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Syringobulbia Extending to the Basal Ganglia

M. C. Valentini,¹ C. Forni,² and M. Bracchi³

Syringomyelia and syringobulbia are diseases in which there is a tubular cavitation of the spinal cord and medulla. CT with intrathecal injection of water-soluble contrast material (CT myelography) and, more recently, MR imaging have led to a revolution in the diagnosis of syringomyelia. Syringobulbia is much rarer than syringomyelia and usually associated with it [1]. Radiologic demonstration of the cavity in syringobulbia is very difficult, since the cavity in the brainstem is narrow and slitlike.

We report a case of associated syringomyelia and syringobulbia in which there is radiologic demonstration of the cavity extending from the lower thoracic cord through the brainstem and basal ganglia up to the left lateral ventricle (syringocephalia).

Case Report

A 23-year-old woman was admitted because of progressive weakness of the left lower limb that, over the preceding year, had extended to the left upper limb. The patient related the onset of symptomatology

to a craniocervical trauma suffered 6 years earlier. During the last months she began to complain of dorsal back pain and urgency of micturition. Neurologic examination revealed a left hemiparesis without facial involvement, brisk deep-tendon reflexes, and left Babinski sign. Cranial nerves were not impaired and sensation was normal. X-ray examination and CT of the cervical spine showed a mild widening of the cervical canal and an ununited C1 posterior arch.

Brain CT examination and myelography were performed (Figs. 1 and 2), and a microsurgical approach through a T2 median laminectomy was carried out 1 month later. Myelotomy showed a high-pressure, fluid-filled cavity within a well-preserved cord parenchyma. Clear CSF flowed from the cavity and continued to flow during the whole procedure. The horizontal branches of a T-shaped silicone rubber catheter were introduced into the cavity in a cephalad and caudad direction, and iopamidol 200, 1.5 ml, was injected through the vertical branch to determine the whole extent of the syrinx. X-ray examination showed opacification of the left lateral ventricle, the head being at a lower level with respect to the dorsal region. The T-tube vertical branch was then introduced caudad to drain into the thoracic subarachnoid space. Tomography was performed immediately after surgery (Fig. 3). CT scans (Fig. 4) showed that in the medulla the cavity was situated ventrally, in the region of the left pyramid, and in

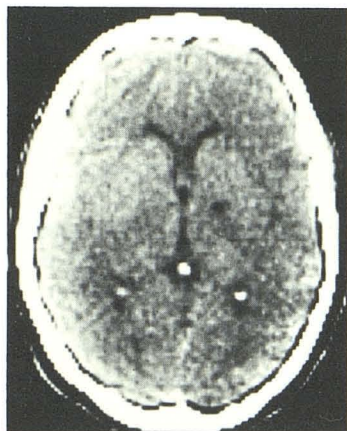


Fig. 1.—CT scan shows small hypodense area in left basal ganglia.

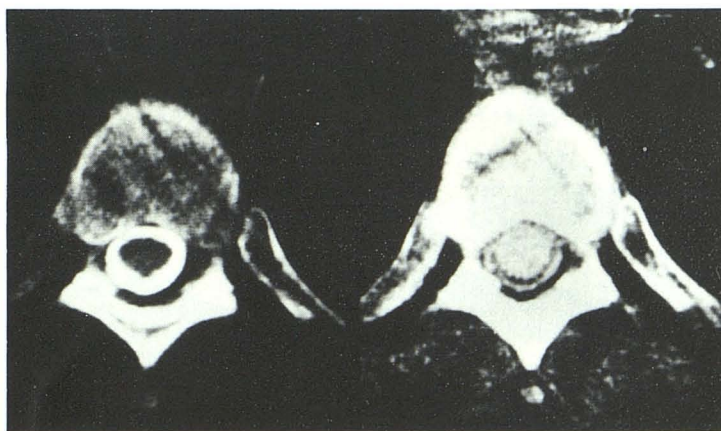


Fig. 2.—A, Lumbar myelogram (iopamidol 300, 16 ml) shows widened dorsal and cervical cord. B, CT scans 1 hr (left) and 6 hr (right) after myelography show filling of cavity.

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¹ Department of Neuroradiology, Centro Traumatologico Ortopedico, Via Zuretti 29, Torino, Italy. Address reprint requests to M. C. Valentini.

² Department of Neurosurgery, Centro Traumatologico Ortopedico, Torino, Italy.

³ Department of Neuroradiology, Istituto Neurologico, Milano, Italy.

the pons between the corticospinal tract and the pontine nuclei. No direct communication with the fourth ventricle was seen. In the mesencephalon the cavity occupied transversally the basal part of the left peduncle, in the region of the substantia nigra.

Subsequent CT controls showed that the whole ventricular system, as well as the basal cisterns and the subarachnoid spaces of the vault, became opacified. The cavity within the basal ganglia remained opacified for 48 hr, thus verifying that there was a slight CSF stream through it.

The patient had immediate postoperative relief of dorsal pain and of micturition urgency, but no improvement of the hemiparesis. A

long-term control performed 9 months later demonstrated that aside from an improvement of the left leg paresis, deep-tendon reflexes had also become brisk in the right leg. Moreover, spasms of the left sternocleidomastoid muscle, fasciculations of the tongue, and mild left facial hypoesthesia had appeared, demonstrating a rostral evolution of damage to the neural pathways.

An MR study* performed in the same period showed that the syringomyelic cavity was wide, with an upper limit at C1. No alterations could be detected in the medulla, while a narrow cavity was recognizable in the pons, the mesencephalon, and the basal ganglia (Fig. 5).

Discussion

Syringobulbia is frequently a cranial extension of the cervical syringomyelia. According to the literature, the extension of the cavity to the pons or to a higher level is extremely rare [2-6]. In 1906, Spiller [7] described a case of syringomyelia extending through the medulla, the pons, and the cerebral peduncle to the upper part of the internal capsule. All the cases described were autopsy studies. To our knowledge, our case is the first with an in vivo radiologic demonstration of a syringomyelic cavity extending to the basal ganglia and communicating with the lateral ventricle.

In 1932 Jonesco-Sisesti [8] described three possible positions of the cavity in syringobulbia: (1) anterolateral directed from the floor of the fourth ventricle external to the hypoglossal nucleus; (2) from the fourth ventricle along the median raphe; and (3) ventrally situated, between the pyramid and the inferior olive (the rarest location).

* Philips Gyroscan 0.15-T resistive magnet.

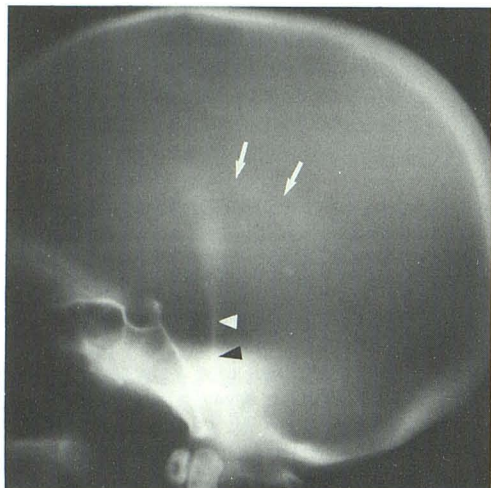


Fig. 3.—Tomogram after direct opacification of cavity (iopamidol 200, 1.5 ml) shows syrinx extending through brainstem and basal ganglia (arrowheads). Opacification of left lateral ventricle (arrows) is also evident.

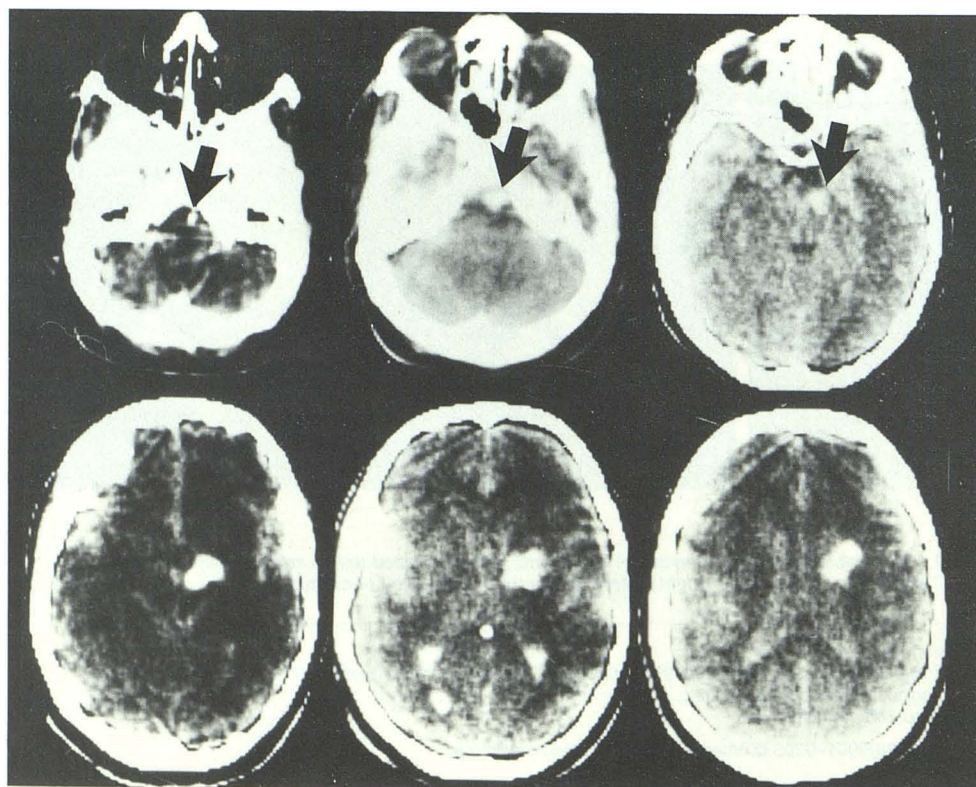


Fig. 4.—CT scans show syrinx extending to ventral part of medulla, pons, cerebral peduncle (arrows), and internal capsule.

Fig. 5.—MR images reveal small cavity in left cerebral peduncle and in left basal ganglia (arrows).



In our patient, as in Spiller's case [7], the cavity lies ventrally, as in the third group described by Jonesco-Sisesti [8]. If we assume that syringobulbia is the cranial extension of syringomyelia [9], this location, remote from the fourth ventricle, could explain the exceptional cranial extension of the cavity. It is interesting that the patient did not have the classical syringomyelic dissociated anesthesia but a left hemiparesis, which can be related to a lesion at the level of or below the pyramid decussation, while the cavitation of the left basal ganglia appeared to be asymptomatic. In our case the syringomyelia-bulbia-cephalia is associated with a tonsillar ectopia and an ununited C1 posterior arch. It is worth emphasizing that the onset of the symptoms was related to a cervical trauma, as has been described for many other cases in the literature [10]. In our case the trauma likely had a precipitating effect in producing a further extension of a former asymptomatic cavity [11].

The radiologic findings also deserve some comments. The demonstration of syringobulbia is rare and difficult because of the small size of the slitlike cavity in the brainstem. As syringobulbia is generally the cranial extension of the syrinx of the spinal cord, it is likely that spinal cord syringography is the best technique for showing it [12–15]. This method has become more informative with water-soluble contrast materials due to their miscibility with CSF [16–18]. In fact, in our case, the water-soluble contrast medium opacified the whole cavity and the ventricular system, while a little air bubble could not overcome the narrow portion of the cavity at C1, because of its high surface tension. MR clearly showed the syringomyelic cavity in the spinal cord; it showed less clearly the cavity in the mesencephalon; but it could not demonstrate the smallest cavity in the brainstem. Increased spatial resolution in MR units, as is now taking place, will also be likely to show such a small cavity [19]. The greater ability to demonstrate small lesions in the parenchyma, as well as the absence of bony artifacts, make MR the examination of choice in suspected syringobulbia.

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