Preoperative Demonstration of Postinflammatory Syringomyelia

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Syringomyelia secondary to arachnoiditis is a rarely reported entity, which has not been described in the radiographic literature. A series of eight patients who had either postinfectious, posthemorrhagic, iatrogenic, or idiopathic arachnoiditis was examined recently with metrizamide computed tomography (CT). These patients presented with progressive neurologic deficits that could not be attributed to the level of arachnoid changes on myelography. On immediate and/or delayed CT scans, metrizamide accumulation within the central part of the cord was identified, both at and above the level of the arachnoiditis. In some patients, the cord diameter was normal. Syrinx cavities were demonstrated and shunted at surgery with subsequent partial relief or arrest of the symptoms. The mechanism of development of postinflammatory syringomyelia is discussed.

Syringomyelia may result from craniocervical junction obstruction and anomalies, tumor, trauma, and postarachnoiditis [1]. Because of the variation in the clinical and myelographic appearance of arachnoiditis, the diagnosis of syrinx associated with arachnoiditis is difficult [2] and has rarely been reported. However, the advent of high-resolution computed tomography (CT) and its use with water-soluble contrast media has made the diagnosis of syrinx more reliable [3]. We review the entity of syringomyelia associated with arachnoiditis and present several recent cases of syrinx associated with arachnoiditis diagnosed preoperatively with metrizamide CT.

Materials and Methods

Eight patients with syringomyelia associated with arachnoiditis were studied. The arachnoiditis resulted from previous meningitis [2], intrathecal hemorrhage [1], or surgery [3], or it was idiopathic [2]. Ages were 30–66 years. In general, patients were studied because of progression of symptoms or extension of symptoms beyond the myelographic limits of known arachnoiditis.

Metrizamide CT was performed after myelography or as a primary study. If a myelogram was desired, it was done with routine technique. Afterward, the patient was placed in a semiupright position in bed. Following a 4–6 hr delay, the patient was rolled 360° several times and then scanned in the supine position with the neck flexed to avoid spillage of contrast material into the basal cisterns. If myelography had been done on the previous day, a primary metrizamide CT study was performed. In such cases, dilute metrizamide (3–5 ml, 170 mg I/dl) was injected under fluoroscopic control. The CT scan immediately followed. If a syrinx was suspected clinically and it was not detected on initial scans, a second 6 hr and even 24 hr delayed scans were obtained.

All patients were scanned with a G.E. 8800 CT/T scanner with 120 kVp, 320 mA, and scan speed of 9.6 sec. In all cases, a high-resolution extended-scale ReView software program was used for excellent intrathecal detail and contrast resolution.

Representative Case Reports

Case 1

A 32-year-old woman had progressive cranial nerve deficits. Eight years before she had a craniectomy for a presumed trapped fourth ventricle. At operation there were severe adhesions in the posterior fossa. The fourth ventricle was enlarged but not obstructed. Because of recent progression of new symptoms, a brainstem glioma was suspected, and metrizamide CT was performed (fig. 1). It revealed left basilar cistern adhesive changes, an enlarged fourth ventricle, and a syrinx extending into the medulla and upper cervical cord. A repeat craniotomy revealed an enlarged fourth ventricle and syringobulbia, which did not communicate. Again, adhesive changes were present in the posterior fossa. No tumor was evident. After shunting of the syrinx, there was improvement in the cranial nerve deficits.

Case 2

A 48-year-old man was admitted with progressive back and leg pain and increasing leg weakness. A previous thoracic laminectomy for suspected cord tumor revealed only extensive chronic arachnoiditis. A myelogram (fig. 2A) demonstrated a complete block at T9. The irregular appearance of the contrast material was consistent with arachnoidal adhesions. However, tumor could not be excluded. Metrizamide CT (figs. 2B and 2C) 6 hr later added considerable information. At the level of the block, a large syrinx cavity filled with contrast material and extended inferiorly to the conus. The cord was normal in size. Posterior to the cord was an area of ossification, so-called arachnoiditis ossificans [4]. Subsequent drainage of the syrinx led to partial relief of the symptom complex.

Case 3

A 66-year-old man was admitted with progressive arm weakness. Previously, he had undergone several Pantopaque myelograms and thoracic lumbar decompressive laminectomies for spinal stenosis.

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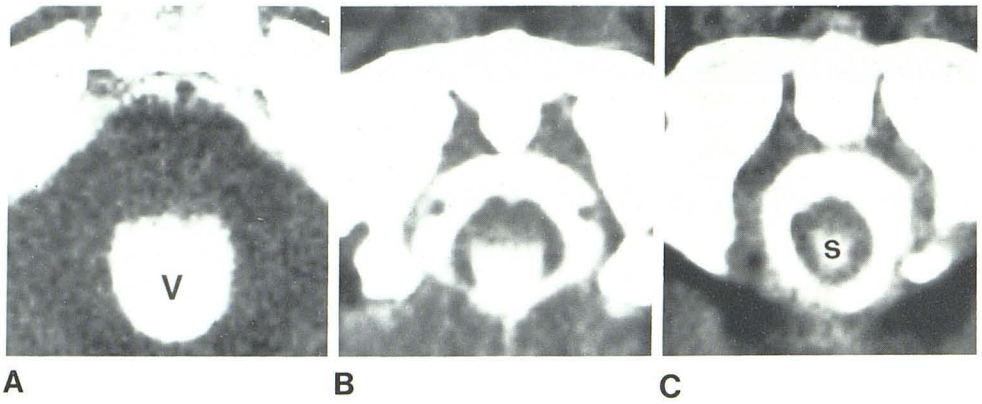

Fig. 1.—Case 1. Syringobulbia associated with arachnoiditis in 32-year-old woman with progressive cranial nerve deficits. Metrizamide cisternogram revealed basilar cistern adhesions. **A**, Metrizamide CT scan through posterior fossa. Fourth ventricle (V) is enlarged. **B** and **C**, Through medulla and upper cervical cord. Syrinx (s) extends from lumen of fourth ventricle into medulla and upper cervical cord. At surgery, there was no communication between syrinx and fourth ventricle.

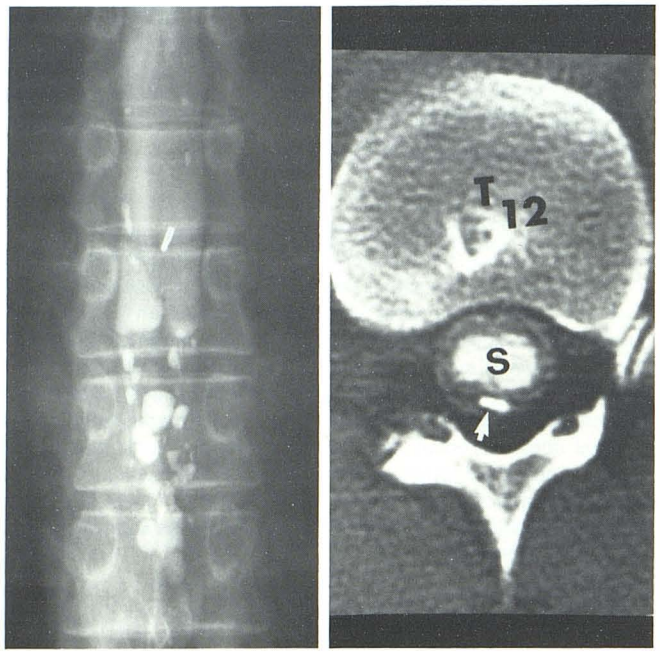

Fig. 2.—Case 2. Block and syrinx with arachnoiditis in 48-year-old man with progressive leg weakness over several years. **A**, Metrizamide myelogram with C1–C2 puncture. Irregular complete block at T9. Residual Pantopaque below block. **B**, CT scan at T12. Contrast material within syrinx (s) extends from T9 to conus. Note low-density rim of cord and thickened arachnoid surrounding syrinx. High density posterior to cord (*arrow*) represents ossification, or so-called arachnoiditis ossificans, proven at surgery.

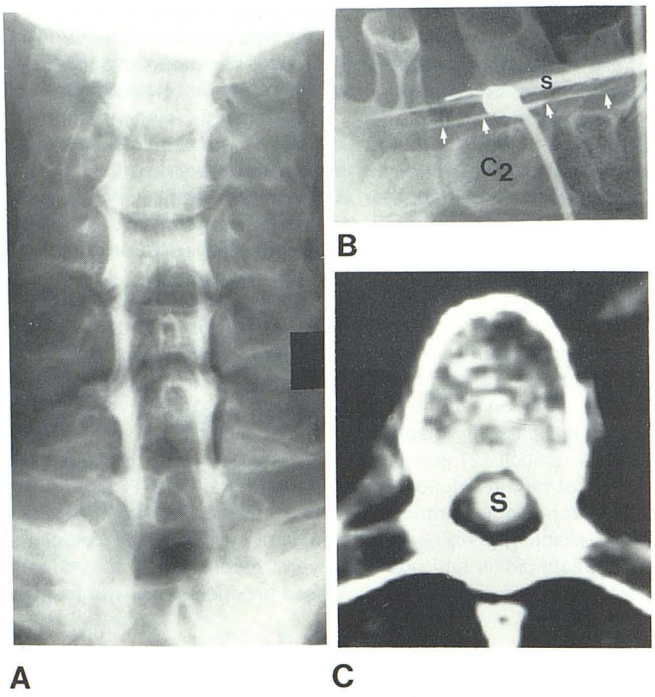

Fig. 3.—Case 3. Direct syrinx puncture in 66-year-old man with severe arachnoiditis and progressive arm weakness. **A**, Cervical myelogram. Cervical cord is slightly enlarged. **B**, Lateral cervical spine film after direct C1–C2 puncture of syrinx. Two contrast layers are within syrinx (s) and subarachnoid space (*arrows*). **C**, Immediate CT scan. Syrinx (S) extends from C2 to conus. Caudal extent allowed thoracic rather than cervical shunting of syrinx.

Myelographically, severe arachnoidal adhesions were seen in the thoracic and lumbar thecal sacs. These did not explain his arm difficulties. His cervical metrizamide myelogram (fig. 3A) revealed a slightly enlarged cord. The delayed CT scan was equivocal for syringomyelia. Because of the patient's symptoms and slightly enlarged cord, a syrinx was strongly suspected and, therefore, a syrinx puncture was performed. Using lateral fluoroscopy, 2 ml of metrizamide 170 mg I/dl was injected at the C1–C2 level (fig. 3B). Two layers of contrast material were seen within the spinal canal, one within the central syrinx and another within the subarachnoid space. To determine the extent of the syrinx an immediate CT scan was obtained (fig. 3C). Because the syrinx extended to the thoracic level, shunting of the syrinx avoided the brachial plexus with excellent results.

Discussion

Postinflammatory syringomyelia represents cavitation of the spinal cord resulting from arachnoidal adhesions. In 1869, Charcot and Joffroy [5] first described cervical cord cavitation associated with arachnoiditis. Since then, only 14 pathologically or surgically proven cases have been reported, none in the radiologic literature. This probably does not reflect the true incidence of this entity, but rather a previous inability to diagnose the lesion. In the past, a syrinx complicating arachnoiditis was suspected only on clinical

grounds because of symptoms extending beyond the limits of myelographic abnormality. Surgical exloration was needed to secure the diagnosis. The advent of metrizamide CT allows visualization of the intramedullary contrast-filled cavity as well as the adhesive arachnoid changes.

Several theories have been proposed to explain the occurrence of syringomyelia secondary to arachnoiditis [3, 6–9]. The proposed theories are centered on two basic mechanisms: (1) alteration of cerebrospinal fluid (CSF) and (2) vascular compromise. Appleby et al. [7] reported several cases of syringomyelia associated with arachnoiditis. They believed that the cavitation resulted from adhesive blockage of the fourth ventricle. They theorized that the interference with drainage from the fourth ventricle leads to an increased intraventricular pressure and central canal dilatation with eventual parenchymal cavitation. To explain the fact that the syrinxes and central canal dilatation were usually limited to the spinal cord and often did not extend up to the fourth ventricle, they proposed that the adhesive changes around the upper cord and medulla prevented canal dilatation in these areas. Aboulker [8] believes that cavitation may develop in the cord without increased pressure and dilatation of the central canal. Rather, a stenosis at the level of the foramen magnum from the arachnoiditis alters the normal ascent of the CSF, leading to an increase in the CSF pressure around the cord. The CSF then is forced into the cord through the parenchyma, posterior roots, and arteries. The final result is cord edema and eventual cavitation. In support of this, a recent study by Dubois et al. [9] demonstrated that indeed there is penetration of fluid, specifically metrizamide, into the spinal cord.

The premise that arachnoidal adhesions lead to vascular compromise and eventual ischemic cavitation has gained support from both experimental and clinical studies [6, 10]. In 1954, McLaurin et al. [10] produced chronic adhesive arachnoiditis in dogs by injecting kaolin into their cisternae magnae. The animals were sacrificed and examined between 9 days and 4½ months postinjection. Severe arachnoiditis in the posterior fossa and around the spinal cord was found in all cases, and, in more than half of the animals, there was some degree of cord cavitation. The subarachnoid vessels were embedded in thick fibrous adhesions and appeared constricted. Because posterior fossa and basal cisterns were involved in the process, an element of fourth-ventricular obstruction might have entered into the cavity formation. However, animals sacrificed early showed patchy necrotic lesions of the posterior cord at the gray-/white-matter junction. These early lesions were patchy, discontinuous, and not related to the central canal or fourth ventricle. Dogs sacrificed later had larger posterior cord cavities that were continuous with the fourth ventricle. They concluded that: (1) cavitation of the cord results from vascular insufficiency produced by constriction of the vessels in the course of the fibrotic development; (2) the posterior location of the cavity was consistent with the work of other investigators [11], who found that terminal branches of the anterior and posterior spinal arteries are in the posterior cord, and the effects of vascular compromise would center on this area; and (3) the cavities often enlarge with time, which leads to communication with the central canal of the cord.

Barnett [6] added support to the thesis that vascular compromise from adhesive arachnoiditis can lead to ischemic necrosis and cavitation. After reviewing the literature, he identified seven cases of reported syringomyelia associated with arachnoiditis involving the spinal cord without extending above the foramen magnum. He added seven cases from his experience. He found that the cavities had no communication with the fourth ventricle or subarachnoid space, and he concluded from studying the pathology of these cases that vascular compromise plays a role in the production of the syrinxes.

From the experimental and clinical evidence, it seems likely that syringomyelia may result from either of the two basic mechanisms described. If the inflammatory process involves the posterior fossa, CSF dynamics may be altered, causing central canal dilatation or cord edema. In addition, arachnoid adhesions may cause vascular compromise resulting in eventual ischemic cavitation.

We reviewed eight recent cases of syrinx associated with arachnoiditis. Preoperative metrizamide CT demonstrated cavities in the medulla, cervical, and/or thoracic cord. Myelography was the first step in the evaluation of all of these patients with suspected syrinxes except for the case of syringobulbia (case 1) where the initial clinical diagnosis was brainstem glioma. The myelogram is useful in excluding other causes of the presenting symptoms and in localizing the level of the block, cord enlargement, or arachnoid adhesions.

The findings of syrinx associated with arachnoiditis are characteristic on metrizamide CT scans. The syrinx appears as a contrast-filled cavity within the substance of the cervical and thoracic cord (figs. 2 and 3) or the medulla (fig. 1). The cord at the level of the syrinx may be normal (fig. 2), small, or large (fig. 3). The syrinx may be restricted to a short section of the cord or even extend the entire length of the cord (case 3). In addition to the cavitation, there is evidence of arachnoid adhesions in these cases. Arachnoidal adhesions in the cervical thecal sac present on CT as arachnoidal thickening (fig. 2) or obliteration, calcification (fig. 2), block, or cyst. In the lumbar thecal sac, there is blunting of the root sleeves, clumping of the roots, peripheral adherence of the roots to the dura, block, or cyst.

Occasionally, for unknown reasons, a syrinx will not fill with contrast material even with delayed scanning. If there is a strong clinical or myelographic suspicion of a syrinx in such a case, a direct puncture of the syrinx at the appropriate level followed by immediate CT scanning will demonstrate the location and extent of the cavity (fig. 3). The demonstration of the extent of the syrinx is helpful not only in diagnosis but also in planning the level of surgical exploration and drainage. If the cavity extends from the cervical cord into the thoracic cord, the neurosurgeon usually elects to shunt the syrinx below the brachial plexus to avoid possible injury.

Finally, in addition to establishing the presence and extent of a syrinx, CT is useful in demonstating associated pathology accompanying arachnoid adhesions and excluding other causes of cord enlargement or block. Not uncommonly, arachnoid cysts occur with arachnoiditis. Barnett [1] noted five arachnoid cysts associated with the 14 syrinxes secondary to arachnoiditis that he reported. We discovered one arachnoid cyst in our series of syrinx patients.

Surgery in advanced cases of arachnoiditis is generally contraindicated. However, shunting of a syrinx associated with arachnoiditis has been helpful in alleviating symptoms or arresting progression of disability in reported cases in the literature and in this series.

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