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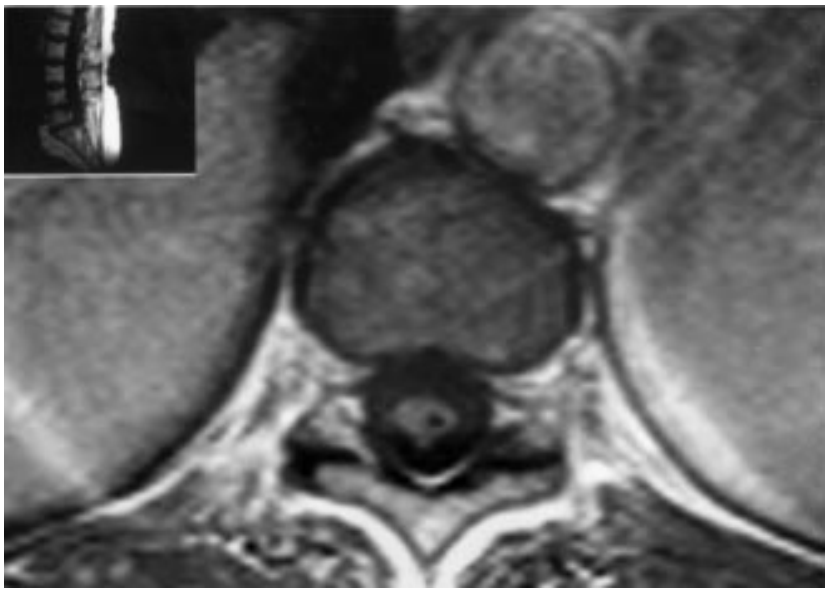
Attacks of pain in the leg from classic syringomyelia

Neurologists should not be surprised if the cause of a disorder is remote from its effect, such as a parasagittal meningioma causing foot dragging, or a sacral ependymoma leading to deafness as the first sign of siderosis of the nervous system. Yet we were for some time bewildered by the following problem.

A 67 year old woman consulted us in 1996 for attacks of pain in the left leg that had started in 1978 and had gradually increased in severity and frequency up to three to four attacks an hour, each lasting a few minutes and so severe that she had to stop anything she was doing to nurse her pain. It was sharp and stabbing in character and radiated from the gluteal region to the groin, and further down to the lateral part of the upper and lower leg. Between attacks she was completely free of pain. The stabs also woke her up at night, six or seven times. In 1988 investigations at another hospital had established the diagnosis of cervical syringomyelia, secondary to tonsillar ectopia (Chiari I malformation); a year later a syringopleural drain was inserted. Subsequently the syrinx collapsed, but the attacks of pain continued unabated. Examination (in 1996) showed normal power and sensation in the arms, a thoracic kyphosis, and on the left side of the trunk a suspended level of hypaesthesia and hypalgesia, extending from just under the nipple down to the left leg, as far as 10 cm below the knee; vibration sense was abolished below the sternum on both sides. Power in the legs was normal; the tendon jerks were very brisk on the right and sluggish on the left, both plantar responses flexor. Repeated MRI studies confirmed a collapsed syrinx in the cervical region, extending throughout the thoracic cord, and deviating to the left at the level of the lumbar cord (figure). Many analgesic, antidepressive, antiepileptic, and antiarrhythmic drugs had previously failed or failed again in our hands, as did sympathetic blockade or transcutaneous electrostimulation. Implantation of an epidural stimulator gave considerable relief, the intensity of the pain decreasing by more than half; this was sustained up to the last contact, nine months after implantation.

Leg pain with syringomyelia has so far been reported only in patients with a syrinx confined to the lumbosacral cord, secondary to trauma or tumour.[1 2] In our patient bouts of pain in the leg were associated with classic syringomyelia, which starts at the level of the cervical cord and results from obstruction of the CSF flow at the foramen magnum, most commonly by tonsillar ectopia.[3 4] We propose that the pain resulted from disturbed impulse transmission in the posterior grey matter at the left side of the lumbar cord, the pain partially matching the area of sensory loss. It is not unusual for symptoms to correspond to paracentral cavitations only, the central part of the syrinx being clinically silent.[5] Dysaesthetic pain from a paracentral syrinx is known to occur in the arm.[6] Also in those cases the response to surgical treatment is unpredictable.[6] Epidural stimulation was the only measure that made our patient's life again bearable.

We are grateful to Dr HE van der Aa (Enschede) for referring the patient and to AL Liem (St Antonius hospital, Nieuwegein) for implantation of the epidural stimulation device.



T1 weighted MRI at the level of the D10-D11 intervertebral disc. Hypointense lesion in the left dorsolateral region of the lumbar cord, corresponding to the posterior horn.

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Anti-GQ1b and anti-GT1a IgG antibodies in a patient with acute demyelinating polyradiculoneuropathy without ophthalmoplegia

Anti-GQ1b IgG antibodies have been often detected in the serum of patients with Miller Fisher syndrome or Guillain-Barré syndrome with ophthalmoplegia. These antibodies may participate in the development of ophthalmoplegia. Our patient with acute demyelinating polyradiculoneuropathy had no ophthalmoplegia despite having anti-GQ1b and anti-GT1a IgG antibodies in her serum.

An 80 year old woman was admitted because of weakness of her limbs. Ten days before admission to our hospital, after a common cold, she developed difficulty standing and paraesthesia in her lower legs. Within a few days, walking and prolonged sitting were nearly impossible and her arms became weak. On the day of admission, she developed dysphagia. There was no diplopia or ptosis at any time.

Her ocular and facial muscles were normal. She had difficulty in drinking water. Her limbs were weak (upper limbs MRC grade 3/5 and lower limbs 2 to 3/5). Deep reflexes were diminished or abolished. Pathological reflexes were not elicited. Superficial and deep sensation were abnormal below the knees. Stool culture was negative for *Campylobacter jejuni*. The CSF contained 159 mg protein/dl and no cells.

The right tibial motor nerve conduction velocity (MCV) was slow and the peroneal nerve did not respond. The median and ulnar nerves showed delayed distal latencies. Slowing of sensory nerve conduction velocity (SCV) was found only in the median nerve. F waves could not be elicited from the median and tibial nerves. A sural nerve biopsy on the 24th day showed loss of myelinated fibres without cell infiltration.

Dysphagia disappeared on the 27th day. Strength in her upper limbs recovered to MRC grade 4/5 a week later. One month after admission, she could walk with a little help. The protein concentrations in her CSF were 82 and 57 mg/dl after a month and four months respectively. Three months later, the median and tibial MCVs were still slow and could not be elicited in the peroneal nerve.

The distal and F wave latencies in both the median and ulnar nerves were delayed. F waves could not be elicited in the peroneal or tibial nerves. Median SCV did not improve. The patient was discharged with no disability in activities of daily life four months after admission.

The patient's serum was tested at intervals for antiganglioside antibody activities by enzyme linked immunosorbent assay (ELISA).[1] GM1a, GM2, GM3, GD1a, GD1b, GT1a, GT1b, and GQ1b were prepared from bovine brain.[2] The antigenic solution contained 20 pmol/50 µl of each ganglioside. Each patient's serum was tested in triplicate at 1:100 dilution, and binding detected with horse radish peroxidase conjugated goat antihuman IgM or IgG antibody (Jackson Immunoresearch Laboratories, Inc, West Grove, PA, USA) at 1:1000 dilution. On admission, the serum reacted with both GQ1b and GT1a at a dilution of 1:3200. These two antibody activities, expressed as optical densities, fell to 1/2 in parallel with her improvement, but they could still be detected in her last blood sample (at 16 weeks). IgG antibodies against the other gangliosides were not detected at any time, nor were IgM antibodies to any ganglioside detected.

To determine whether these two antibodies react independently with each ganglioside or react with a common epitope sharing with GQ1b and GT1a, an absorption study was performed using GQ1b or GT1a coated polystyrene beads as described previously.[1] The titre of anti-GQ1b IgG antibody decreased when preincubated with GT1a coated polystyrene beads, and vice versa for the anti-GT1a IgG antibody. Based on these results, we hypothesised that these antibodies reacted with a common epitope sharing GQ1b and GT1a.

Chiba *et al*[3] found anti-GQ1b IgG antibodies in patients with Miller Fisher syndrome and also with Guillain-Barré syndrome with ophthalmoplegia. They showed immunochemically that GQ1b was rich in the paranodal regions of the human oculomotor, trochlear, and abducens nerves. These anti-GQ1b IgG antibodies also reacted with GT1a. Yuki *et al*[4] found anti-GQ1b IgG antibodies in eight patients with acute paresis of the extraocular muscles but without ataxia. These results suggested that anti-GQ1b IgG antibodies had a part in the development of ophthalmoplegia. We have also detected anti-GQ1b and anti-GT1a IgG antibodies in six patients with Miller Fisher syndome and two with Guillain-Barré syndrome accompanied by ophthalmoplegia.We also found that these antibodies reacted with a common epitope sharing GQ1b and GT1a (unpublished data). Kimura *et al*[5] have already reported a patient with acute relapsing sensory dominant polyneuropathy without ophthalmoplegia who had an anti-GQ1b IgG antibody which did not react with GT1a. Further clarification of the relation between anti-GQ1b IgG antibody and the development of ophthalmoplegia in Miller Fisher syndrome or Guillain-Barré syndrome is required.

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Can trauma alone to the trigeminal root relieve trigeminal neuralgia? The case against the microvascular compression hypothesis

Idiopathic trigeminal neuralgia (ITN) is increasingly regarded as being due to microvascular compression of the trigeminal sensory root, either by an artery or a vein close to the brainstem.[1] Yet vascular contacts are found in entirely asymptomatic cases, no vascular contacts are found in some asymptomatic patients, and, most importantly, ITN suddenly switches off, even for years, only to return later, in the face of continuing vascular compression.[2] Despite this, microvascular decompression produces immediate virtually complete (98–100%) relief in 82% of the cases and 64% after 10 years.[3] Adams has suggested that, ITN being a hyperfunctional disorder of the brainstem, microvascular decompression "produces chronic trauma to a sensitive zone of the cranial nerve... by the dissection necessary and by the manipulation required microvascular decompression produces sufficient trauma to achieve interference of normal functioning of that nerve", thus dampening the abnormal brainstem activity responsible for ITN.[2] Failure to achieve an initial result or early recurrence would imply insufficient surgical trauma.[2] We evaluated this hypothesis in our patients submitted to microvascular decompression. Forty one out of 410 patients with ITN have had keyhole microvascular decompression in the posterior fossa from December 1986 to February 1996 at the neurosurgical pain relief unit of the University of Turin. Twenty six patients showed pronounced arterial or arterovenous compression, three distally (group 1), three mild or disputable contacts (group 2), two slight and three pronounced venous compressions (group 3), five with arachnoiditis, two with a sharp root kink, four distally (group 4). In four cases (group 5), no anomaly whatsoever could be found, despite adequate magnification and careful exploration; vessels were noted close to the root, but no contact or groove could be seen. Thus the root was gently "massaged" with a microdissector; no other manoeuvre was attempted. These four patients included one woman and three men, with a mean age of 59 (range 54–66). All had typical ITN which at some point could no longer be controlled by drugs at adequate dosage. In all other cases, the vessels were separated from the root by interposing Surgicel; the arachnoiditis was dissected to free the root and, in one patient with slight venous compression partial (25%) rhizotomy was also carried out. In several patients, "massage" of the trigeminal root was performed.

All patients were relieved by surgery. Follow up disclosed the following recurrences: two in group 1 (follow up 4 months-9 years), none in group 2 (follow up 2.3–5 years), none in group 3 (follow up 1.1–5 years), two in group 4 (follow up 1–6 years), one in group 5 six years postoperatively (follow up 6 months-8 years). Importantly, one of the group 1 recurrences was operated on and no vascular contact whatsoever could be found (partial rhizotomy was elected). The only group 5 recurrence could be controlled by drugs. Whereas this was seen in other groups as well, all four group 5 patients showed transient slight to moderate hypaesthesia in two to three branches after surgery, a hallmark of trauma.

In 1961, Taarnhøj reported that simple manipulation of the trigeminal root by running a nerve hook along the root obtained 60% long term pain free results over a mean of 6.5 years (longest follow up 10 years).[4] Gardner and Miklos also obtained 67% pain free results over 4.5 years by manipulating the trigeminal sensory root at the point of crossing the apex of the petrous bone,[5] suggesting that manipulation not at the root entry zone (as suggested by Jannetta), but peripherally at the petrous apex produces as good results as microvascular decompression. As no other report considered the problem, the microvascular compression hypothesis for ITN gained favour. Our data suggest that (1) trauma alone can ensure long term relief; (2) this relief is similar between patients showing microvascular compression and those without; it should be stressed that several patients of the microvascular compression group were "massaged" to ensure a control with the group without.

The trigeminal nerve root is surrounded by many arteries. In 60% of the roots, the trigeminal vessels form arterial rings encircling at least half of the root or its entry zone in asymptomatic patients,[6] justifying the frequency of vascular contacts at large. Despite a plethora of vessels reported to compress the trigeminal nerve in a recent series,[3] initial relief could not be achieved in 18% of the patients. Recently, a group reported electrophysiological data supporting the concept of trauma during microvascular decompression, despite attempts to limit the procedure to simply moving the vessel.[7] The trauma hypothesis would explain recurrences of successful microvascular decompression in which no new compression is found (for example, our case and see Yamaki *et al*[8]) at re-exploration.

Adam's contention is being appreciated in the case of hemifacial spasm. Payner and Tew,[9] discussing their results with microvascular decompression for this disorder, stated that "...perhaps there is truth that the early success in most patients results from minor trauma... to support this theory, additional analysis is needed of the long term follow up in patients without vascular compression and...treated by "manipulation" alone". This is what has been done in this study.