



**Syringomyelia associated with intradural
extramedullary masses of the spinal canal.**

R M Quencer, T el Gammal and G Cohen

AJNR Am J Neuroradiol 1986, 7 (1) 143-148

<http://www.ajnr.org/content/7/1/143>

This information is current as
of July 31, 2025.

Syringomyelia Associated with Intradural Extramedullary Masses of the Spinal Canal

Robert M. Quencer¹
 Taher El Gammal²
 Gil Cohen¹

Three cases of syringomyelia associated with intradural extramedullary tumors of the spinal canal are reported to demonstrate their radiographic features, to postulate a mechanism for their formation, and to describe the clinical deterioration occurring in two patients immediately after metrizamide myelography. It is believed that the mechanism by which these syrinx cavities formed was similar to the mechanism of syrinx formation within chronically injured spinal cords. The cystic cavities may extend for considerable distances away from the extramedullary mass and may be present above and/or below the mass. The development of a syringomyelia in the face of an intradural extramedullary tumor may not be rare, and it is suspected that with the more frequent use of delayed metrizamide CT and now magnetic resonance imaging, syringomyelia will be found as an occasional consequence of this type of mass.

The development of a spinal cord cyst in association with an extramedullary mass is considered extremely rare. In the literature only eight cases of suggested or documented syringomyelia associated with extramedullary intraspinal lesions have been reported [1, 2]. In none of those patients were the radiographic findings illustrated or described. We recently examined three patients in whom this combination of findings was demonstrated on myelography and delayed metrizamide computed tomography (CT). Our object in this report is threefold: (1) to illustrate and describe the often confusing radiographic images that may occur when a syringomyelia and an intradural extramedullary mass occur together, (2) to propose a pathologic mechanism that could account for the development of the syrinx, and (3) to offer a warning that clinical deterioration after myelography in these types of cases may occur as a result of accumulation of metrizamide within the syrinx.

Case Reports

Case 1

A 42-year-old woman noted the onset of paresthesias involving her left leg 3 years before admission. One year later, because of progressive weakness and numbness of her left leg, she underwent an L4-L5 laminectomy for suspected canal stenosis at another hospital and experienced no relief of her symptoms. She became increasingly weak in both legs, and at the time of admission, she required a walker for ambulation. In addition, she complained of urinary incontinence and intrascapular pain. Physical examination revealed bilateral leg weakness, a T5 sensory level, bilateral ankle clonus, and increased muscle tone in her legs. Metrizamide myelography (figs. 1A and 1B) showed a widened thoracic and cervical spinal cord and a lobular filling defect in the canal at C5-C6. Immediate metrizamide CT (figs. 1C and 1D) confirmed a mass and an enlarged spinal cord. Delayed metrizamide CT (fig. 1E) showed findings consistent with syringomyelia. After myelography the patient became increasingly weak in her legs, and she was treated with corticosteroids. Twenty-four hr later she had returned to her premyelographic neurologic status. Because of the confusing nature of the myelographic and metrizamide CT findings an intravenously enhanced CT scan of the spine was obtained (fig. 1F), which showed a mass within the spinal canal at C5. At surgery,

Received March 6, 1985; accepted after revision June 25, 1985.

¹ Department of Radiology (R-130), University of Miami, Jackson Memorial Medical Center, P.O. Box 016960, Miami, FL 33101. Address reprint requests to R. M. Quencer.

² Department of Radiology, Medical College of Georgia, Augusta, GA 30902.

AJNR 7:143-148, January/February 1986

0195-6108/86/0701-0143

© American Society of Neuroradiology

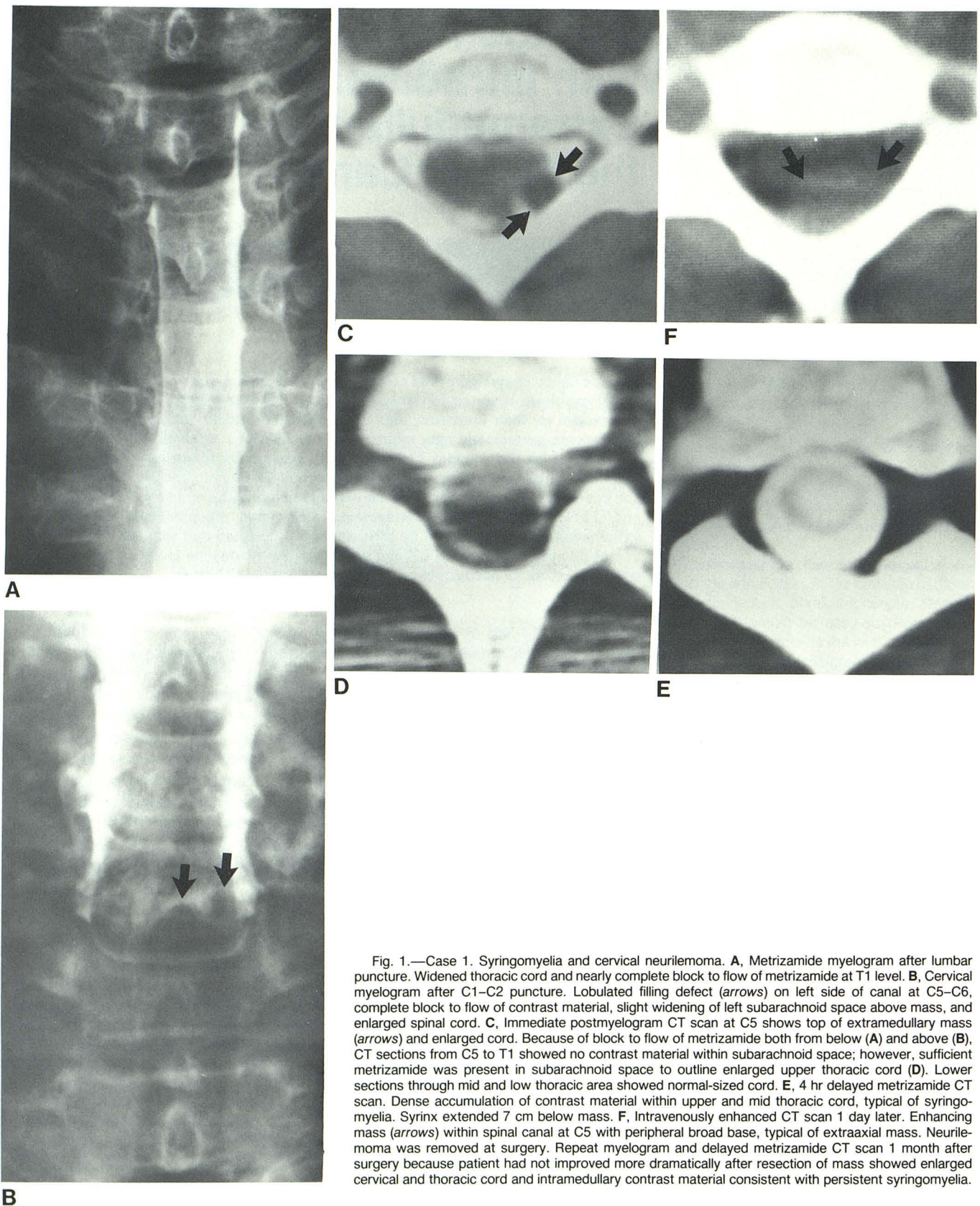


Fig. 1.—Case 1. Syringomyelia and cervical neurilemoma. **A**, Metrizamide myelogram after lumbar puncture. Widened thoracic cord and nearly complete block to flow of metrizamide at T1 level. **B**, Cervical myelogram after C1–C2 puncture. Lobulated filling defect (arrows) on left side of canal at C5–C6, complete block to flow of contrast material, slight widening of left subarachnoid space above mass, and enlarged spinal cord. **C**, Immediate postmyelogram CT scan at C5 shows top of extramedullary mass (arrows) and enlarged cord. Because of block to flow of metrizamide both from below (**A**) and above (**B**), CT sections from C5 to T1 showed no contrast material within subarachnoid space; however, sufficient metrizamide was present in subarachnoid space to outline enlarged upper thoracic cord (**D**). Lower sections through mid and low thoracic area showed normal-sized cord. **E**, 4 hr delayed metrizamide CT scan. Dense accumulation of contrast material within upper and mid thoracic cord, typical of syringomyelia. Syrinx extended 7 cm below mass. **F**, Intravenously enhanced CT scan 1 day later. Enhancing mass (arrows) within spinal canal at C5 with peripheral broad base, typical of extraaxial mass. Neurilemoma was removed at surgery. Repeat myelogram and delayed metrizamide CT scan 1 month after surgery because patient had not improved more dramatically after resection of mass showed enlarged cervical and thoracic cord and intramedullary contrast material consistent with persistent syringomyelia.

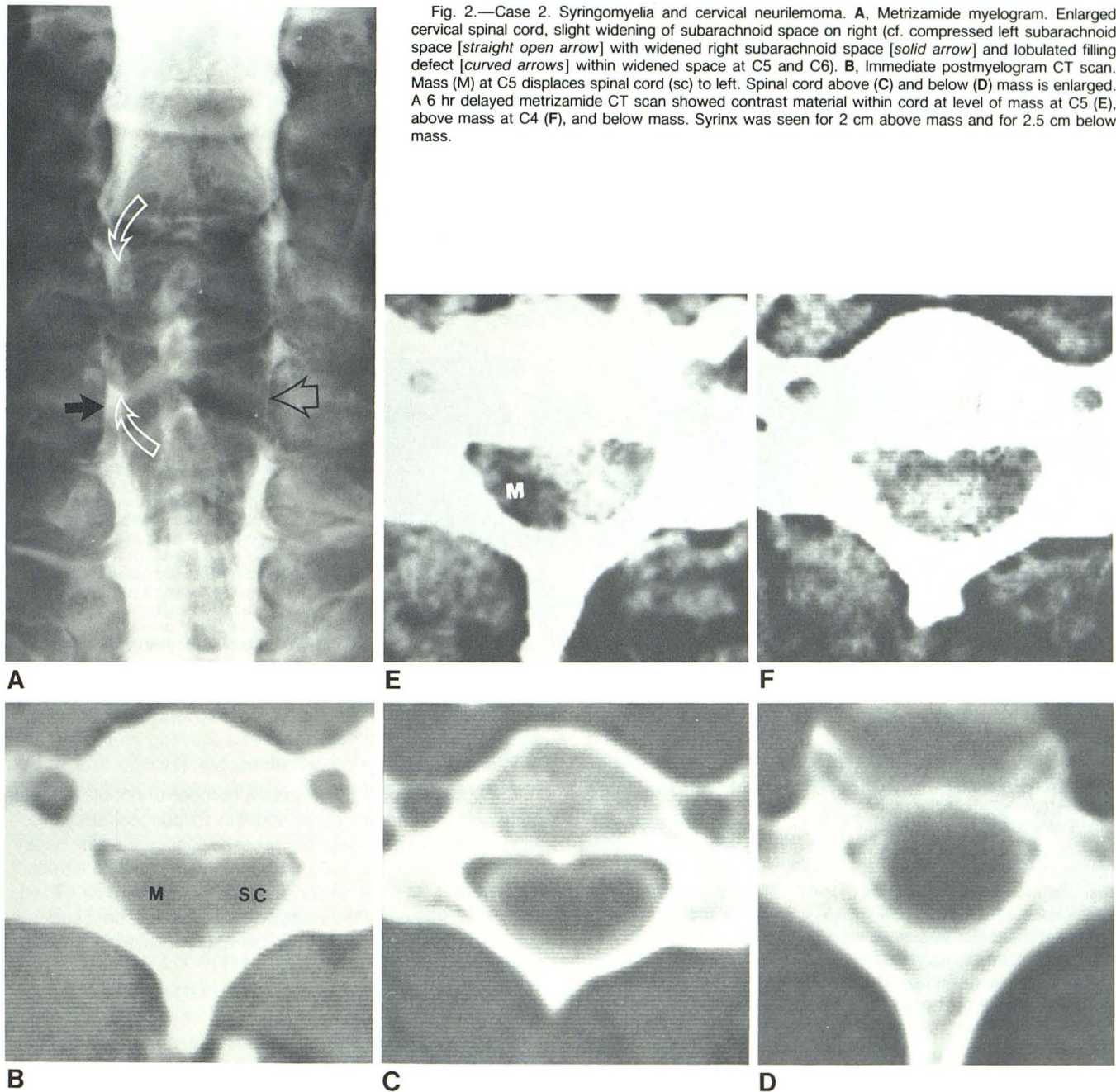


Fig. 2.—Case 2. Syringomyelia and cervical neurilemoma. **A**, Metrizamide myelogram. Enlarged cervical spinal cord, slight widening of subarachnoid space on right (cf. compressed left subarachnoid space [straight open arrow] with widened right subarachnoid space [solid arrow] and lobulated filling defect [curved arrows] within widened space at C5 and C6). **B**, Immediate postmyelogram CT scan. Mass (M) at C5 displaces spinal cord (sc) to left. Spinal cord above (C) and below (D) mass is enlarged. A 6 hr delayed metrizamide CT scan showed contrast material within cord at level of mass at C5 (E), above mass at C4 (F), and below mass. Syringomyelia was seen for 2 cm above mass and for 2.5 cm below mass.

a C5–T3 laminectomy was performed, and a large neurilemoma arising from the left dorsal root ganglion at C6 was excised. No attempt was made to enter the cord to drain or shunt the underlying syringomyelia. Over the next month there was moderate and gradual improvement in the strength of the patient's leg and she began to void spontaneously. Because there had not been a more dramatic change in her neurologic status, myelography and metrizamide CT were repeated, and an enlarged cord with an abnormal collection of intramedullary contrast consistent with a persistent syringomyelia was seen. Despite this finding, it was decided to follow her clinically because of her overall neurologic improvement.

Case 2

A 56-year-old woman was admitted to the hospital with complaints of right scapular pain, numbness of the right hand, and loss of strength in her right arm and hand. On physical examination, she was found to have atrophy of the right thenar muscles, decreased grasp of the right hand, a hyperactive right biceps reflex, and neck pain on hyperextension. In a fall 5 years before, the patient injured her neck and fractured her right clavicle, and because of this previous history of trauma, she was suspected of having a cervical radiculopathy secondary to either a herniated cervical disk or degenerative spon-

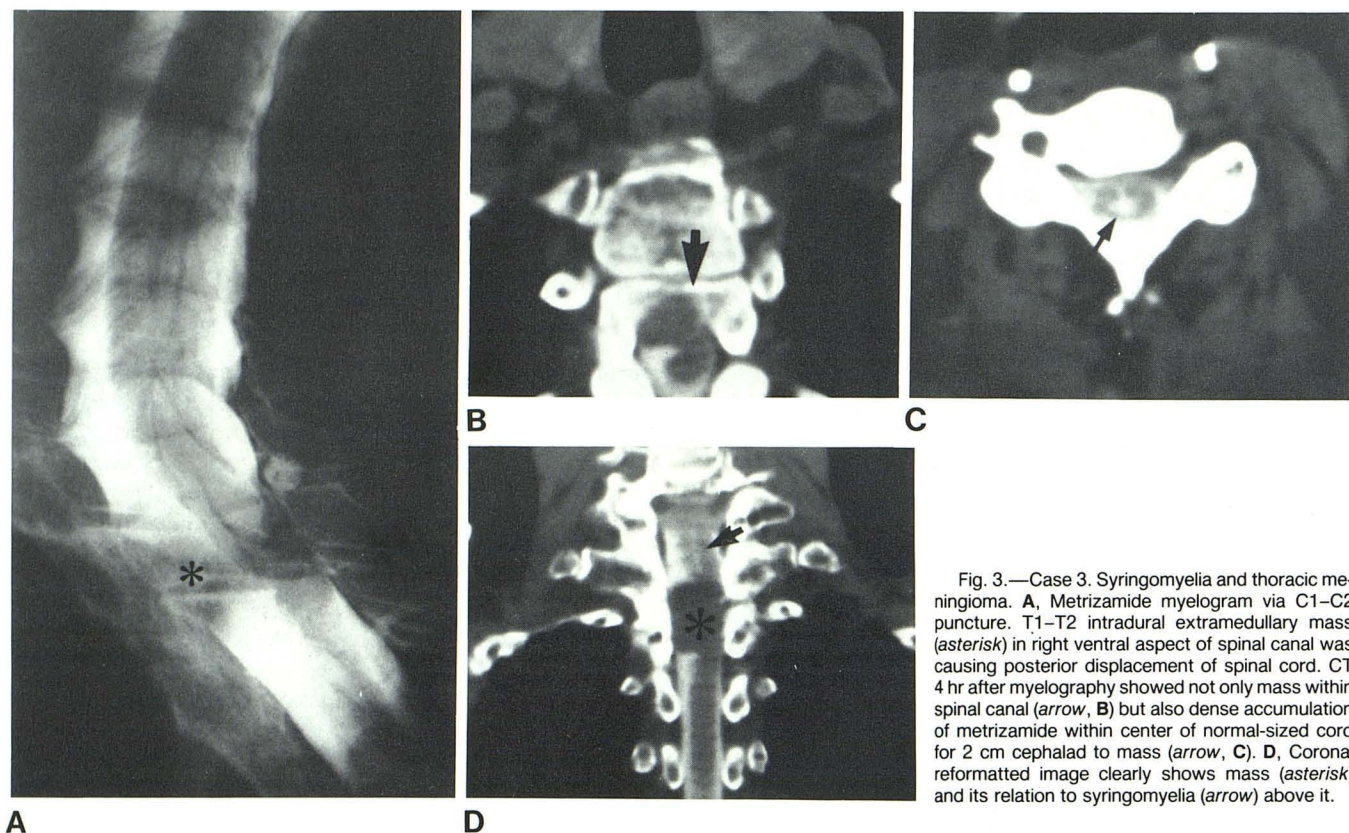


Fig. 3.—Case 3. Syringomyelia and thoracic meningioma. A, Metrizamide myelogram via C1–C2 puncture. T1–T2 intradural extramedullary mass (asterisk) in right ventral aspect of spinal canal was causing posterior displacement of spinal cord. CT 4 hr after myelography showed not only mass within spinal canal (arrow, B) but also dense accumulation of metrizamide within center of normal-sized cord for 2 cm cephalad to mass (arrow, C). D, Coronal reformatted image clearly shows mass (asterisk) and its relation to syringomyelia (arrow) above it.

dylosis. Metrizamide myelography (fig. 2A) showed an enlarged cervical cord and a filling defect on the right at C5–C6. CT immediately after the myelogram (figs. 2B–2D) confirmed a mass and an enlarged cord. Because of the uncertain nature of the enlarged cord, a delayed metrizamide CT scan (figs. 2E and 2F) was obtained and showed findings consistent with a syringomyelia at, above, and below the intradural mass. After myelography, the patient experienced only headaches and nausea. At surgery, a benign neurilemoma was removed. No attempt was made to aspirate or shunt the associated syringomyelia. Postoperatively, the patient did well, regaining strength in her right arm and hand and losing the numbness of the right hand and the pain in her right scapula.

Case 3

A 63-year-old woman had a 9 month history of progressive leg weakness and paresthesias and a 3 week history of urinary incontinence. On examination the patient had a T5 sensory level, diminished strength in both legs, left greater than right, hyperactive reflexes in her legs, and bilateral Babinski signs. Myelography showed a mass in the canal at T1–T2 (fig. 3A). There was no evidence of spinal cord enlargement. A CT scan 4 hr later (figs. 3B–3D) showed the mass and an associated syringomyelia above the mass. Six hr after the myelogram the patient's bilateral leg weakness worsened and she was placed on corticosteroids. Twelve hr later she had improved to her premyelographic status. At surgery, an anterolateral intradural meningioma, which extended from T1 to T2, was removed. Although the patient made a satisfactory postoperative recovery, she still demonstrated continued weakness and spasticity on a 6 month follow-up examination.

Discussion

Syringomyelia may result from a number of neoplastic and nonneoplastic conditions. In addition to congenital syringohydromyelia, cysts may be present within intramedullary spinal cord tumors [1, 3–6], form in conjunction with intracerebral tumors [1], be associated with arachnoiditis [7, 8], develop in previously traumatized cords [9–12], be a sequela of cord ischemia [13, 14], or be a consequence of long-standing spinal cord compression [1].

A syringomyelia forming as a result of spinal cord compression by an intraspinal extramedullary tumor has been considered rare, and reports of such cases have only sporadically appeared in print. In a review of the world's literature up to 1973, Barnett and Rewcastle [1] reported seven cases of pathologically proved syringomyelia associated with extramedullary tumors, five of which were thoracic and two of which were cervical in location. In six of these, the syringomyelia was of considerable length, extending well beyond the local area of the tumor to include the cervical and thoracic cord. Since that time, only one similar case has been reported [2], and in that patient a thoracic meningioma at T9 was found at surgery to be associated with a syringomyelia of the conus medullaris. Neither that patient nor any of the other reported cases was studied with CT in conjunction with myelographic contrast agents.

The diagnosis of a cystic cavity within the cord has been most reliably made by the use of CT of the spine 4–24 hr after myelography with a water-soluble contrast agent. Re-

ports of false-negative CT scans, in which a proved syrinx failed to fill [15], and false-positive CT scans, in which dense contrast material within the spinal cord turned out not to be a cystic lesion [10], have made this imaging method less than 100% reliable. Magnetic resonance imaging does offer hope that these cysts may be more easily and reliably demonstrated [16] than with metrizamide CT; however, it is possible that if these cysts become filled with fluid that is not exactly the equivalent of cerebrospinal fluid (CSF), signals of intensity similar to cord tissue, tumor, or edema may result. In those instances, identifying the cyst could be difficult.

In our three cases, delayed metrizamide CT showed not only the extramedullary mass but also evidence of dense concentration of contrast material within the spinal cord, consistent with syringomyelia. Two patients (cases 1 and 2) had dilated spinal cords, but in one case (case 3), the cord was not enlarged. Had Pantopaque rather than a water-soluble contrast agent been used, the diagnosis of a syringomyelia in cases 1 and 2 would have been more difficult and would have to have been based strictly on the presence of an enlarged cord, while in case 3 the diagnosis of a syringomyelia would not have been possible. Since intraoperative sonography was not used during surgery and a myelotomy was not performed, we do not have absolute proof that a syringomyelia was present. Nonetheless, the typical appearance on delayed metrizamide CT and the fact that cavitation such as this has been reported to occur in lesions that compress the spinal cord [1] make us confident that an intramedullary cystic cavity was indeed present in each case. The only other possible pathologic entity that would explain this type of metrizamide uptake by the spinal cord would be focal myelomalacia directly adjacent to the compressive intraspinal lesion; however, the relatively long midline accumulation of contrast material is typical of syringomyelia and not myelomalacia.

The pathophysiologic mechanism by which these cysts fill is, we believe, similar to the series of events that cause cysts within previously traumatized spinal cords to fill [9]. Long-standing constant compression of the spinal cord at the level of an intradural extramedullary tumor may lead to local nerve tissue and connective tissue damage manifested, in part, by enlargement of the Virchow-Robin spaces. The presence of these dilated extracellular, perivascular spaces in conjunction with altered CSF dynamics at the level of the tumor creates a situation in which abnormal amounts of CSF can enter the spinal cord. Eventually, over a long period of time, a cystic cavity, which may be in continuity with the central canal, gradually enlarges. As a result, the syrinx that forms may be located below the mass (case 1), both above and below the mass (case 2), or just above the mass (case 3). An alternative, but less attractive, theory to explain the presence of a syrinx would be that of ischemia caused by compression of the cord's vasculature by the mass. Animal experiments [13, 14] have shown cord cavitations, invariably located in the central gray matter, that may result from ischemia. Were, however, ischemia *per se* the underlying mechanism causing the syringomyelia, it is unlikely that the cysts would extend for significant distances away from the intraspinal mass. In addition, we do not agree with Blaylock [2] that the syringomye-

lia forms as a result of extrachoroidal production of CSF that then fills a dilated and obstructed central canal. By his theory the syrinx would always be located caudad to the extramedullary mass. Our cases 2 and 3 show that the syrinx may also be located either partly or totally superior to the mass. Unlike a congenital syringohydromyelia, the CSF in the type of syringomyelia we have described is not in free communication with the ventricular system and subarachnoid space, and we speculate that under these circumstances gradual enlargement of the syringomyelia with time, prior to tumor removal, may occur. Although removal of the tumor results in decompression of the cord and may improve the CSF dynamics within the spinal canal, it still will not alter the irreversible damage done to the cord, and its removal may not result in total decompression of the dilated intramedullary cavity. Our postulate appears to be confirmed by case 1, in which we showed a persistent syringomyelia 1 month after the removal of the spinal neurilemoma.

Although all of our patients' signs and symptoms are best explained by an extramedullary mass, rather than syringomyelia, we believe that the immediate postmyelographic clinical deterioration in two of our patients (cases 1 and 3) may have resulted from the combined presence of an intraspinal mass, which had caused a complete (case 1) and partial (case 3) myelographic block, and the syringomyelia. Because of this situation, metrizamide, a known neurotoxic agent, passed through the cord in a relatively concentrated volume and accumulated within the syrinx. Therefore, the spinal cord was bathed on the inside and the outside by metrizamide that was not in free communication with the entire subarachnoid spaces. In those two patients, neurologic deterioration was manifested by strikingly diminished strength in the legs that required steroid administration. Return to their premyelographic neurologic status occurred in 24 hr in case 1 and in 12 hr in case 3. Certainly the spinal tap and removal of CSF could not be blamed for the postmyelographic neurologic deterioration since a C1-C2 puncture, that is, above the level of the intradural extramedullary mass, was performed either alone (case 3) or in combination with a lumbar puncture (case 1). As a result of our experience with these cases, we suggest that whenever neurologic deterioration occurs after myelography in which a compressive spinal cord lesion is identified, delayed metrizamide CT be performed because an underlying syringomyelia may be found.

The radiologic diagnosis of a coexisting intradural extramedullary mass and syringomyelia may be difficult on routine myelography. As figures 1A, 1B, and 2A show, the widened cord may partially mask the presence of an extramedullary lesion, which can be diagnosed on delayed metrizamide CT. On the basis of our experience, we suggest that this combination of pathologic entities may be more common than has been previously believed and that the increasing use of delayed metrizamide and magnetic resonance may in time reveal the true incidence of this combination of lesions.

REFERENCES

1. Barnett HJM, Rewcastle NB. Syringomyelia and tumors of the nervous system. In: Barnett HJM, Foster JB, Hudgson P, eds.

- Syringomyelia*. London: Saunders, 1973:261-301
2. Blaylock RL. Hydrosyringomyelia of the conus medullaris associated with a thoracic meningioma. *J Neurosurg* 1981;54:833-835
 3. Quencer RM, Tenner MS, Rothman LM. Percutaneous spinal cord puncture and myelocystography. Its role in the diagnosis and treatment of intramedullary neoplasms. *Radiology* 1976;118:637-644
 4. Quencer RM. Needle aspiration of intramedullary and intradural extramedullary masses of the spinal canal. *Radiology* 1980;134:115-126
 5. Kan S, Fox AJ, Viñuela F, Barnett HJM, Peerless SJ. Delayed CT metrizamide enhancement of syringomyelia secondary to tumor. *AJNR* 1983;4:73-78
 6. Ferry KJ, Hardman JM, Earle KM. Syringomyelia and intramedullary neoplasms. *Med Ann DC* 1969;38:363-365
 7. Simmons JD, Norman D, Newton TH. Preoperative demonstration of postinflammatory syringomyelia. *AJNR* 1983;4:625-628
 8. Barnett HJM. The pathogenesis of syringomyelic cavitation associated with arachnoiditis localized to the spinal canal. In: Barnett HJM, Foster JM, Hodgson P, eds. *Syringomyelia*. London: Saunders, 1973:245-260
 9. Quencer RM, Green BA, Eismont FJ. Posttraumatic spinal cord cysts: clinical features and characterization with metrizamide computed tomography. *Radiology* 1983;146:415-423
 10. Quencer RM, Morse BMM, Green BA, Eismont FJ, Brost P. Intraoperative spinal sonography: adjunct to metrizamide CT in the assessment and surgical decompression of posttraumatic spinal cord cysts. *AJNR* 1984;5:71-79, *AJR* 1984;142:593-601
 11. Seibert CE, Dreisbach JN, Swanson WB, Edgar RE, Williams P, Hahn H. Progressive posttraumatic cystic myelopathy: neuro-radiologic evaluation. *AJNR* 1981;2:115-119, *AJR* 1981;136:1161-1165
 12. Rossier AB, Foo D, Naneedy MH, Wang AM, Rumbaugh CL, Levine H. Radiography of posttraumatic syringomyelia. *AJNR* 1983;4:637-640
 13. Woodward JS, Freeman LW. Ischemia of the spinal cord. An experimental study. *J Neurosurg* 1956;13:63-72
 14. Wilson CB, Bertran V, Norrell HA, Hukuda S. Experimental cervical myelopathy. II. Acute ischemic myelopathy. *Arch Neurol* 1969;21:571-589
 15. Aubin ML, Vignaud J, Jardin C, Bar D. Computed tomography in 75 clinical cases of syringomyelia. *AJNR* 1981;2:199-204
 16. Pojunas K, Williams AL, Daniels DL, Houghton VM. Syringomyelia and hydromyelia: magnetic resonance evaluation. *Radiology* 1984;153:679-683