Case Report

Post-traumatic syringomyelia following complete neurological recovery

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Objective: To describe the later neurologic deterioration secondary to the appearance of a post-traumatic syringomyelic cavity, in a patient who, in the initial phase, had an incomplete spinal cord lesion (ASIA C), which improved to ASIA E.

Methods: A 52-year-old male patient who, at the age of 19 (1965), suffered a spinal cord injury. He presented with a fracture of the sixth and seventh cervical neurological segment at the time of the lesion, evolving to ASIA E. Nine years after the traumatism, he began to feel pain accompanied by a sensory and motor deficit.

Results: With the aid of myelography and MRI, the existence of a syringomyelic cavity was detected, which extended from the fourth to the seventh cervical segments. The patient was operated on, on various occasions, placing a syringo-subarachnoid shunt. The neurological status of the patient continued to deteriorate and, at present, he has a complete lesion below the fourth neurological cervical segment with a partially preserved sensitive area up to T1.

Conclusion: The development of the syringomyelic cavity could be one of the causes of later neurologic deterioration in patients with traumatic spinal cord injury with neurological recovery '*ad integrum*' in the initial phase of spinal cord injury.

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Introduction

Post-traumatic syringomyelia (PTS) is an infrequent complication of traumatic medullary lesions; its incidence ranges between 1 and 4% in symptomatic patients (patients with symptoms of syringomyelia) and it can occur a few months after the initial lesion or even several years later.^{1–4} The incidence of PTS increases when diagnosis is obtained by neuroimaging in subjects with no associated neurological symptomatology. It has also been described in patients with a background of vertebral fracture with or without transient neurological deficit at the time of the lesion.

The mechanism by which the syringomyelic cavity is formed is not fully known but, in general, the most accepted theory is the one proposed by Williams, who considers it is produced but the combination of focal necrosis and the pressure gradients in the cerebrospinal fluid (CSF) at the level of the lesion (the wave and aspiration theory).^{4,5}

Clinical symptoms produced by the syringomyelic cavity are not specific and can appear above or below the initial neurological level; they include pain or paresthesias, sensory disturbances, weakness, spasticity, autonomic system dysfunction and bladder

disorders. Generally, this symptomatology develops insidiously, but sometimes it can appear abruptly with a Valsalva manoeuvre such as produced by coughing.

Magnetic resonance is the recommended diagnostic technique, its main limitation being that it does not distinguish between a simple cystic cavity and one which will develop into a syringomyelic cavity.

We report a case of a patient with post-traumatic syringomyelia, the clinical symptoms of which appeared 9 years after the medullary lesion; a lesion which had progressed to complete neurological recovery ASIA E.⁶

Case report

A 52-year-old male patient who, at the age of 19 (1965), suffered spinal cord injury as a consequence of a plunge. He presented with a fracture of the sixth and seventh vertebrae with an incomplete lesion, ASIA C, below the seventh cervical segment. The neurological lesion evolved to ASIA E (complete recovery) and, on discharge, the patient did not present any limitation insofar as physical performance: he had global muscular balance at 5, no sensory deficit, voluntary sphincter control, no erectile dysfunction and he even practiced certain sports.

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At the age of 28 (9 years after trauma) he began to feel pain at the level of his left arm, later accompanied by a sensory and motor deficit in the left leg which later spread to all four extremities. At this time he underwent surgery, being submitted at a C5–C6 and C6–C7 disc extraction; his evolution led to progressive neurological deterioration but maintaining the ability to walk without use of aids until 1981. At this time, with the aid of myelography, the existence of a syringomyelic cavity which extended from the fourth to the seventh cervical segments was detected, with a complete block at the lower border of the seventh cervical vertebrae. Surgery was performed consisting of a C7 laminectomy, opening the dura and finding at C7 level an intramedullary cyst cavity that was sectioned and a syngo-subarachnoid shunt was placed. A complete block by adhesions of the subarachnoid space was discovered at the top of the cyst cavity.

Neurological deficit continued to increase, but the patient was still able to walk with the aid of a correcting splint on his left leg and with two walking sticks.

At the age of 43 (1989) he was seen at our center for the first time; he had an incomplete lesion below the fourth neurological cervical segment, ASIA C, and he was able to move in a manually propelled wheelchair. Magnetic resonance demonstrated a cavity extending from C1 to C7 vertebral segments with divisions in the interior and signs of arachnoiditis (Figures 1 and 2). Surgery was performed once again to drain the cavity, and a cyst-subarachnoid shunt was placed.

The patient's neurological status continued to deteriorate and, at present, he has a complete medullary lesion below the fourth cervical segment with a partially preserved sensitive areas up to T1; he depends on an electrically propelled wheelchair to move. He has no voluntary sphincter, and an evacuation rhythm every 48 h has been established with the aid of laxatives; he is dependent on the aid of a third person for carrying out his daily activities.

The last magnetic resonance study performed in July 1998 showed a syringomyelic cavity extending from C1 to T1, with intramedullary gliosis and signs of arachnoiditis in the posterior perimedullary space from C4 to T1.

Discussion

The present article describes the case of a patient with post-traumatic syringomyelia that appeared 9 years after a medullary lesion secondary to a cervical fracture and incomplete SCI with complete neurological recovery.

The literature published on this subject provides eight cases of similar characteristics; thus, Barnett and Jousse⁷ review six cases of PTS (two of them presented severe arachnoiditis without vertebral fracture in the area of the initial traumatism), and Bleasel *et al*⁸

Figure 1 T1 MRI (sagittal). Cavitation on the cervical spinal cord extending from C1 to C7 vertebral segments

describe another two. However, most of the PTS series recently published^{9,10} are of patients with incomplete spinal cord injury; no other cases of complete neurological recovery have been described.

At present it is believed that the incidence of syringomyelia is similar in complete and incomplete lesions, with no relation to the severity of the initial spinal cord injury. Backe *et al*¹¹ describe a similar incidence among patients with an ASIA classification motor index lower or higher than 50. Nevertheless, the incidence of syringomyelia after complete spinal cord injury is two or three times higher than with incomplete lesions in other series.^{9,12,13} Rossier *et al*¹ describe an incidence of 3.9% and 2.4% after complete and incomplete lesion, respectively.

Several theories explain the pathogenesis of this condition, the most widely accepted being the one proposed by Williams^{4,5,14} based on the concept of initial formation of a cystic cavity at the level of the fractured vertebral area, followed by a less well-understood mechanism of extension and growth of the cavity. Initially, the cavity is formed by a process of hemorrhagic necrosis, liquefaction of the hematoma and liberation of lysosomal enzymes, asso-

Figure 2 T1 MRI (axial). The syrinx can be seen located mainly on the right side of the spinal cord

ciated with the phenomenon of tethered cord produced by arachnoiditis. Cavity formation frequently begins at the level of the fracture in the grey matter between the dorsal horns and posterior columns, which is a relatively avascular zone between the dorsal and ventral arterial supply. The secondary extension of the cavity is due to an increase in epidural venous pressures, causing intracordal fluid movements (theory of 'slosh' and 'suck').

According to this mechanism, posttraumatic sub-arachnoid scarring and adhesions around the syrinx cause blockage of the normal cerebrospinal fluid wave that is continually being produced by epidural venous pulsations. This pulse pressure is consequently transmitted to the intraparenchymal posttraumatic cystic cavity. Williams^{4,5,14} suggests that this repetitive sloshing or cerebrospinal fluid against the cyst may explain the frequently seen extension of the syrinx above or below the level of cord damage.

Our patient had significant arachnoiditis at the site of the trauma that contributed to the growth of the cavity.

The point where the cavity begins corresponds to the level of the fracture, generally coinciding with the

lower part of the cavity. The literature describes the existence of syringomyelic cavities with a caudal extension to the origin of the vertebral fracture. The close relationship between the syringomyelic cavity and the vertebral fracture supports the thesis that the latter is a relevant factor for the development of the cavity, as occurs in the case we present.^{12,13}

Post-traumatic syringomyelia appears both in patients submitted to conservative therapy (as in our patient) and in those who have undergone surgery in the acute phase of the spinal cord injury; a similar incidence has been described for both groups. No large series comparing both groups exists in the literature, and even the series with surgically-treated patients are scarce.

Another point discussed in relation to the development of the cavity is its correlation with the degree of medullary canal stenosis. Wang *et al*⁹ found no correlation between them, but in recently published articles such as those by Schurch *et al*² and Perrouin-Verbe *et al*,¹⁵ a relation between both factors in pointed out, thus supporting Williams' pathogenesis theory.

The time interval between spinal cord injury and the onset of symptoms is variable and can occur months or even years after the lesion, 9 years in the case of our patient. It has been described to occur up to 40 years later.⁸

According to most authors the most frequent symptoms of onset are pain, which is initially unilateral, followed by changes in sensitivity, motor deficit and areflexia, all of which correspond to the symptoms observed in our patient.

At present, magnetic resonance (MR) is the best technique for the study of syringomyelia. In general, the cavity is eccentrically sited and extends through several vertebral segments, oscillating between three and 10 segments depending on the series (seven segments in our case).^{9,16,17}

The role of surgery is still controversial in relation to this condition. In general, surgical treatment is recommended in cases of progressive syringomyelia with neurological deterioration, and conservative therapy is reserved for non-progressive asymptomatic syringomyelic cavities, with periodic clinical magnetic resonance follow-up.^{12,14,17} Williams^{4,5,14} recommended that the syrinx should be shunted to a low pressure system such as the pleural or the peritoneal cavity for effective drainage, and an internal posttraumatic gibbus or arachnoiditis, if present, should be dealt with at the same time. According to him, abolition of a pressure gradient and free flow of the cerebrospinal fluid are vital factors in minimising the risk of recurrence of syringomyelia.

Good results have been described by drainage of the cavity with the aid of a syngo-subarachnoid shunt, although the latter tends to obstruct rather frequently (as in our case), leading to recurrence of the cavity. At present, the preferred procedure is to establish a shunt between the syringomyelic cavity and a low pressure

system implanted at thoracic or abdominal level, resulting in a much lower incidence of obstruction in comparison to previous types of shunt.^{13,17,18}

In general, the results with surgery are favourable. All series agree as to the relief of pain in almost all cases, followed by improvement of the motor deficit; the least observed improvement is the recovery of sensitivity, although the series presented by Lyons¹³ and El Masry³ describe a postoperative recovery of sensitivity of 91% and 73%, respectively. In the case of our patient, both operations were followed by worsening of the neurological level.

In summary, few cases of appearance of PTS years after a medullary lesion in patients with complete neurological recovery (ASIA E) have been described. Despite the fact that this condition is infrequent, it must be considered in all patients with a history of vertebral fracture, since it can be underestimated and even overlooked sometimes due to the time it takes to develop.

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