Research Article

A Microvascular Decompression Coupled to an Unilateral Intradural Rhizotomy for Cervical Dystonia (Spasmodic Torticollis) Results On 30 Patients

Franco Caputi[*], Raffaelino Roperto, Luciano Mastronardi

Department of Neurosurgery, S. Filippo Neri Hospital, Rome, Italy

[*]**Corresponding author:** Franco Caputi, Department of Neurosurgery, S. Filippo Neri Hospital, Rome, Italy via Marco Besso 34, 00191 Roma, Italy.

Abstract

Background: Surgery remains an effective option to treat Cervical Distonia or Torticollis despite the advance of conservative treatments. Neurologists favor Deep Brain Stimulation (DBS), on the assumption that there is a basal ganglia dysfunction. However, traditionally torticollis has been treated with Sternocleidomastoid Muscle (SCM) denervation intervening on the 11th nerve either intradurally or at the entrance into the SCM muscle fibers. Furthermore, there is well-documented literature on 11th nerve vascular conflict as a cause of torticollis. In our 30 cases, we resolved a vascular conflict with the 11th nerve and severed its C1-C2 rootlets to denervate the SCM and obtain an immediate release of its pathologic spasm and contraction. We exposed the upper cervical spinal cord and the 11th nerve with a novel minimally invasive unilateral approach.

Methods: We retrospectively evaluate 30 patients operated for CD. They had a unilateral compromise of SCM muscle which resulted from EMG and MRI studies, compatible with a vascular conflict. These patients underwent a C1-C2 hemilaminectomy, with a unilateral enlargement of the occipital rim. We disregarded the complexity and pattern of head deviation. We theorized a conflict between the 11th nerve and the vertebral artery or PICA. In some of them, we could identify such anomalies on preoperative MRI.

Results: A conflicting artery was found in all cases. It was the vertebral artery in 27 cases, the PICA in 2 and anterior spinal artery in 1. Overall a positive outcome (head realignment with a preserved range of motion without a need of further medication) was obtained in 23 patients (76,6%). Abolishing abnormal activity in the affected SCM corrected complex patterns of head deviation and lead to an improvement of 85% as measured on the Toronto Western Spasmodic Torticollis Rating Scale (TWSTRS).

Conclusion: A reduced unilateral approach focusing on the 11th nerve rootlets that innervate the abnormal SCM and resolving the underlying vascular conflict is an effective and acceptable way to treat a subgroup of CD.

Introduction

Spasmodic torticollis is a form of focal dystonia resulting from uncontrolled hyperactivity of neck muscles, mainly the SCM. Contemporary authors name it CD. According to Albanese CD "It is characterized by sustained or intermittent muscle contractions of the cervical muscle that cause abnormal, often repetitive, movements, postures, or both" [1]. Hyper-active muscles do not synchronize their activity with head and neck movement and are out of voluntary control. The head and neck may be simply rotated (torticollis), tilted (laterocollis), retroflexed (retrocollis) or anteflexed (antecollis), but deviations my combine [2-4].

As the most compromised muscle is the SCM, innervated by the 11th cranial nerve, denervating procedures focusing on intradural accessory nerve and cervical roots have been practiced since the late nineteenth and early twentieth centuries [5]. In 1920, McKenzie described the intradural section of the anterior cervical roots in combination with the section of the spinal accessory nerve [6]. Taking a different approach, Bertrand popularized the use of peripheral denervation procedure by sectioning the 11th nerve branches under the SCM [7]. In recent years Deep Brain Stimulation (DBS) has gained approval as first-line surgical treatment for CD to desynchronize an altered electrical activity in the sensorimotor loop [8,9]. In fact, previously neurosurgeons had operated on the basal ganglia by stereotactic surgery. Mundinger between 1972 and 1986, had operated 162 patients with CD reporting good results [10]. Since then studies have demonstrated nonspecific structural abnormalities in the basal ganglia by Voxel-Based MRI Morphometry (VBM) and Diffusion Tensor Imaging (DTI), but specific histological and histochemical alterations have not been demonstrated yet [11-13].

However, since the SCM and other hyperactive muscles are driven by the 11th nerve, we wondered if this nerve itself could be primarily affected. Jannetta had shown the relevance of a vascular conflict in triggering abnormal firing from a cranial nerve as it happens for trigeminal neuralgia, facial spasm and even torticollis [14,15]. Such results have been widely confirmed [16,17]. As earlier investigator supposed, it suggests that, a peripheral mechanism may drive muscle control anomalies typical of CD [15,16,18]. This study regards CD of 11th nerve origin as a special entity which responds to microvascular decompression with intradural rhizotomy through a unilateral approach.

Clinical Material and Method

We studied 30 patients, 18 women and 12 men, chosen among 60 evaluated for torticollis, who were unsatisfied with conservative treatment. Selected patients had a clear involvement of SCM on one side (Figure 1), 24 with torticollis (rotocollis) and a variable degree of lateral shift, 5 with laterocollis and one antecollis coupled to kyphosis (Figure 1B). We paid special attention to shoulder elevation /anterior displacement, as it indicated a postural adjustment and a possible progression of the disease toward scoliotic deformities, which could hamper the result of surgery. Such deformities with passing time could become fixed and irreversible because of osteoligamentous changes. The duration of the disease ranged from 2 to 16 years, with a mean of 6 years. The average time from onset to diagnosis was of 2.8 years. The last 20 patients were tested on a Toronto Western Spasmodic Torticollis Rating Scale (TWTRS) with an average score on torticollis severity of 29, a disability score 24 and a pain score of 12. All subjects were under the age of 60, with a mean age of 41.

Figure 1: A typical patient before and after surgery.

Figure 1B: One of the patients at the onset of symptoms (A and B), and seven years later (C), immediately before surgery. It shows how postural deformities progress with time. The results achieved after 11th nerve microvascular decompression and SCM denervation, 5 days from surgery (D).

Symptomatology

In each patient wry neck had begun with a subtle sense of tension and occasional spasms that got worse with stress. Symptoms subsided, at least initially, during the night or while lying down. Pain was not the initial concern for any of them, but almost every patient was later studied for cervical pathology and often referred for orthopedic consultation. By patients' own accounts, symptoms evolved and head deviation progressed with passing time. Symptoms were always characterized by muscle spasms fluctuating over the day, with moments of break and others of intensified distress and pain. For three patients pain had become a characterizing element together with muscle tension and was described as intolerable. For the majority of patients, pain, wasn't a concern. Younger people cited changes to their social habits as the main complaint. Although not always administered, Minnesota Multiphasic Personality Inventory revealed depressive tracts in 7 patients; out of the whole group of torticollis patients, 19 were under therapy with psychotropic drugs. Only two patients had movement disorders outside of the cervical musculature, both with facial tics. One of them had an associated vascular conflict with

his facial nerve; this was clearly evident on MRI scans. No one had swallowing problems and no patient reported a family history of dystonia. Another patient had a huge lipoma over the neck which had no apparent effect on torticollis. The lipoma was later removed. A second patient had an internal carotid artery aneurism as an occasional finding, which was embolized before surgery on torticollis.

Previous Therapy

Prior to surgery, all patients had received conservative treatment which included, along an almost identical scale of complexity, antirheumatic agents and pain medication, muscle-relaxants, benzodiazepines, antiepileptics, antidepressants, and antipsychotic drugs. Among the antiepileptic drugs, the most widely used were the γ – Aminobutyric Acid (GABA) enhancing agents. Everyone at some point entered a therapeutic protocol with a botulinum toxin treatment. Twenty seven had been treated in such a way for years with declining and overall unsatisfactory results; three had only be treated for few months without benefit. Two patients had a premotor cortex stimulation without results and electrodes had been withdrawn at least one month prior to our surgical procedure. No one had DBS. Therapeutic attempts included physical therapy in every patient and occasionally biofeedback, sometime with psychological supportive measure. Reporting on the effectiveness of previous treatments, three-quarters of the patients stated that they had had the most benefit from botulinum toxin, but wasn't satisfactory. Benzodiazepines and physiotherapy were the most common adjuvant, often complemented with pain medication.

Clinical Examination

Clinical examination included a careful evaluation of patterns of head deviation. Only one SCM had to be involved (Figure 1). We noted which one was affected by spasms and the direction to which the chin pointed; this was considered the main indicator of rotatory deviation. We identified four pattern of deformities: purely rotational (torticollis), head tilt (laterocollis), head anteflexion (antecollis) or retroflexion (retrocollis), or a combination. Often, accompanying contractions were observed on the splenius either on the affected side or on the other side. We interpreted these as attempts to readjust head deviation and look ahead. Thus it happened that in 23 patients the shoulder opposite to the contracted SCM elevated and displaced anteriorly with an accompanying sagittal or lateral shift. Such complex adjustment brought scoliotic deformities demonstrable on plain Xray in 26 patients. Only one patient, presented the characteristic feature of antecollis, with marked kyphosis generated by the forward bending of the head. Also in this patient there was a unilateral contraction of the SCM (right side) as shown in Table 1. One other patient presented with retrocollis, with relevant involvement of the trapezius together with a contracted SCM. We never evaluated patients with general dystonia, probably because patients with complex deformities and demonstrable encephalic anomalies (i.e. metabolic abnormalities or other encephalic pathology) were never referred by the neurologists who had been following the patients.
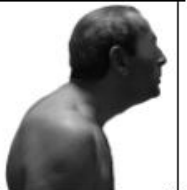

Muscle Involved	Rotation only	Involving shoulders	Antecollis
Left SCM 16	3	13	
Right SCM 14	1	12	1

Table 1: This table reports the involved SCM and how often the deformity extended to the shoulder girdle or produced a forward bending . Only 4 patients had a pure rotational deformity, the remaining 26 had a more complex deformity.

Electromyography

Every patient had electromyography showing evidence of spontaneous and burst suprasegmental activity. They had stopped treatment with botulinum toxin in the previous three months. Those selected for the surgery had a prominent compromise of SCM on one side, with electrical hyperactivity, and lack of inhibition when activating the contralateral SCM. Normally, when the head rotates, the SCM which promotes the movement becomes active, while the contralateral SCM relaxes and becomes electrically silent (inhibited). On the contrary, we observed that if the patient tried to rotate the head to correct the deviation, it would hasten bursting activity on the affected SCM (lack of inhibition). An abnormal activity was usually recorded in the paraspinal muscle, but a possible involvement of other muscles, mainly the contralateral splenius did not affect the surgical choice as long as head deviation was consistent with a sustained contraction of the compromised SCM.

Radiologic Studies

A cervical spine Xray was often done as an initial diagnostic evaluation early in the course of the disease. It was useful to rule out structural anomalies, either acquired or congenital, which could justify head deviation, as for a rotary sub-luxation. It often showed spondylotic ridges and rotoscoliotic changes consequential to muscle hyperactivity. Routine chest X-rays detected the result of compensatory posture showing scoliotic and kyphotic curves of the thoracic spine. Five patients had spiral CT scans with 3 D reconstruction to better evaluate bone structure and the pulling direction of contracted muscles, which would show an increased transverse diameter. Every patient had an MRI including sequences in Time of Flight (TOF) and balanced steady-state to evaluate the intradural transition of Vertebral Artery (VA) and possible conflict with the 11th nerve, the McKenzie branch, or the 1st cervical root. Figure 2 clearly shows a contact between the vertebral artery and the 11th nerve in one of the operated patients. While the vertebral artery and its relation to the 1st anterior cervical root were always visible, we could see rarely contact between the vertebral artery and the 11th nerve. We never visualized the McKenzie branch. Head and neck deviation with lack of symmetry between left and right, plus spasms and discomfort of examined patients made exams longer than usual and affected results impeding fine evaluation of the region of interest. However, the MRI was effective to rule out or describe concomitant spinal pathology and exclude medullary lesions. Eleven patients had cervical discopathy or osteophytes, out of surgical interest. Since our surgical strategy only considered unilateral and limited denervation, swallowing studies were done only in two patients; they resulted normal or slightly altered.
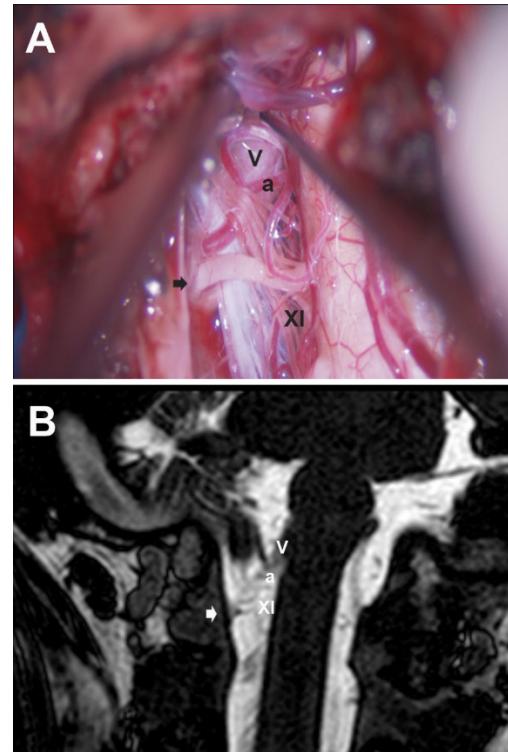

Figure 2: The operative finding (A) in correlation with the preoperative RM (A) in the same patient. It shows the vertebral artery (V) crossed by the posterior root (white arrow), the anterior spinal artery (a) and the 11th nerve (XI).

Surgical Technique and Relevant Anatomy

The goal of surgery is to explore and clear any vascular conflict between the vertebral artery and the 11th nerve, identifying and sectioning the eleventh nerve rootlets which contract the SCM muscle on the affected side. Thus, the exposure is unilateral on the side of the pathologically tightened SCM. The operation is performed under general anesthesia, without the use of a muscle relaxing agent. The patient is positioned prone, in a Concord position with three pins head rest to flatten the cervical lordosis. Needle electrodes are mounted to monitor the SCM (mastoid, sternal and clavicular branch) and the ipsilateral splenius and the trapezius. Stimulation and recording are done with Nymbus, by Imodia. The suboccipital muscles are divided at the midline keeping the incision as small as possible, but enough to expose the spinal cord on one side. We save the spinous processes, although sometime we plane their asperity, for an unobstructed view on the surgical field. Hemilaminae from C1 to C3 are removed, C1-C2 in the last 10 patients. A small sub-occipital craniectomy on the

same side is required to enlarge the occipital foramen and allow a better exposure of the upper part of the spinal accessory emerging underneath the cerebellar tonsil. At this point, we bring in the operating microscope and open the dura with a linear incision, through the foramen magnum, exposing posterior roots with 11th nerve crossing over the dentate ligament. The vertebral artery is not immediately visible, hidden by the dentate ligament, by arachnoid band or both.

We do not expose yet the anterior roots, and always stimulate nerve rootlets pertaining to the 11th nerve while recording the resultant muscle contraction. This nerve is composed of a spinal part which originates as low as the fifth cervical metamer through spinal rootlets arising from the motor cells, in the lateral part of the anterior horn of spinal cord. It runs parallel to the spinal cord, underneath the posterior roots and above the dentate ligament to enter the cranium, where it merges with a cranial part and exits through the jugular foramen.

The 11th nerve can be observed along its course as it appears beyond the cerebellar tonsil and descend parallel to the cord and over the dentate ligament as already said. Conflict, if present, can now be noted and evaluated as the nerve crosses the vertebral artery. Usually the artery enters at C1 level with frequent asymmetry compared to the other side, as seen in pre-operatory MRI. Difference would regard its course and diameter. Most of the time this artery pushes from below on the nerve fixed by arachnoid bands, then the artery crosses the nerve and goes up to the clivus. Rarely it is the posterior cerebellar artery which loops too far below the tonsil that mingles and conflict with spinal accessory nerve rootlets. In one case it was the anterior spinal artery to conflict with 11th nerve, as in Figure 3.
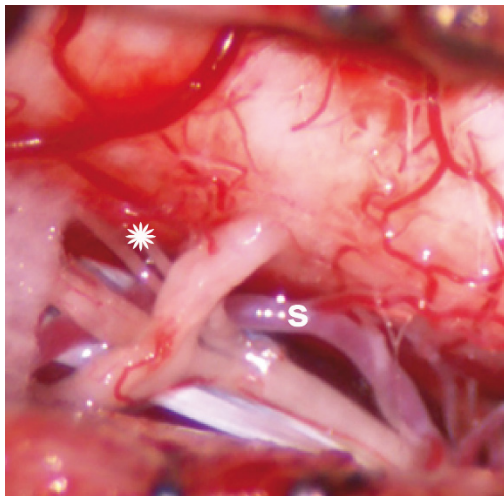

Figure 3: This picture shows a conflict between eleventh nerve rootlets and anterior spinal artery.

Only spinal rootlets which activate the affected SCM are sectioned. Once this part is completed, the dentate ligament is released exposing the anterior roots at each corresponding level. Again, only rootlets directed to the affected SCM are severed. We never found a positive electromyographic response below C1, so we no longer expose the anterior roots below C1. Special attention should be paid to the occasional presence of the McKenzie branch, send out by the eleventh nerve rostral to the vertebral artery. It descends to join the anterior C1 nerve root at the exit from the spinal canal. Spinal rootles to the trapezius are spared, as this muscle is usually not involved with spasmodic torticollis. The diaphragmatic response has been never searched or seen. Once the 11th nerve is more mobile because pertaining rootles from the spinal cord have been severed, the vascular conflict is resolved interposing SURGICEL® FIBRILLAR™ (Ethicon). The closure follows the standard procedures.

Post-Operative Course

All patients but one were kept in the hospital for a mean period of 8 days. The improvement of their posture in all but one was immediately apparent and progressed with passing time, generally within one month. All were sent for a rehabilitation program that included active exercises especially focusing on potentiating corrector muscles and stimulating proprioceptive feed-back [19]. It wasn't necessary to assist the recovery of the patient's ability to turn the head toward the direction opposite to the denervated SCM. Movement towards this side was always present before surgery and reappeared as the head re-aligned. On the contrary movement against the spastic SCM was limited before surgery but widened with time after a successful surgery. One patient who remained in hospital much longer had a CSF fistula with subsequent hydrocephalus, which had to be treated by V-P shunt without neurological sequelae. His head position and posture normalized. Another patient had a CSF fistula, which was treated by local wound care and CSF drain. He did not require wound revision. No patient had post-operative swallowing problems and no one died as a result of surgery or post-operative complication.

Long Term Results

The first patient treated with such unilateral approach was operated in 2006, we since totaled thirty patients on whom we can report. Other patients were operated earlier on both sides through a bilateral laminectomy according to the standard approach as described by Friedman and al. in 1993 and are not included in this group [18]. Patients in this study have been followed by the operating surgeon for at least one year and have been seen and questioned by an independent staff member not involved in their surgical or medical management. Follow up evaluation considered torticollis severity score, pain and disability as specified by the TWTRS. Results were considered excellent if severity score was below 6, disability was below 3 and no pain. Results were rated fair if severity score was below 15 but above 6, disability was below 7 and pain was present but not above 2. According to this

evaluation method, 23 (76,6%) patient were considered to have achieved an excellent results, 5 (16,6%) a fair result and two (6,6%) were considered a failure. Among the "fair" group was considered the patient who developed post-operative hydrocephalus but still regained a normal head alignment. Twenty two patients (73 %) did not complain spasm any more, 6 (20%) still reported some muscle tension, 2 (6,6%) remained with spasm. Patients with fair results still required botulin toxin treatment, which had better efficacy compared to treatment prior previous to surgery and decreased their severity score of a mean of 4 points, their disability score of a mean of 3 point and brought their pain to 0, thus further improving the result of surgery.

When we only considered 20 patients in whom we could couple preoperative and post-operative TWTRS score, a more detailed and effective post-operative changes came out. Indeed significant changes affected deformities, disabilities and pain scales (ANOVA, $P < 0.01$). Prior duration of disease did not seem to affect the result of the procedure, probably because patients were relatively young and arthrosic changes were relatively mild. Thoracic scoliosis always disappeared or ameliorated after surgery and the following rehabilitation process, except for the patient considered as a failure. A picture of a patient before surgery, 1 and 2 and 8 years after surgery is shown as an example (Figure 4). All patients but one regained full range of motion recovering the ability to turn toward the denervated SCM, which as a result of surgery would not obstacle head rotation on this direction. Rotation to the opposite side was maintained by the compensatory mechanisms of other muscles while lateral bending and circumduction were unaffected.

It was impressive to note that the elevated acromial tip contralateral to the contracted SCM, an almost constant indicator of a compensatory attitude consequential to the torticollis, realigned so the shoulder on both side were leveled and symmetric to each other (Figure 4). Once a good result was achieved, rotatory deviation of the trunk disappeared. Results remained unmodified or improved in 23 patients followed for more than two years. One patients had dropped to the fair results.

Figure 4: This picture shows a progressive correction of the deformities after surgery: B at one year and C at 2 years and 8 years follow-up

Discussion

It may be controversial to regard CD as an entity caused by 11th nerve dysfunction. In fact, movement disorders experts consider dystonia as an anomaly in the circuits of the basal ganglia that project to the motor cortex [20].

Its neuroanatomical substrate is not well understood [21,22]. Clinically, it is classified in two main categories: primary dystonia, almost always idiopathic, sometimes due to a genetic defect, and secondary dystonia, second to another morbidity. DYT1 dystonia, an autosomal dominant movement disorder, is the best known hereditary form, but with a low penetrance [23-26]. Current views also separate generalized dystonia from focal dystonia. CD is the most common form of focal dystonia and affects 6-9 persons in every 100000. It prevails in females (male to female ratio 1.4:2.2) with a peak age of onset between 40 and 45 years [27-29]. Symptoms may include head jerks, but head deviation is always a distinctive feature. It corresponds to Spasmodic Torticollis, as defined by the former nomenclature. For many years it has been considered a neurological enigma [2].

DBS focuses on faulty basal ganglia circuits depolarizing and normalizing hyperactive structures; in fact, those patients who improve after DBS, return to a pre-operatory condition when stimulation is turned off. Unfortunately, no specific histochemical

change in the basal ganglia directly relates to CD and it is unknown how the supposed functional and metabolic disturbances alter the basal ganglia equilibrium [21,30].

Structural alterations on MRI studies have been found not only in the basal ganglia but also in the thalamus, motor cortex, premotor cortex, frontal, temporal and parietal cortices, visual system, cerebellum and brainstem. Several authors favor the idea that different brain regions constitute an interconnected pathologic circuit, but it is not known what disrupts normal functionality in this complex network [31]. DBS could block the electric oscillations in specific frequency bands expressing motor malfunction [32]. For the purposes of the following discussion, it is worth noting that affected muscles are innervated by the 11th nerve and by the first three anterior cervical roots. Both the nerve and the first cervical roots easily conflict with the vertebral artery or one of its rami. In fact, the vertebral artery enters the thecal sac in close proximity with the 1st cervical root and sometimes within its sleeve [33]. Because of this, traditional therapeutic attempts have focused on the 11th nerve with good results. Furthermore it has been repeatedly reported that the 11th nerve may be affected by a vascular conflict in the same way as the fifth nerve does [15-18]. Sudden ephaptic electrical discharges within trigeminal fibers cause trigeminal neuralgia, as ephaptic discharges within 11th nerve fibers may induce muscle spasms [34]. This similarity is relevant to explain at least some forms of CD.

Commonly, the first line treatment of CD relies on Botulinum Neurotoxin (BoNT). The effect of a single dose will last 3-4 months with a maximum effect after the third week. It has largely eliminated many of the long-term complications of torticollis, such as contractures and radiculopathy [35,36]. Unfortunately, despite overall good results, this treatment does not achieve a permanent and definite head alignment and does not always satisfy patients' requests. It may cause dysphagia and sometimes the immunoresistance develops, especially if treatments are frequent and booster doses are used [35]. Above all, most patients feel that they are not getting the same benefit they got earlier on with BoNT [37]. Biofeedback may be of help, as the patient learns to silence the hyperactive muscle while viewing his EMG on a display. Permanent good results have been reported in some cases [38,39]. Antipsychotic drugs have been widely used in the past primarily because torticollis has been regarded as a psychiatric disease. However psychiatric support is often required, because the need to cope with the dystonic deformity may bring personality changes.

DBS is considered by many the procedure of choice. It targets the Globus Pallidus Internus (GPi) uni or bilaterally to enforce an inhibitory activity by pallidum on the thalamus and, indirectly, on the hyperactive motor cortex [21]. Hung reported improvement for severity by 54.8%, for disability by 59.1%, and

for pain by 50.4% on the Toronto Western Spasmodic Torticollis Rating Scale (TWSTRS) [40,41]. The subthalamic nucleus has been considered as an alternative target for treatment [42]. DBS improves quality of life in up to 60% of patients (range 40-60%), according to a SF-36 survey [43]. Considering the TWSTRS, the disability improves by 60% and pain by 50% as observed in a follow-up period averaging 32 months [44]. A recent Canadian multicenter, double blind study of 10 patients demonstrated a 43% improvement of dystonia severity score and 59% improvement of TWSTRS total score compared with the pre-treatment values [45].

On DBS converges the enthusiasm of neurologists, mainly for what it promises: the control of the disease by interfering with the same neural mechanisms that produce it. It is to be said that this "interference" is nonspecific because there is no evidence of structural anomalies in the basal ganglia. Instead specific structural anomalies have been found in patients with Parkinson's disease [31].

It is possible that the CD, as a central nervous system disease evolves to a more generalized form. Searching pub med with a query "torticollis followed by generalized dystonia", six papers appear, only one of which refers to the evolution of CD ("Spread of primary dystonia in relation to initially affected region.") [46]. The authors studied 132 patients, selected "if primary focal dystonia was the only sign of neurological disease other than tremor; i.e. in all patients a single body part could be identified as affected at the onset". The authors state that, "after a mean follow up period of 7.5 years, dystonia remained focal in 73% of the cases". In 10 patients (7%) it progressed to a generalized form. Among those with CD, only 3 (5.9%) out of 51 developed generalized dystonia. Nothing is stated about pathogenesis, but interestingly the risk of the of spread of dystonia was lower in patients with positive family history. Friedman reports that in his series of 58 operated patients only one developed signs of generalized dystonia [47]. So, the argument that CD, as a form of a progressive disease worsens with time, has never been proven, but it may be different in genetic distonia [48].

Given the fact that stimulation of the basal ganglia resets and interrupts a pathological circuit, we do not know how and where the disease takes origin. Perhaps, DBS is not so specific as we like to believe and it is reasonable to pursue other treatments that still give good results. Denervating procedures as surgical options for CD are validated by historical evidence. The SCM is always involved in the genesis of torticollis. Surgeons have achieved acceptable results, carefully selecting patients whose denervation of the SCM could be enough to correct the abnormal posture. The earliest attempt was done by Bujalski by ligating the 11th nerve [3]. In following years other surgeons sectioned the 11th nerve or performed intradural rhizotomies to denervate the SCM and muscles compromised by CD [5,6]. This latter procedure has been refined by McKenzie and popularized by Nashold and later

by Friedman et al. who in 1993 reported on 58 patients with a success rate of 74% if torticollis had occurred less than five years prior to the procedure, and of 42% in other cases. They labeled as "success" cases that were subjectively rid of torticollis, were free of muscle spasms, suffered no or minimal pain, had normal or almost normal head position, and had no untoward side effects of surgery. Their method consisted of bilateral intradural denervation cutting 11th nerve rootlets and the anterior cervical roots, up to C3, directed to the SCM and, if warranted, to the trapezius [47].

Among torticollis experts, Bertrand and his followers selectively denervate the SCM aiming at the terminal rami of the 11th nerve [7]. It requires that every peripheral ramus directed to the SCM be cut. Others combine intradural with extradural approach, pursuing better results, but the procedure is more difficult [49,50]. Jho and Jannetta reported in 1995 on 20 patients in whom a vascular conflict caused torticollis, in analogy to facial spasm and conflict with other cranial nerves. They state that 13 (65%) were cured and 4 (20%) improved while others had minimal improvement or no changes [15]. Subsequently, others have confirmed that vascular impingement could be responsible for the spasmodic contraction of the neck muscles resulting in CD. Shima et al. operated on 7 patients out of 22 evaluated. Selection relied on the fact that torticollis of extrapyramidal origin would attenuate at rest, on the contrary for torticollis due to vascular conflict. By doing so, they sectioned "at C1 or C2 the branching root of the 11th nerve that had caused the tight cross contact" with the conflicting artery and fixed the nerve trunk to the dura mater. They did not use any EMG guidance. Improvement was obtained in 2 to 4 weeks with full or satisfactory relief in 5 patients, with improvement in 2 [18]. It is not known how many patients with CD carry a vascular conflict and even sophisticated diagnostic tools rarely prove such anomaly.

We believe many patients with CD carry a vascular conflict, in analogy to what happens with trigeminal neuralgia. The incidence of this is unknown. It may well be that CD represents two different entities, one originating by 11th nerve malfunction (which could be considered true "Torticollis") as opposed to another being true "dystonia". This view, which treat torticollis as a special entity due to peripheral nervous malfunction rather than basal ganglia dysfunction, is reinforced by the experience reported by Bertrand and followers on more than 2000 cases [51,52]. Inspired by these considerations our method consists of unilateral exposure to the 11th nerve on the side of the tightened SCM, as seen on clinical examination and verified through preoperative EMG studies. The MRI study may confirm the side of the tightened muscles, as it appears with larger diameter with respect to the controlateral. Deossiglucose studies may reveal an increased metabolic activity on this same SCM. Unfortunately, it is difficult to reveal a vascular conflict on the 11th nerve. A detailed study is not always possible due to the character of the disease, which worsens under stress or in uncomfortable positions. However, we always found a vascular

conflict. In one patient we were impressed because of the finding of the 11th nerve embedded within dural sleeves at the entrance of the vertebral artery in the thecal sac. CD resolved after freeing the 11th nerve. A similar finding has been reported in an anatomical study [53].

Our opinion is that the 11th nerve is hyperactive and drives a muscle spasm that causes the SCM tonic and phasic contraction with head posture. The motor anomaly, initially confined to the SCM could extend to contiguous muscular groups in a compensatory attempt driven by sensory feedback to offset the postural anomaly. This is to say that anomalous muscular contractions activate nervous pathways in a vicious cycle that finally alters nervous control of the axial musculature. It is a point difficult to prove and to which up until now there is no answer: can a peripheral malfunction drive a pathological change within the central nervous system? From what we know about the pain it could be the case. Neuropathic pain tends to radiate and extend beyond the area of anatomical alteration. It alters and recruits cellular electrical activity within the dorsal horn, thalamus, and pain circuitry. Similarly, malfunction of the 11th nerve may drive pathological changes in the basal ganglia-thalamo-cortical circuits. Our idea is that complex deformities follow head deviation due to the obliquity of the patient's line of sight. To keep a horizontal line of sight, the body is forced to adjust. Shoulder elevation with anterior displacement and lateral or sagittal shift of the head causes scoliosis. Spasms bring a need for continuous adjustment and bodily deformities progress [54]. Deformities of the trunk follow the progression of scoliosis. Based on this, the amnestic finding that head deviation started with SCM spasms is a key consideration when deciding which patients may benefit from surgery directed to the 11th nerve.

Conclusion

We propose a method that combines a denervating strategy with a microvascular decompression through a reduced approach. It may be a better strategy compared to classical denervating procedures or microvascular decompression alone. Compared to the peripheral denervating procedures, our strategy offers the advantage of exploring for vascular conflicts that are not evident by MRI. Patients must be selected according to the pattern of the evolving disease, clinical observation, electromyography and possibly according to metabolic studies with deoxyglucose which should demonstrate a preeminent involvement of one SCM. Our results show that if surgery focuses on the affected side, the outcome is good even when TWSTRS reaches high values. Since as for current classification "torticollis" corresponds to Cervical Dystonia with alteration in the basal ganglia, we wonder if a subgroup justified by vascular conflict on the 11th nerve could involve the basal ganglia and drive their dysfunction with passing time. It happens in chronic pain syndromes as repeatedly reported

[55]. If so, scholars could consider a different nomenclature distinguishing those cases due to 11th nerve dysfunction, especially if others will replicate our results.

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