
Symptomatic cervicogenic headache

R. Delfini¹, M. Salvati², E. Passacantilli¹, E. Pacciani³

¹Chair of Neurotraumatology, Dept. of Neurological Sciences; ²Department of Neurosurgery, Inn Neuromed, Pozzilli (Is), University of Rome "La Sapienza"; ³Ospedale Pediatrico Bambin Gesù, Presidio Di Palidoro (Roma).

Please address correspondence and reprint requests to: Maurizio Salvati, MD, Neurotraumatology, Policlinico Umberto I, Viale del Policlinico, 00100 Rome (Italy).
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ABSTRACT

Cervicogenic headache is a little-known clinical condition whose true importance has only recently been recognized. A number of causes may lie at the basis of the onset of headache (symptomatic cervicogenic headache). However, despite exhaustive attempts, sometimes it is not possible to identify a clear cause responsible for the onset of the syndrome (primitive cervicogenic headache). The genesis of symptomatic cervicogenic headaches sometimes may be easy to identify as a result of a close, pre-existing, cause-effect relationship (i.e. trauma). On other occasions it may be much more laborious to pinpoint the pathology responsible for headache (some cranio-cervical anomalies, etc.). Clinically, it is necessary to perform a thorough preliminary clinical and anamnestic evaluation which can orient subsequent investigations to achieve a diagnosis in the least time possible with the minimum discomfort to the patient and his relatives, not to mention lower costs for society.

Introduction

Headache is a complex syndrome which may originate from a considerable number of pathologies with a very variable genesis (1-12). In the light of this, it is not surprising that not infrequently disorders arising from the cervical spine may also play a role in headache. Pioneer studies in this context were performed by Barré who, as early as 1926 (13), noted this association. When it is possible to identify a lesion at the origin of headache, the term used is symptomatic or secondary cervicogenic headache. Other cases are classified as idiopathic or primary headache.

This study deals with symptomatic or secondary headache (SCH). Obviously, for a headache to be classified as symptomatic or, more precisely, idiopathic, a precise, scrupulous diagnostic protocol must be carried out. In fact, as mentioned at the beginning, at the basis of headache there may be a myriad of pathologies with various topographic locali-

zations and different prognostic significance. The first fundamental step in the study of such a patient is a detailed anamnestic evaluation followed by an equally detailed clinical examination, taking into consideration the patient's psychological status. It is impressive how the use of such investigations, which are simple to perform, makes it possible to save time and diagnostic means and, consequently, money as well. Above all, they can orient the clinician either towards the pathology likely to be responsible for headache or towards an "idiopathic" origin.

In an era where technology is a constant feature of daily life, however, "reason" must prevail over technology at all costs. Subsequently, according to the clinical data collected, the diagnostic investigations deemed most suitable should be performed. The principal diagnostic tool remains that of the radiological examination, both traditional (plain X-ray films: static and dynamic studies, plain tomography) and advanced (multiplanar plus 3-D CT-scan, MRI and MRI-angiography, scintigraphy) (14). However, the importance of neurophysiological tests (SSEP, SMEP, EMG) should not be underestimated (14). In the great majority of cases these studies can give a nosographic definition of the probable causes of headache or, on the other hand, its idiopathic nature. In Table I a brief classification of the diseases mainly responsible for the onset of so-called cervicogenic headache.

The structures involved in the genesis of SCH are numerous and can be grouped into (1, 2, 4):

- components of the vertebral elements (especially the first vertebral segments and cranio-spinal junction, in particular the apophyseal joints, synovial joints, annulus fibrosus of the intervertebral disks, ligaments of the spine, periosteum of the vertebral bodies);
- cervical muscles and their bone attachments;
- cervical nerve roots;
- vertebral arteries.

Table I. Symptomatic cervicogenic headache. Relevant cervical spine pathologies causing cephalaea.

Traumatic pathology
- Fractures
- Dislocations
- Instability
Degenerative pathology
- Disk herniation
- Cervical spondylosis
Congenital anomalies
- Cranio-spinal junction anomalies (Arnold-Chiari malformations, Klippel-Feil syndrome, atlanto-axial dislocation, occipitalization of atlas, basilar impression)
- Syringomyelia
Spondylitis
Rheumatoid arthritis
Neoplastic diseases
- Vertebral and extradural tumors (Metastases, primitive osseous tumors, lymphomas, etc.)
- Intradural-extramedullary tumors (Neurinomas, meningiomas, etc.)
- Intramedullary tumors (Astrocytomas, ependymomas, malignant gliomas, hemangioblastomas, etc.)

A number of theories have been proposed to explain why a lesion in the cervical spine region may cause a cephalic localization of pain (5). The nerve roots, the sympathetic cervical chain and the trigeminal nucleus seem to play a fundamental role in the spread and cranial localization of pain. However, an appraisal of the various series shows a significantly higher frequency of cephalic pain in pathologies involving the first cervical segments rather than the lower ones (1, 2).

We will now deal with the salient features of the main pathologies in more detail.

Cervical spine tumors

Tumors of the cervical spine region include a considerable number of tumors with a diverse histopathogenesis (1, 2, 8, 14). Pain as an early symptom is frequent (cephalea being one of the principal manifestations) and, unfortunately, the absolute non-specificity of this symptom means that at this stage it tends to be underestimated. On other occasions symptoms such as endocranial hypertension or the subarachnoid hemorrhage syndrome, usually indicative of an intracranial pathology, tend to be disorientating. In fact, spinal tumors such as neurinomas, meningiomas and ependymomas may sometimes give rise to symptoms

mimicking endocranial hypertension. Similarly, at the basis of a subarachnoid hemorrhage syndrome there may be an expansive lesion such as hemangioblastoma, neurinoma, ependymoma, or artero-venous malformation.

On the basis of clinical anamnestic indications, diagnostic investigations consist not only of traditional radiographic techniques (in particular for bony lesions), but principally MRI, MRI-angiography and multiplanar plus CT-scan. Nowadays, imaging techniques such as traditional spinal angiography play a very limited role and are only performed for specific pathologies such as spinal angiomas, often for therapeutic purposes (embolization).

Treatment may very radically depend on the histotype. In each case it depends on a common element which, when possible, consists of surgical treatment; depending on its clinical and biological behavior, this may be associated with radio- and chemotherapy.

In these lesions, in particular, early diagnosis is important. In fact, while it is true that some histotypes are not definitely influenced by the therapeutic strategy (above all gliomas and metastases), there can be no doubt that early diagnosis in such cases makes it possible to initiate treatment while the patient's clinical condition is more favorable, with a

better KPS and a lower post- and peri-operative morbidity. Overall, this means a significant improvement in the quality of life and, not infrequently, the length of life. In other histotypes, such as primary malignant bone tumors (osteosarcomas, in particular), an early combined approach may signify a real life-saving strategy.

Rheumatoid arthritis

This multi-district pathology accounts for one of the most important and complex chapters of modern medical treatment (1, 14). Difficulties in controlling the disease coupled with the elevated toxicity of the drugs currently available make the long-term management of patients with rheumatoid arthritis quite difficult. This disease is localized at the level of the upper cervical tract in a high percentage of cases (between 44 and 88%, according to the various series). The cervical localization leads to malalignment and instability of the vertebral elements ranging from the destruction of bone and supporting ligaments by synovial proliferation, to brain stem compression. Atlanto-axial subluxation is a particularly important aspect. Correction of the deformity and instability is frequently carried out surgically and must be performed early to avoid, or at least delay, the onset of an incurable invalidity with enormous costs to society.

Spondylitis

Spondylitis can be divided into vertebral osteomyelitis and discitis (14), and further divided into spontaneous and post-surgical (14). Frequently, the cause is identified as an infection of the urinary tract, respiratory system or soft tissues, an odontoiatric disorder (oral flora) or a previous spinal operation. However, in as many as 37% of cases the source cannot be identified. Not infrequently, the presence of diabetes mellitus, hemodialysis, advanced age or intravenous drug abuse are found to be at the basis. *Staphylococcus (aureus, albus, epidermis)*, *proteus* or *E. coli*, as well as *Mycobacterium tuberculosis*, are the germs responsible for infection. Conservative medical treatment cures 75% to 90% of cases. Surgery is necessary only occasionally.

Cranio-spinal junction anomalies

The cervicogenic headache syndrome affects 26% of patients with bony cranio-spinal anomalies (1). The most frequent pathologies identified in clinical practice are the Arnold-Chiari malformations. In this case, too, early identification resolves the problem and, above-all, avoids definite sequelae or may even be life-saving (14).

Degenerative pathologies

These consist of diseases (cervical disk herniation: C6-C7 and C5-C6 in particular; cervical spondylosis: C3-T1, especially C5-C6) with a high social impact, both in terms of their entity and the functional damage they may cause when they are not identified at an early stage (1, 2, 14). The CHS occurs in a significant percentage of cases. In fact, between 25% and 40% of patients with a degenerative spinal pathology develop cervicogenic headache. The prognosis is excellent if the timing and therapeutic techniques are correct

Traumatic pathology

Traumatic pathologies probably represent the most important category of cervicogenic headache syndrome due to their very high incidence and considerable social impact. Furthermore, they are among the few pathologies which can be avoided in the great majority of cases (14). Paradoxically, they represent an iatrogenous event, if "iatrogenous" is taken to mean the effect of irregular social behaviour. To summarize, we can divide cervical spine traumas into fracture, dislocation and those causing early or late instability, which can be variously combined. The mechanism responsible for the trauma can be either a hyperextension fracture-dislocation or a hyperflexion fracture-dislocation.

Depending upon the spinal segment involved, and taking into account the structural characteristics of each region, traumas can be classified as those involving segments C0 to C2 and those involving segments C3 to C7. In Table II, these various groups of lesions are described in detail. Their evolution depends not only on whether correct first-aid and subsequent treatment is introduced sufficiently early, but also on the extent and

Table II. Symptomatic cervicogenic headache. Traumatic pathology.

Subaxial traumatic pathology (C0 - C2)
- Hyperextension fracture-dislocations
Posterior fracture-dislocation of the dens
Traumatic spondylolisthesis of the axis (hangman's fracture)
Hyperextension sprain (momentary dislocation) with fracture
Hyperextension fracture-dislocation with fracture articular pillar
Hyperextension fracture-dislocation with comminution of the vertebral arch
- Hyperflexion fracture-dislocations
Anterior fracture dislocation of the dens
Hyperflexion sprain (rare, occur a rupture of posterior ligaments)
Locked articular facets with fracture
Teardrop fracture-dislocation
C3-C7 Traumatic pathology
Subluxation and locked facets
Unilateral locked facets
Bilateral locked facets
Teardrop fractures
Hyperflexion sprain
Clay shoveler's fracture
Sciwora

reversibility of spinal cord damage. For this purpose, MRI is essential because, when associated with the main clinical elements, it is able to give a fairly precise prediction of prognosis, both *quoad vitam* and *quoad valetudinem*.

Stabilization of the fracture, either by conservative or surgical management, is the cardinal feature of treatment, accompanied by intensive neuromotor rehabilitation. In order for the patient to return to society at least able "to look after himself", it is essential for treatment to be prompt and adequate. Although it may seem of little importance, there is an enormous difference between a patient able to move about in a motorized wheelchair and a patient unable to do even that for a number of reasons. In economical terms, this means a considerable saving even over a period of just one year.

The efficacy of early treatment with corticosteroids (such as methylprednisolone) is well established, patients obtaining a far better recovery than non-treated patients, or those treated with other steroids. The protocols used are periodically updated in accordance with the various NASCIS studies and are, in any case, easy to follow even in outlying emergency services.

Whiplash injury

Whiplash injury consists of a complex

constellation of symptoms (i.e., neck pain and stiffness, pain radiating to the occiput or fronto-temporal regions) which appear immediately or just a few hours after minor cervical trauma (1, 2, 6, 14). We retrospectively reviewed a group of 162 patients who suffered trauma between January and August 1975 and who were followed up for a period of 23 years. This series was culled from a larger group consisting of 588 patients. Anamnesis on admission was carefully evaluated to identify the onset of whiplash injury and the evaluation of pain (including intensity, limitation of social life, duration).

The average age of the patients was about 35 years, with a prevalence of males (M:F ratio 1.4: 1). Headache and vertigo were present in 22%. In 28% more than 5 diagnostic investigations were performed. Symptoms required medical treatment in 28% of the patients, physiotherapy in 27%. There was a temporary limitation of working activities in 8% of cases. One interesting observation regards the duration of symptoms (including headache) which persisted at 23 years of follow-up in 5% of patients and necessitated medical and physiotherapy. This incidence is far higher than had reported in the literature and warrants further studies on a larger patient population.

Conclusions

Every effort must be made to exclude the presence of cranio-cerebral and cervical pathologies before making a definite diagnosis of primary cervicogenic headache. Some cases require more sophisticated diagnostic investigations before a certain diagnosis can be made. The cost of headache treatment accounts for a substantial portion of the total health expenditures of a society.

Similarly to back and neck pain, cervicogenic headache like-syndromes are a leading cause of morbidity and disability, as well as an important factor in insurance litigation (i.e., whiplash injury and traumatic diseases).

References

1. EDMEDS J: The cervical spine and headache. *Neurology* 1988; 38: 1874-8.
2. EDMEDS J: Cephalées d'origine cervicale. *Rev Prat* 1990: 399-402.
3. PEARCE JMS: Cervicogenic headache: Personal view. *Cephalalgia* 1995; 15: 463-9.
4. BERGER M, GERSTENBRAND F: Cervicogenic headache. In *Handbook of Clinical Neurology*. New York, Elsevier Science Publishers BV, 1986: 405-12.
5. NILSSON N: The prevalence of cervicogenic headache in a random population sample of 20-59 years old. *Spine* 1995; 20: 1884-8.
6. LEONE M, D'AMICO, GRAZZI L, ATTANASIO A, BUSSONE G: Cervicogenic headache: A critical review of the current diagnostic criteria. *Pain* 1988; 78: 1-5.
7. JANSEN J, BARDOSI A, HILDEBRANDT J, LUCKE A: Cervicogenic, hemicranial attacks associated with vascular irritation or compression of the cervical nerve root C2. Clinical manifestations and morphological findings. *Pain* 1989; 39: 203-212.
8. DI LORENZO N, DELFINI R, CIAPPETTA P, CANTORE GP, FORTUNA A: Primary tumors of the cervical spine: Surgical experience with 38 cases. *Surg Neurol* 1992; 38: 12-8.
9. PIKUS HJ, PHILLIPS JM: Outcome of surgical decompression of the second cervical root for cervicogenic headache. *Neurosurgery* 1996; 39: 63-71.
10. IANSEK R, HEYWOOD J, KARNAGHAN J, BALLA JF: Cervical spondylosis and headaches. *Clin Exp Neurol* 1987; 23: 175-8.
11. MICHLER RP, BOVIM G, SJAASTAD O: Disorders in the lower cervical spine. A cause of unilateral headache? Case report. *Headache* 1991; 31: 550-1.
12. VINCENT M, LUNA RA: Cervicogenic headache. Josey's cases revisited. *Arq Neuropsiquiatr* 1997; 55: 841-8.