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Diagnosing cervicogenic headache

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Abstract The notion that disorders of the cervical spine can cause headache is more than a century old, yet there is still a great deal of debate about cervicogenic headache (CEH) in terms of its underlying mechanisms, its signs and symptoms, and the most appropriate treatments for it. CEH is typically a unilateral headache that can be provoked by neck movement, awkward head positions or pressure on tender points in the neck. The headaches can last hours or days, and the pain is usually described as either dull or piercing. Convergence of the upper cervical roots on the nucleus caudalis of the trigeminal tract is the most commonly accepted

neurophysiological explanation for CEH. In most cases, CEH is caused by pathology in the upper aspect of the cervical spine, but the type and exact location of the pathology varies substantially among individual cases. Anaesthetic blocks may be necessary to confirm the diagnosis of CEH, showing that the source of pain is in the neck. Differential diagnosis is sometimes a challenge because CEH can be mistaken for other forms of unilateral headache, especially unilateral migraine without aura. Neuroimaging and kinematic analysis of neck motion may aid in diagnosing difficult CEH.

Keywords Cervicogenic headache • Neck pain • Headache

Clinical picture

Since the first cases of cervicogenic headache (CEH) were identified [1], considerable progress has been made. Particularly in the last decade, there have been advances in therapeutic approach and in defining the clinical picture and diagnostic criteria. As repeatedly stated, CEH is a syndrome, not a disease or an entity *sui generis*. It constitutes a "final common pathway" for pain stemming from several neck disorders. These may involve such structures

as nerves, nerve root ganglia, uncovertebral joints, intervertebral disks, facet joints, ligaments, muscles and so on [2, 3]. Pain may accordingly originate at different levels, including the lower part of the cervical spine [4]. CEH comprises all headaches stemming from the neck with the possible exception of specific headache entities (e.g., a subgroup of chronic paroxysmal hemicrania (CPH) with mechanical precipitation of attacks) [3].

CEH has been defined, in principle, as a unilateral headache without sideshift. In the upgrading of the CEH diagnostic criteria [5], the strict unilaterality criterion has been softened. In clinical practice, patients with bilateral headache may be acceptable (like "the unilaterality on two sides" in tic douloureux) [6, 7]. Because CEH is a syndrome, the pathologic process can, probably not so infrequently, be reproduced on the contralateral side. In these cases, a positive response to appropriate anaesthetic blockades might be essential also in clinical practice (not only in scientific diagnostic work-up), mainly in order to exclude tension-type headache (TTH). Even in the more regular unilateral case, pain may eventually spread to the opposite side when headache becomes severe, while remaining stronger on the original side [5]. The typical unilaterality is probably clearest at attack/exacerbation onset. In CEH, therefore, headache may be strictly unilateral in the most typical and diagnostic case, or it may have a unilateral preponderance; as far as we are concerned, it will not occur solely on the side opposite to the usual one [8].

Other, equally important, diagnostic features are the symptoms and signs of neck involvement. Such signs are mechanical precipitation of attacks (both iatrogenically and subjectively induced), reduced range of motion in the neck — in one or more directions, diffuse ipsilateral neck/shoulder/arm pain of non-radicular nature or, occasionally, arm pain of radicular nature (Table 1). Iatrogenically induced pain similar to the spontaneous one may be elicited by external pressure over tendon insertions in the occipital area. Pressure along the course of the major occipital nerve, over the groove immediately behind the mastoid process, and over the upper part of the sternocleido-mastoid muscle on the symptomatic side

may also provoke similar pain. Intrinsic precipitation mechanisms may be activated by neck movements and/or sustained, awkward head positioning during sleep or during wakefulness (such as when washing the ceiling, speaking to one's neighbour at a table during a party, and so forth). Ipsilateral shoulder/arm symptoms may be even more frequent than they seemed to be initially [9]. Not infrequently patients are encountered with marked, more or less constant arm pain of a non-radicular nature [8]. In these cases, the underlying pathology possibly resides in the lower part of the cervical spine (C5 and so on). However, these phenomena are not infrequently of low intensity, and may be more like a discomfort than a pain. Such phenomena may in the occasional case have their own temporal pattern, more or less independent of the headache attacks. The side-locked unilaterality of the headache combined with the ipsilaterality of the arm pain provides rather compelling evidence that headache on such occasions stems from neck structures, but not necessarily only from bony structures.

The duration of attacks/exacerbations varies widely (from a few hours to a few weeks), with a strong tendency toward chronicity; CEH is not infrequently episodic in the initial phase, becoming chronic-fluctuating later on. The pain of attack starts in the neck, eventually spreading to the oculofrontotemporal area, where, during the acme, it may be as strong as or even stronger than in the occipital region [2, 5]. The duration of pain episodes is most frequently longer than in common migraine; the pain intensity is moderate, non-excruciating, unlike cluster headache and usually of a non-throbbing nature.

 Table 1 Cervicogenic headache: CEHISG diagnostic criteria [5]

Major criteria

Ia. Precipitation of head pain, similar to the usually occurring one: Ia1) by neck movement and/or sustained, awkward head positioning, and/or: Ia2) by external pressure over the upper cervical or occipital region on the symptomatic side. Ib. Restriction of the range of motion (ROM) in the neck. Ic. Ipsilateral neck, shoulder or arm pain of a rather vague, non-radicular nature, or – occasionally – arm pain of a radicular nature. II. Confirmatory evidence by diagnostic anaesthetic blockades. III. Unilaterality of the head pain, without sideshift. Head pain characteristics IV. Moderate-severe, non-throbbing pain, usually starting in the neck. Episodes of varying duration, or: fluctuating, continuous pain. Other characteristics of some importance V. Only marginal effect or lack of effect of indomethacin. Only marginal effect or lack of effect of ergotamine and sumatriptan. Female sex. Not infrequent occurrence of head or indirect neck trauma by history, usually of more than only medium severity. Other features of lesser importance VI. Various attack-related phenomena, only occasionally present, and/or moderately expressed when present: a) nausea, b) phono- and photophobia, c) dizziness, d) ipsilateral "blurred vision", e) difficulties swallowing, f) ipsilateral oedema, mostly in the periocular area.

I. Symptoms and signs of neck involvement*

^{*}It is obligatory that one or more of the phenomena Ia-Ic are present

Autonomic symptoms and signs, like photo- and phonophobia, nausea, vomiting and ipsilateral periocular oedema, are infrequent – and mild if present – and some of them, like vomiting, are clearly less marked than in common migraine [2, 3, 5, 8–10]. In a study by Vingen and Stovner [11], light- and sound-induced discomfort and pain thresholds have been measured in patients with TTH, CEH and in headache-free controls. It is striking that patients with CEH showed a greater photophobia on the symptomatic than on the non-symptomatic side, whereas no such differences were found in TTH and unilateral headaches [11].

Difficulty swallowing is another, rarely occurring, associated phenomenon [1, 4]. There have also been cases with features consistent with a CEH picture, but with additional dizziness and even with vertebral drop-attacks; such patients may benefit from surgical interventions, such as an anterolateral approach toward the cervical spine, *ad modum* Jung. These patients may constitute another clinical subgroup, namely the "vertebral artery type" [8].

Diagnostic criteria

In the revised diagnostic criteria [5] (Table 1), the importance of symptoms and signs of neck involvement has been further stressed. Mechanically precipitated attacks – or pain similar to that of an attack – subjectively and/or iatrogenically induced, is an obligatory requirement for a certain/definite diagnosis, as is the positive anaesthetic blockade effect. Unilaterality without sideshift is highly desirable in scientific works. The lack of Ia criterion will clearly reduce the validity of the diagnosis. It has been proposed that the presence of Ib and Ic, II), and III) criteria, as in the previous version, may be consistent with a "provisional"/tentative diagnosis [5]. In the revised criteria [5], among the "Other characteristics of some importance", the lack of a complete response to indomethacin, sumatriptan and ergotamine has also been introduced.

Although CEH is not, in principle, a post-traumatic headache, a history of neck/head trauma should still be considered to be of potentially pathogenetic importance, especially if it is of more than "only medium severity" and has a putative whiplash mechanism [3, 5].

A history of a long-lasting, strictly unilateral headache is suggestive of CEH, in particular if in a female subject. The temporal, that is the "non-clustering", but chronicfluctuating pattern, and the severity and the non-throbbing nature of the pain (usually moderate and non-excruciating) distinguish CEH from other unilateral headaches, such as cluster headache and CPH. Hemicrania continua (HC) and migraine without aura may represent differential diagnostic problems. An appropriate anamnesis and accurate neurological examination, showing a reduced ROM [12] and precipitation mechanisms, are fundamental elements in distinguishing this headache from others. The combination of pain first felt in the neck and then spreading unilaterally to the frontal area on the same side fortifies the suspicion that one may be faced with a case of CEH. The site and radiation of pain, the temporal pattern and the mechanical precipitation of attacks, both iatrogenically and subjectively, are important aspects of the clinical picture and may help in distinguishing between CEH on the one hand and migraine and TTH on the other [13, 14]. In patients with bilateral pain, but still with a preponderance on the usual side, anaesthetic blockades become mandatory even in clinical practice. In order to single out the correct level of affection, the blockades should be directed to the nerve or nerves where the pain most likely originates/is elicited, on the side of prevailing pain [15].

The IHS diagnostic criteria

In the new IHS classification the criteria for headache associated with neck disorders has been largely revisited (Table 2) [16]. The headache is termed for the first time: "cervicogenic headache" and not "cervical headache".

Table 2 Cervicogenic headache: IHS diagnostic criteria [16]

Diagnostic criteria

- A. Pain, referred from a source in the neck and perceived in one or more regions of the head and/or face, fulfilling criteria C and D
- B. Clinical, laboratory and/or imaging evidence of a disorder or lesion within the cervical spine or soft tissues of the neck known to be, or generally accepted as, a valid cause of headache
- C. Evidence that the pain can be attributed to the neck disorder or lesion based on at least 1 of the following:
 - 1. demonstration of clinical signs that implicate a source of pain in the neck
 - 2. abolition of headache following diagnostic blockade of a cervical structure or its nerve supply using placebo or other adequate controls
- D. Pain resolves within 3 months after successful treatment of the causative disorder or lesion

However, already under letter "A", it is stated that the pain can be in "...one or more regions of the head and/or face...". In other words, there may be only facial pain and not a headache. Unfortunately, the causes of this headache are only vaguely known, and the finer mechanism largely unidentified. Instead of - under letter "B" - "...known to be or generally accepted as...", this item should probably have been left alone. Fantasy and the creativity of devoted future researchers are relied upon to uncover the causes and not limit them to what is "known". Moreover, it is impossible to decide what is "known/accepted". Another shortcoming of the new classification is under letter "B", where it is stated: "Clinical, laboratory and/or imaging evidence...". As written here, it can be interpreted as meaning: imaging evidence would suffice, giving no importance to any headache. Even under letter "C" 1, which suffices for the fulfilment of criterion "C", it is stated: "...pain in the neck" while under letter "D": "pain" is unspecified. Seemingly, a headache does not have to be present.

A minor importance has been deserved the property of CEH to be precipitated. Only in the notes and not in the IHS criteria are the important ipsilateral shoulder/arm symptoms mentioned, which aid in distinguishing CEH from a central affection. These are only some examples of the many shortcomings of these criteria. Unfortunately, these criteria – as the previous ones – may be somewhat unsuited for clinical headache work, and even less so for epidemiological headache work. In retrospect, the 1983 description of CEH may have constituted a ride, a new escalating interest in headache stemming from the neck.

A further refinement of the current diagnostic IHS criteria might make it possible to avoid the existing, partial overlap of CEH and migraine/TTH. We have the feeling that extensive use should be made of the greater occipital nerve – and other blockades – in the routine work-up of CEH, both non-classifiable cases and the mixed forms, in order to improve the efficiency of the current diagnostic system.

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