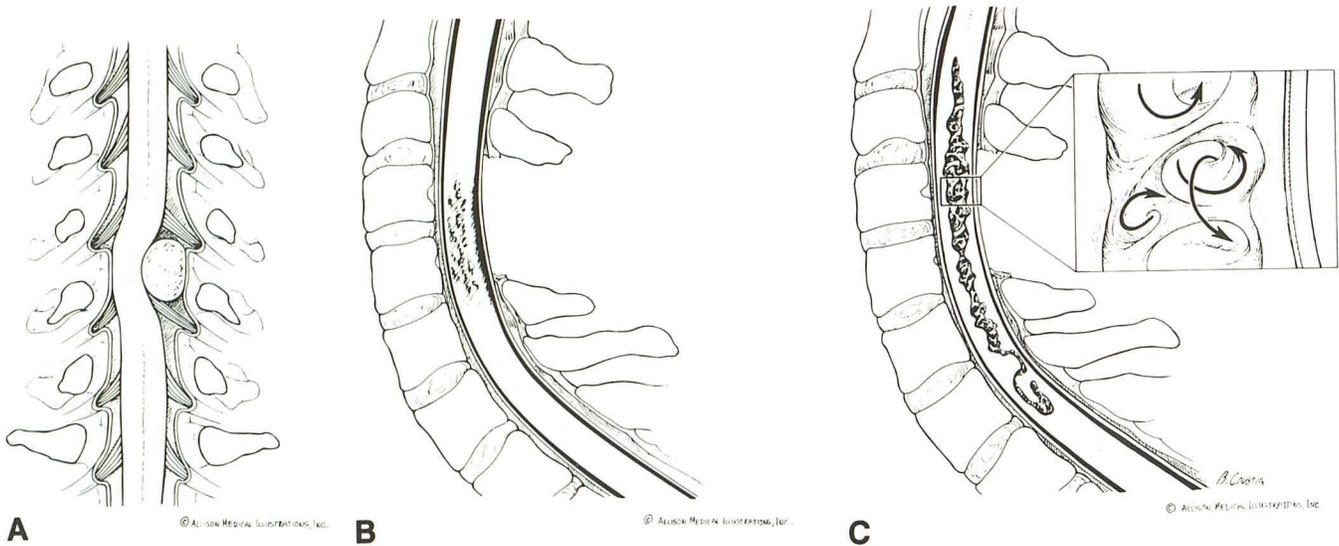


Fig. 6.—Diagrammatic representation of the events that are postulated to lead to the postoperative formation of syringomyelia after the removal of extramedullary lesions.

A, Drawing showing a coronal section through cervical spine. Extramedullary mass to left of cord is shown with intramedullary edema and microcystic changes (shaded area) due to long-standing compression of cord.

B, Sagittal view of postoperative cervical spine seen after total extramedullary tumor removal. The cord remains deformed and small cysts are present secondary to coalescence of microcystic changes.

C, Diagram of late postoperative period. The multiple small cysts have joined and formed large, irregular cavities with multiple septations. Multiple adhesions within subarachnoid space, which form at level of prior surgery and may be responsible for directing pulsatile forces to the cord, are not shown in this illustration. The transmitted pulsations can create intracystic fluid turbulence (arrows in inset), causing expansion of cyst both rostrally and caudally. During periods of active syrinx expansion, this turbulence may manifest as a flow void on T2-weighted images (see text).

images are more likely to be those that are undergoing active expansion at the time of examination. These are the patients that may benefit most from syrinx decompression (e.g., cases 2 and 3). On the other hand, MR characteristics suggesting the presence of nonpulsatile fluid may represent syrinx cavities that are not undergoing active expansion. In those cases the clinical symptoms and the presence (case 1) or absence of an enlarged cord will determine the therapeutic approach. We do not have an explanation of why, at times, an intramedullary cyst may undergo active expansion both clinically and radiologically and at other times the same cyst may be relatively inactive. In our series, all three patients operated on for syrinx decompression improved clinically. With a nondilated cord, the presence (case 4) or absence (case 5) of an intrasyrinx flow-void phenomenon can give the clinician an indication of possible active syrinx enlargement even when the immediate decision is not to perform surgery. Those patients with flow void might be watched more carefully for signs of clinical deterioration.

In conclusion, we believe MR is the imaging method of choice for the evaluation of patients with new onset neurologic symptoms after the removal of a previous extramedullary mass. Not only can the possibility of recurrent lesion be excluded but the presence of syringomyelia can be documented. Evaluation of the signal intensity from the syrinx when combined with the clinical presentation can supply additional information that may be useful in determining the need for syrinx decompression.

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