Surgical treatment of non-responsive cervicogenic headache

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ABSTRACT

Objective.

102 patients suffering from long-lasting, very severe cervicogenic headache (CEH), non-responsive to physical or drug therapy, were surgically treated.

Methods.

Different diagnostic procedures and their significance for determining the advisability of surgery are summarised. The particular importance of the patient history and local anaesthesia together with the clinical examination is outlined. Different surgical approaches are described: ganglionectomy, ventral and dorsal decompressive operation.

Results.

CEH can be triggered by vascular or scar tissue compression of the C2 root and ganglion and irritation of other upper cervical nerve roots (C3,C4). Vascular compression is caused by: (a) the sinusoidal venous plexus, which surrounds the ganglion and nerve root like a cuff and may be dilated upon raised venous pressure; (b) further on by arterial loops throbbing against the ganglion; and (c) (rarely) by arteriovenous (AV) malformations. Nerve fibre degeneration is demonstrated morphologically by electron optical investigation. Afferences from ganglion C2 to the brain stem, as documented by experimental investigation on cats using the injection of HRP into the C2-ganglion, can explain the reference of pain from the neck to the fronto-ocular region and could at the same time elucidate the genesis of accompanying symptoms.

Degenerative diseases such as disc protrusion and retrospondylosis have been shown to be other trigger mechanisms evoking CEH, as is well known from facet joint arthrosis. Degenerative diseases usually cause dura compression with narrowing of the spinal canal and frequently, in addition, instability. Evocation of CEH could be explained by the irritation, by those degenerative diseases, of structures with pain-conducting nerve fibres (facet joint capsule, nerve root, longitudinal ligaments, spinal dura, disc).

About 80% of our surgically treated patients were relieved of pain or improved during a long period of follow up. The recurrence of degenerative alterations with new irritation from pain-conducting structures is thought to be responsible for the recurrence of headache. Further surgical approaches for the treatment of patients with the recurrence of pain are discussed.

Conclusion

Various surgical treatments are suggested to treat long-lasting severe CEH in patients not responsive to any physical or drug therapy.

Introduction

The decision for surgical treatment generally requires the failure of physical therapy to improve the patient's disease to an acceptable extent. All the patients whom we treated surgically suffered from intractable, non-responsive cervicogenic headache (CEH). The diagnosis of CEH was made in accordance with the revised definition of CEH, which was published by Sjastaad and co-workers in 1998 (12).

The most important prerequisite for surgical therapy of CEH is to find out the origin or - perhaps better called - the trigger mechanism of the patient's headache. For a long time the concept of headache with a cervical origin was not accepted, since the trigger mechanisms were unknown. Tumors or vascular diseases, especially from the vertebral artery, were held to be responsible for this headache. Both of these diseases are very rare, however, in comparison to the higher frequency of CEH.

Even today we have knowledge of only a few mechanisms which could trigger CEH. Relatively well established is the relevance of degenerative changes of the upper cervical facet joints, especially C2/C3 (1, 2) and also the relevance of irri-

tation or compression of the upper nerve roots and ganglions, especially the C2 root (4-10).
The fact that degenerative diseases of the cervical spine could contribute to the origin of cervicogenic headache was denied vehemently for a long time. For the surgical treatment of CEH to be undertaken, however, it is essential to search for and find the precipitating factors causing this headache. To find these trigger mechanisms, an extensive diagnostic procedure is essential. This procedure is described in the following essay.

Diagnostic procedures
Anamnesis, clinical examination, imaging procedures, and - very importantly - local anaesthesia, are the tools currently available to find these trigger mechanisms. The search is focused on cervical pain-provoking and pain-conducting structures. These structures are the nerve root, disc, dorsal and ventral ligament, facet joint capsule and, last but not least, the dura mater.

Anamnesis
Patients coming into the neurosurgical department are very selected. Most have been tormented by pain lasting for years without acceptable improvement. Upon asking the patients for the anamnesis, they report unilateral, rarely moderate but frequently severe headache, usually without sideshift but sometimes spreading across the midline. In some patients suffering from disc protrusion and retrospondylosis combined with spinal canal narrowing or stenosis, we find bilateral headaches as well.
The patient's headache starts in the neck and spreads over the temporal or parietal to the fronto-ocular region. The pain remains in the initial stage sometimes for hours, but usually for a few days. In a later stage in some cases the headache becomes a continuous pain. Either this pain is felt as a basic pain - continuing, often with considerable strengthening - or as an intolerably strong continuous pain. Sustained position or neck movement and shoulder-neck-overload can provoke the patient's headache. Related symptoms such as nausea (sometimes with vomiting), dizziness or ataxic abasia and blurred vision have been found in different combinations but without any differential diagnostic value. Patients with pathologic changes of the upper cervical nerve roots reported tearing, conjunctival injection or periocular ipsilateral oedema more frequently.
The anamnesis is the most important part of the examination and requires the most time to complete.

Clinical examinations
The neurologic as well as the neurophysiologic examinations are important to exclude the possible presence of space-occupying lesions of the brain, for example pituitary tumor, especially if the patient reports frontal headache not originating from the neck.
The neuro-orthopaedic examination indicates movement disorders and its segmental origin, if performed by an experienced clinician. Active and passive neck movement can precipitate attacks or pain aggravation, as can pressure on specific tenderpoints in the cranio-cervical region. These tenderpoints are the greater and minor occipital nerves (GON and MON), tendon insertions of the sternocleidomastoid muscle, and the dorsal rami at the transversal processes. These findings are not specific for the localisation of any trigger mechanism of pain, but do serve to underline the fact that structures of the neck are responsible for the patient's headache.

Imaging procedures
X-rays should be carried out first of all, in flexion and extension as well as at rest. With these functional x-rays, the extent of segmental mobility can be measured using the method described by Dvorak *et al*. (3). This investigation is very important to detect any one-segment hypomobilities in patients with multi-segmental degenerative changes such as ligamental calcification, osteochondrosis, retrospondylosis, facet arthrosis or reactive segmental muscular hypertension, and at the same time to discover any single segment hypermobility due to disc degeneration or facet joint instabilities.
CT findings are of less importance for the diagnosis of CEH in my opinion; they are helpful to demonstrate disc protrusion, retrospondylosis or uncarthrosis and for the exclusion of a primary or secondary vertebral tumor.
MRT is known to be useful for studying soft tissue structures. The soft cervical tissues are of special interest because of their capacity as pain-conducting nerve fibres. C fibres and A delta fibres can be found in the disc, the dorsal and ventral ligament, the facet joint capsule, spinal nerve roots and - in my opinion very important - in the cervical dura mater. MRT shows disc protrusion, dorsal ligament thickening, dura compression with spinal fluid stenosis and, additionally, nerve root compression. Specialised radiologists can also find alterations of the facet joint capsule. These findings necessitate our next investigation.
Local anaesthesia, either peridural or on the symptomatic side - with all due consideration for its potential problems - was found to be helpful in elucidating the context between each of these innervated structures and the origin of pain. The differentiated local anaesthesia of pain-mediating structures and the relief of headache, frequently including the accompanying symptoms, explains the location and trigger mechanism of this headache. If these blockades are carried out in an exact manner, then local anaesthesia can confirm the diagnosis and the indication for surgery. As far as possible the blockade should be combined with pain-stimulating procedures, both without and after performing local anaesthesia. In this way the context between specific pain-conducting structures and CEH can be certified.

Methods
Patients
Since 1979 102 patients suffering from severe CEH were treated by surgery. Many of these patients were at risk of suicide because of the severity of their pain. Altogether 65 female and 37 male patients were treated. C2 ganglionectomy was performed on 38, ventral decompression on 56, and dorsal decompression on 8 patients. Ventral or dorsal decompression was performed on 42 and 7 females, respectively; thus female patients had a clear preponderance over males. Among the patients undergoing ganglionectomy we found an insignificant preponderance of 22 males over 16 females.

C2 ganglionectomy
C2 ganglionectomy was performed in 38 patients suffering from the effects of ganglion or nerve root irritation or compression. All had a typical history and neuro-orthopaedic symptomatology. The patients suffered from one-sided hemicranial attacks without sideshift, sometimes for hours but usually for days. After years the attacks changed in some cases to become continuous pain. Pain attacks were accompanied by a single or different combinations of concomitant symptoms, including conjunctival injection, tearing, nasal hypersecretion, and dizziness (or perhaps more precisely, ataxic abasia, visual disturbances, or - very rarely - single drop attacks with unconsciousness). The clinical neuro-orthopaedic examination showed the above mentioned symptoms. The indication for surgical treatment was confirmed by the responsivity to local anaesthesia of the C2 root, pre-conditioned non-responsivity to drugs, and the lack of acceptable improvement following physical therapy.
Surgical findings. During the operation compression of the nerve root and ganglion was found, frequently caused by the surrounding venous sinusoidal plexus. Arterial loops throbbing against the ganglion were found only rarely. An arterio-venous malformation was seen in 2 cases. Scar tissue or a thickening of the atlanto-epistrophic inter-arcual ligament was also found in some patients.
Electron optical findings. Electron optical investigation was performed on the ganglions and nerve fibres. Signs of nerve fibre degeneration, nerve fibre regeneration, and collagen fibre production were found. These pathological changes were considered to be the result of chronically repeated local injury of the C2 ganglion and root by the above mentioned vascular compressions.
Afferences to the brain stem. Searching for an explanation of the accompanying symptoms, experimental investigations were carried out on cats to study afferences from the C2 ganglion to the brain stem. Wheat germ agglutinated horse radish peroxidase was injected into the C2 ganglion and, 48 hours after injection and after fixation of the animal by perfusion, the brain stem was excised. Light microscopic investigations showed afferences to the dorsal horn of the spinal cord at the C2 level, descending fibres at the C3 level, and ascending fibres to the cuneat nucleus, as expected. Interestingly, we found afferences to the caudal spinal trigeminal nucleus, the dorsal motor nucleus of vagus, the inferior vestibular nucleus, and to the nucleus X. The relevance of the nucleus X is not yet clear, but connections from nucleus X to the III and IV cranial nerves are known. That could explain the accompanying symptoms. It may also be the explanation for the referred pain from the neck to the fronto-orbital region.

Ventral decompression
Ventral decompression with one- or two-segment fusioning was performed in patients suffering from severe headache over a long period of time. It was carried out if imaging procedures demonstrated less than two segments of ventral spinal stenosis caused by disc protrusion or bulging combined with retrospondylosis. The patient's headache therefore stemmed from the neck, beginning there and spreading over the temporal and/or parietal region to the fronto-ocular region. Neurological examinations were not of additional value for the diagnosis of headache. About 70% of our patients also showed radicular symptoms. Because of the predominantly medial site of the protrusion, local anaesthesia was performed peridurally (PDA). In determining the indication for surgery, PDA was the most important diagnostic procedure next to anamnesis and the imaging findings.

Dorsal decompressive laminectomy and laminoplasty
Dorsal decompression was usually performed from the lamina C2 to C6 or C7 by laminectomy and laminoplasty (11). Each sawed lamina was moved dorsally about 3 or 4 mm and than fixed again with screws. This surgical procedure was indicated in patients with more than two-segment disc protrusion and retrospondylosis. Experience in the treatment of myelopathy led to the development of this surgical approach. The clinical symptomatology and the imaging findings were identical with the above-mentioned patients undergoing ventral decompression. None of these patients had experienced any benefit from physical or drug therapy, but were totally relieved of pain by PDA for as long as the effect of the local anaesthesia remained.

Results
Relief of pain
One hundred and two patients severely tormented by CEH pain were operated on in different ways. Summarising and considering all these operations, a 100% relief of pain was achieved in about 80% of our patients. An improvement of between 60% and 80% was seen in about 15%. Pain remained unaffected in about 6%.
Looking separately at the operative procedures we find that among 38 patients who underwent C2 ganglionectomy, 26 experienced a complete relief of their pain and 6 were improved. The complete relief of pain lasted for a mean period of 44 months, and the improvement for a mean period of 25 months. Six patients showed no response to ganglionectomy. After ventral decompression and fusioning in 56 patients, the pain was totally relieved in 48, and 8 were improved by more than 50% for a mean period of 14 months. Thus, all patients were treated successfully within the follow-up period. Dorsal decompressive laminectomy and laminoplasty were performed on only 8 patients. For a mean period of 5 months 7 patients were relieved of their pain, while in 1 the pain was lessened by more than 50% for 13 months.

Recurrence of pain
After C2 ganglionectomy (n = 38) a recurrence of pain (50% or less) was seen in 8 patients. Only 2 patients experienced a total recurrence of pain after a mean period of 45 months. Six of the 8 patients who underwent dorsal decompression suffered recurrences of pain: in 2 the pain was ≤ 50% and in 4 it was > 50% after a mean period of 5 months.

Conclusion
C2 ganglionectomy
Focusing our interest on those 8 patients with a recurrence of pain of 50% or less, local anaesthesia of the upper nerve roots C3 or C4 showed good results after the

recurrence of headache. The anaesthetic block achieved relief or improvement from pain as long as the anaesthetic effect remained. Because of the presence of numerous motor fibres we refrained from ganglionectomy of these roots. After radiofrequency neurotomy, patients showed improvement or total relief from pain. This highlights the fact that not only C2, but also the upper cervical nerve roots C3 and C4 play a role in the development of CEH. Electron optical investigation convincingly showed pathological nerve fibre alterations as a result of chronically repeated injury caused by irritation and compression of the described vessels. The afferences from the C2 root to the brain stem explain the reference of pain from the neck to the first branch of the trigeminal nerve, as well as the origin of the accompanying symptoms. The post-operative relief of pain over a long period confirms the important role of the upper nerve roots in evoking headache.

Ventral or dorsal decompressive operation

About 80% of the patients suffering from very severe pain obtained significant and long-lasting benefit in terms of their personal and social life after the decompressive operation. The surgical procedure certainly does not cure the degenerative disease. Therefore, it is not surprising that the development of new degenerative changes in a further segment could lead to new pain. New degenerative changes were often seen to develop next to the fused segment. After a further decompressive operation on patients with such new pain, their headache was improved or relieved again. Months or years after the operation many of our patients were asked whether they would agree to undergo the operation again, knowing their post-operative results. The spontaneous approval of the vast majority of patients encourages us to continue to use this surgical procedure in cases of severe CEH.

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