P1: KWW/KKL P2: KWW/HCN QC: KWW/FLX T1: KWW
GRBT050-121 Olesen- 2057G GRBT050-Olesen-v6.cls August 18, 2005 23:22

Chapter 121

Disorder of the Skull and Cervical Spine

Hartmut Göbel and John G. Edmeads

HEADACHE ATTRIBUTED TO DISORDER OF CRANIAL BONE

Internation Headache Society (IHS) code and diagnosis: 11.1 Headache attributed to disorder of cranial bone

World Health Organization (WHO) code and diagnosis:

G44.84 Headache or facial pain associated with disorders of cranium, cranial and facial structures, cranial nerves, neck, and spine

G44.840 Headache associated with disorders of the cranial bone (M80-M89.8)

Short description: Pain in the head or face caused by a lesion within the cranial bone

EPIDEMIOLOGY

Most disorders of the skull (e.g., congenital abnormalities, fractures, tumors, metastases) are not usually accompanied by headache. Exceptions of importance are osteomyelitis, multiple myeloma, and Paget disease. Headache may also be caused by lesions of the mastoid and by petrositis. No epidemiologic data are available on headaches due to lesions of the cranial bone.

ANATOMY AND PATHOPHYSIOLOGY

Headache can result from skull lesions that chiefly involve the periosteum (Fig. 121-1), and which are rapidly expansile, are aggressively osteoclastic, or have an inflammatory component (41,51).

Most skull lesions are asymptomatic (58) and are discovered incidentally on roentgenograms or other imaging procedures performed to investigate conditions such as fibrous dysplasia, osteomas, epidermoid cysts, metastatic cancers, hemangiomas, eosinophilic granulomas, and Paget disease of the skull. Hemangiomas, eosinophilic granulomas, and the rare aneurysmal bone cysts may present with a tender swelling on the calvarium but not with spontaneous headache.

Relatively few skull lesions produce headache. Multiple myeloma often presents with bone pain anywhere in the body, and skull deposits are sometimes a source of such pain. The multiplicity of the deposits and the proclivity of the myeloma cells to produce osteoclast-activating factor likely account for the production of head pain by this particular bone tumor. Osteomyelitis produces spontaneous head pain because of its rapid evolution and its inflammatory component. Although most cases of Paget disease of the skull are asymptomatic, remodeling of bone, by producing basilar invagination, may cause headache either through traction on the upper cervical nerve roots or by the production of cerebrospinal fluid pathway distortion with hydrocephalus.

CLINICAL FEATURES

The IHS criteria for headache attributed to disorder of cranial bone (Revised International Classification for Headache Disorders [ICHD-II]) are as follows:

A. Pain in one or more regions of the head or face fulfilling criteria C and D.

B. Clinical, laboratory, and/or imaging evidence of a lesion within the cranial bone known to be, or generally accepted as, a valid cause of headache.

C. Pain develops in close temporal relation to and is maximal over the bone lesion.

D. Pain resolves within 3 months after successful treatment of the bone lesion.

MANAGEMENT

Skull lesions causing headache usually require neurosurgical treatment. If necessary, surgical excision can serve to confirm the diagnosis and retard the progression of

P1: KWW/KKL P2: KWW/HCN QC: KWW/FLX T1: KWW
GRBT050-121 Olesen- 2057G GRBT050-Olesen-v6.cls August 18, 2005 23:22

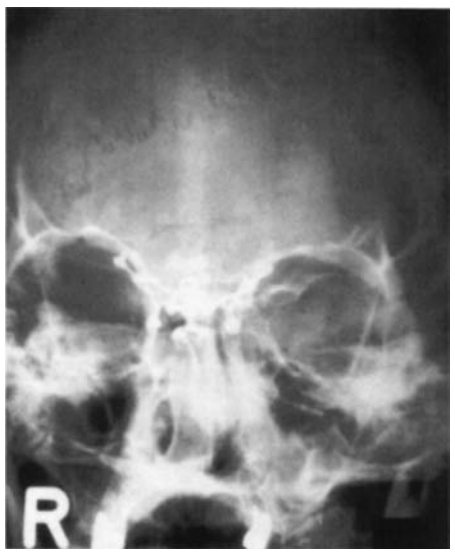

FIGURE 121-1. Headache due to skull fractures after a horse kick.

neurologic dysfunction and head pain. Apart from specific medications that are directed at the underlying disease, nonopioid and opioid analgesics may be used for pain relief.

HEADACHES ATTRIBUTED TO DISORDER OF NECK

IHS code and diagnosis:
11.2 Headache attributed to disorder of neck
11.2.1 Cervicogenic headache
11.2.2 Headache attributed to retropharyngeal tendonitis
11.2.3 Headache attributed to craniocervical dystonia
WHO code and diagnosis:
G44.841 Headache associated with biomechanical lesions of cervical spine (M99.x1)
G44.842 Headache associated with retropharyngeal tendinitis (M79.8)
Short description: Headaches originating from disorders of neck structures and the headache can be relieved by treatments directed at the neck.
Other terms: Cervical headache

Epidemiology

Estimates of the true prevalence of cervicogenic headache require reliable criteria that define the condition. It is insufficient to label posterior headache in the presence of degenerative C-spine disease as cervicogenic because such changes can be found in virtually all people over 40 years of age (14,17).

Prevalence estimates of cervicogenic headache range from 0.4 to 2.5% of the general population to 15 to 20% of patients with chronic headaches (26). Cervicogenic headaches affect patients with a mean age of 42.9 years, have a 4:1 female disposition, and tend to be chronic (26).

TABLE 121-1 Pain-Sensitive Structures of the Neck

Vertebral column
Apophyseal joints
Atlanto-occipital (condylar) joints
Annulus fibrosus
Spinal ligaments
Vertebral periosteum
Cervical muscles
Cervical nerve roots and nerves
Vertebral arteries
Carotid arteries

From Edmeads J. The cervical spine and headache. *Neurology* 1988;38:1874-1878, with permission.

Anatomy and Pathophysiology

Diseases or dysfunction of the neck causes headache when three conditions are met (17):

1. Implicated cervical structures are pain sensitive.
2. The referral pain pattern from neck to head is physiologically based.
3. The disease or dysfunction is identifiable and verifiable.

Nociceptive cervical structures include (2) joints, periosteum, and ligaments of the cervical spine; muscles around the cervical spine; cervical nerve roots and nerves; and vertebral arteries (Table 121-1). Referred pain from these cervical structures to the head is achieved by:

1. C2 sensory root and its extensions, greater and lesser occipital nerves allowing pain referral to the back of the head.
2. Possibly the C1 sensory root (although some authorities believe it to be nonexistent or inconsequential) allowing pain referral to the vertex or frontal head region (33).
3. Connections between the tentorial branches of the ophthalmic division of the trigeminal nerve and posterior fossa branches of C2 allowing pain referral from C2-innervated structures to the front of the head.
4. The descending spinal tract of the trigeminal nerve, intermingling impulses from the upper cervical segments with those from cranial nerve V and allowing pain referral from these segments to the head (34). There is no known physiologic basis for cephalic referral of pain originating from cervical segments below C4.

The *cervical causes of headaches* are listed in Table 121-2. Occipital or suboccipital pain can be the presenting complaint in 26% of developmental anomalies of the craniovertebral junction and upper cervical spine, such as basilar invagination, congenital atlantoaxial dislocation, and separate odontoid; in cases such as these, stretching of

P1: KWW/KKL P2: KWW/HCN QC: KWW/FLX T1: KWW
GRBT050-121 Olesen- 2057G GRBT050-Olesen-v6.cls August 18, 2005 23:22

TABLE 121-2 Cervical Causes of Headache

Accepted causes	■ Developmental anomalies of the craniovertebral junction and upper cervical spine ■ Tumors of craniovertebral junction and upper cervical spine (primary and multiple myeloma) ■ Paget disease of the skull with secondary basilar invagination ■ Osteomyelitis of the upper cervical vertebrae ■ Rheumatoid arthritis of the upper cervical spine ■ Ankylosing spondylitis of the upper cervical spine ■ Traumatic subluxation of the upper cervical vertebrae ■ Retropharyngeal tendinitis ■ Craniocervical dystonias
Controversial causes	■ Cervical disk disease and spondylosis ■ "Whiplash" injuries[a]
Currently unaccepted causes	■ Posterior cervical sympathetic syndrome of Barré ■ Migraine cervicale syndrome of Bartschi-Rochaix

[a] Bogduk's "third occipital headache" and Sjaastad's "cervicogenic headache" are not specific disease entities, but rather syndromes or reaction patterns said to result from a variety of lesions.

upper cervical nerve roots can be seen at surgery. On the other hand, Klippel-Feil anomaly does not produce head or neck pain unless there is excessive mobility between vertebrae rostral to the congenital fusion, with secondary facet joint changes. Also, cervical spina bifida does not cause headache unless associated with soft tissue anomalies such as Arnold-Chiari malformation, with or without hydrocephalus (Fig. 121-2).

Acquired lesions of the craniovertebral junction and upper cervical spine, such as primary tumors (meningioma, schwannoma, ependymoma), Paget disease of the skull with secondary basilar invagination, osteomyelitis of the upper cervical vertebrae, and multiple myeloma of the skull base or upper cervical vertebrae may produce headache by erosion of the pain-sensitive structures or traction on upper cervical nerve roots (Fig. 121-3). Blows to the head or even forceful sneezing may produce rotatory subluxation of the atlas, which, through irritation of synovial joints, causes persistent occipital headache. Rheumatoid arthritis of the upper cervical spine produces headaches through a variety of mechanisms, including inflammation of the synovial atlanto-occipital and atlantoaxial joints and stretching of upper cervical ligaments and nerve roots caused by atlanto-axial subluxation secondary to attenuation of the transverse ligament of the odontoid (this also may occur in ankylosing spondylitis).

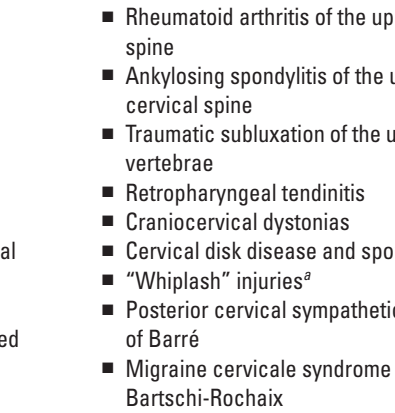

FIGURE 121-2. Arnold-Chiari malformation with persistent movement-dependent headache and neck pain.

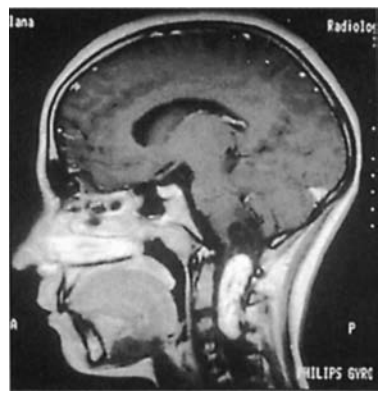

FIGURE 121-3. Intraspinal tumor (cervical ependymoma) with increasing cervicogenic headache.

Cervical spondylosis and *cervical disc disease* are not universally accepted to cause headache because they typically involve the lower cervical discs and vertebrae. Also, their almost ubiquitous existence in people over 40 years old makes it difficult to establish causality or even strong association. Restriction of movement in the lower cervical regions may lead to excessive "play" in rostral apophyseal joints (e.g., at C2–C3 or C3–C4), and could cause head pain (Fig. 121-4), but evidence for this hypothesis remains elusive.

Trauma including "whiplash" injuries is not a widely accepted cause of headache. Clearly, many people with an extension–flexion injury of the neck experience self-limited neck, occipital, and occasionally frontal pain, which clears within days or a few weeks. Likely, this pain results from injury to the upper cervical ligaments and muscles. Others believe that shearing injuries of the long axons in the brainstem and upper cord may disrupt central pain and other regulatory mechanisms, allowing the emergence of headache (5,28,54,58,60).

P1: KWW/KKL P2: KWW/HCN QC: KWW/FLX T1: KWW
GRBT050-121 Olesen- 2057G GRBT050-Olesen-v6.cls August 18, 2005 23:22

FIGURE 121-4. Patient suffering from cervicogenic headache after cervical spine fusion. From Göbel H, ed. *Botulinum Toxin A in der speziellen Schmerztherapie.* Uni-Med-Verlag: Bremen, 2004, with permission.

Clinical Features

The IHS diagnostic criteria for cervicogenic headache (30) (ICHD-II) are as follows:

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 one of the following:
  1. Demonstration of clinical signs that implicate a source of pain in the neck[2]
  2. Abolition of headache following diagnostic blockade of a cervical structure or its nerve supply using placebo or other adequate control
D. Pain resolves within 3 months after successful treatment of the causative disorder or lesion.

The ICHD-II includes those footnotes or qualifiers to the criteria for cervicogenic headache. These are:

1. Tumors, fractures, infections, and rheumatoid arthritis of the upper cervical spine have not been validated formally as causes of headache, but are nevertheless accepted as valid causes when demonstrated to be so in individual cases. Cervical spondylosis and osteochondritis are NOT accepted as valid causes fulfilling criterion B. When myofascial tender spots are the cause, the headache should be coded under 2. Tension-type headache.
2. Clinical signs acceptable for criterion C1 must have demonstrated reliability and validity. The future task is the identification of such reliable and valid operational tests. Clinical features such as neck pain, focal neck tenderness, history of neck trauma, mechanical exacerbation of pain, unilaterality, coexisting shoulder pain, reduced range of motion in the neck, nuchal onset, nausea, vomiting, photophobia, etc., are not unique to cervicogenic headache. These may be features of cervicogenic headache, but they do not define the relationship between the disorder and the source of the headache.
3. Abolition of headache means complete relief of headache, indicated by a score of 0 on a visual analog scale (VAS). Nevertheless, acceptable as fulfilling criterion C2 is $\geq$90% reduction in pain to a level of $<$5 on a 100-point VAS.

While there is nothing specific about headaches from craniovertebral anomalies, they tend to exhibit some suggestive features, such as posterior location, triggering by flexing of the neck or by coughing and straining, and sometimes a pronounced postural component that may mimic low-pressure headaches (35). Depending on what neural anomalies are associated, there may be complaints of vertigo, facial numbness, limb weakness, or ataxia, as well as neurologic findings related to the upper cervical nerve roots, lower brainstem, or upper cervical cord.

As with the congenital atlantoaxial and upper cervical anomalies, acquired disorders tend to produce occipital headaches that are worsened or triggered by neck movements or straining, although often the postural element of the headache is not so evident (Fig. 121-5). In addition,

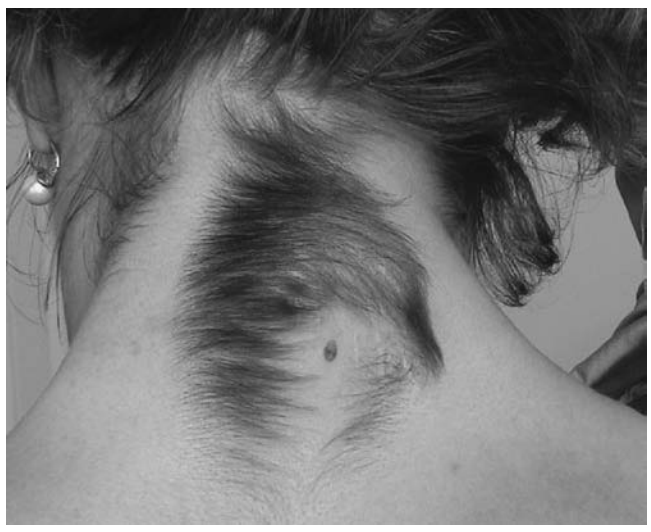

FIGURE 121-5. Patient suffering from cervicogenic headache attributed to meningomyelocele. From Göbel H, ed. *Botulinum Toxin A in der speziellen Schmerztherapie.* Uni-Med-Verlag: Bremen, 2004, with permission.

P1: KWW/KKL P2: KWW/HCN QC: KWW/FLX T1: KWW
GRBT050-121 Olesen- 2057G GRBT050-Olesen-v6.cls August 18, 2005 23:22

aids to diagnosis include the stigmata of the underlying diseases encountered in the history, examination, and investigations. When examining patients with rheumatoid arthritis who have headache, care should be taken when the patient is asked to flex the neck, because occasional fatalities have resulted from compression of the medulla by the odontoid, which, no longer bound to the atlas by the transverse ligament, fails to move away from the brainstem on anteflexion of the cervical spine.

Researchers who teach that cervical spondylosis is an important cause of headache describe the headache of cervical disc disease as being posterior, often unilateral, sometimes constant, nonthrobbing, and aggravated by neck movements and occasionally by coughing and straining (42).

There is no specific phenotype that describes headache of cervical origin. The quality of pain is variably described and the headache could be unilateral or bilateral (1,8–13,30,36,47–50,55,59).

Furthermore, pericranial tenderness and myofascial tender spots are elicited (32) but in themselves do not provide evidence of cervicogenic headache. Lastly, thickening of the skin (pinch-and-roll sign) may be observed in patients with cervicogenic headache (40) but the reliability and validity of this sign are unknown.

Sjaastad and colleagues have suggested that many patients with cervicogenic headache report a fairly uniform headache profile pointing to the neck as the origin of the headache (46–50). The profile includes: (a) unilateral pain (always on the same side) beginning in the neck and eventually spreading to the oculofrontotemporal areas, where the maximum pain often is "located"; (b) moderate and nonthrobbing pain, intermittent or continuous, and provoked by neck movements or sustained awkward head postures or by pressure over the posterior neck or occiput; (c) nonradicular neck, shoulder, and arm pain; (d) reduced range of motion in the cervical spine; (e) female gender; (f) a history of head or neck trauma; and (g) transient relief by anesthetic blockade of the major occipital nerve or the C2 root. Nausea, vomiting, phonophobia or photophobia, edema or flushing around the eye, visual blurring ipsilateral to the pain, and difficulty swallowing are some accompanying symptoms that are reported but are not necessary for the diagnosis.

Sjaastad and colleagues carefully stipulated that "cervicogenic headache" is not a disease entity but rather a reaction pattern (46–50). Also, they emphasize that the headache may emanate from several structures or processes in the neck, including bone and soft tissue. However, the authors took a controversial position when they concluded that cervicogenic headache is extremely common and may be misclassified as migraine without aura.

Reports of the cervical spine investigations in patients with suspected cervicogenic headache have been inconsistent (46–50). Sjaastad and colleagues found no differences between radiographs of the cervical spine of patients with cervicogenic headache and those of age- and sex-matched controls (46–50). Pfaffenrath and colleagues found reduced mobility in the upper cervical spine in cervicogenic headache patients compared to controls (43).

MANAGEMENT

An important first step in the diagnosis of cervicogenic headache is to identify *symptomatic forms* and treat the underlying cause(s), if possible. Ipsilateral blockade of the C2 root or greater occipital nerve allows the differentiation between cervicogenic headache due to irritation of the C2 root, where a short-lived benefit is observed, and primary headache syndromes such as migraine and tension-type headache. Symptomatic pharmacologic treatment and surgical or chiropractic procedures have not been very successful in leading to a significant improvement or remission. Furthermore, side effects of analgesics, antiinflammatory drugs, and muscle relaxants preclude long-term treatment. Small, noncontrolled case series have reported moderate success with surgery and injections. Physiotherapy, including muscle relaxation techniques and psychotherapy, can provide some relief but systematic studies are lacking. Finally, a few randomized controlled trials and a number of case series support the use of cervical manipulation, transcutaneous electrical nerve stimulation, and botulinum toxin A injections (26).

HEADACHE ATTRIBUTED TO RETROPHARYNGEAL TENDINITIS

Retropharyngeal tendinitis is an uncommon condition of unknown etiology that is characterized by the acute onset of upper cervical and occipital pain, aggravated by neck movements (especially extension) and accompanied by pain on swallowing and, in the early stages, tenderness in the sides of the upper neck, mild to moderate fever, and often an increased erythrocyte sedimentation rate. Radiographs of the cervical spine show increased thickness of the C1–C4 prevertebral soft tissue, sometimes with calcification (computerized tomography may show this better). Upper carotid dissection should be ruled out.

CLINICAL FEATURES

The IHS diagnostic criteria for headache attributed to retropharyngeal tendonitis (ICHD-II) are as follows:

A. Unilateral or bilateral nonpulsating pain in the back of the neck, radiating to the back of the head or to the whole head and fulfilling criteria C and D.

P1: KWW/KKL P2: KWW/HCN QC: KWW/FLX T1: KWW
GRBT050-121 Olesen- 2057G GRBT050-Olesen-v6.cls August 18, 2005 23:22

TABLE 121-3 Main Clinical Characteristics of Focal Dystonias of the Head and Neck Accompanied by Pain

Form	*Main Clinical Characteristics*	*Pain Criteria*
Mandibular dystonia	Dystonia of the jaw-closing or jaw-opening muscles. The principal symptoms are the defective jaw position and the painful spasms of the masticatory muscles. Patients may bite their own tongue. In severe cases, dystonia of the jaw-closing muscle in particular leads to heavy wear of the teeth, or even loss of teeth due to their breaking out.	Owing to hypercontracted muscles, pain is frequent and often involves great suffering; as a secondary symptom, a mandibular joint syndrome or oromandibular dysfunction may occur and maintain pain; pain often corresponds to tension-type headache.
Pharyngeal dystonia	Pharyngeal dystonia or spasmodic dysphagia occurs very rarely as isolated focal dystonia. More frequently, it is a symptom of the Meige syndrome, or it accompanies lingual and laryngeal dystonia. Even if it dominates the overall clinical picture, dystonia of other muscle groups will also be found as a rule.	Very annoying sensation of cramp and tension
Torticollis spasmodicus (TS)	The main symptom is the abnormal movement or position of the head; depending on the dominant direction of movement, a distinction is made between rotatory TS (the most common variant), laterocollis, and retrocollis or anterocollis. Combinations of these positions are found in about 66% of patients.	Pain in neck region: pain is a principal symptom of the condition; the pain is caused by local contractions and secondary vertebrogenic changes, in rare cases going as far as vertebral luxation with paraplegia; responds well to botulinum toxin and nonsteroidal antirheumatics
Lingual dystonia	Involuntary sticking out of tongue; continual rolling movements of the tongue, which can be observed particularly well with the mouth open; sometimes accompanying oral movements	Occasionally accompanied by pain
Segmental craniocervical dystonia	Combinations of the cranial and cervical dystonias described above; a combination of blepharospasm with an oromandibular dystonia is most common (Meige syndrome)	

B. Swollen prevertebral soft tissues, in adults measuring >7 mm at the level between C1 and C4 (special x-ray technique may be required)
C. Pain is aggravated severely by bending the head backwards.
D. Pain is alleviated within 2 weeks of treatment with nonsteroidal anti-inflammatory drugs (NSAIDs) in their recommended doses.

MANAGEMENT

The symptoms and the prevertebral swelling subside over several days, although resolution may be accelerated by using NSAIDs (20,21).

HEADACHE ATTRIBUTED TO CRANIOCERVICAL DYSTONIA

Headache attributed to craniocervical dystonia is included in the second IHS classification for the first time (22,30). Pain arising from *craniocervical dystonia* is either due to continuous contraction of muscles, or it may occur as a result of secondary irritation of neural structures, for example, at the emergence of the occipital nerves, induced by muscular hyperactivity. If the condition is prolonged, it may give rise to degenerative changes of the cervical spine, mandibular joint, or dentition, which may cause additional local pain. The continuous contraction may lead to hypertrophy of the affected muscles.

Dystonia can be primary (*idiopathic*) or secondary (*symptomatic*). Focal dystonias in the head and neck region (Table. 121-3) can be *cranial* (blepharospasm, spasmodic dysphonia, mandibular, or lingual dystonia) or *cervical dystonias* (torticollis). The incidence of *craniocervical dystonia* (CCD) is 5 to 15 per million (45).

Under criterion B, the IHS classification lists abnormal movements or defective posture of neck or head due to muscular hyperactivity as a headache cause. According to criterion D, the pain must disappear within 3 months after successful treatment. Treatment with botulinum toxin A is the therapy of first choice. Thus, for the first time botulinum toxin A also acquires diagnostic significance within headache therapy.

P1: KWW/KKL P2: KWW/HCN QC: KWW/FLX T1: KWW
GRBT050-121 Olesen- 2057G GRBT050-Olesen-v6.cls August 18, 2005 23:22

CLINICAL FEATURES

The IHS diagnostic criteria for headache attributed to craniocervical dystonia (ICHD-II) are as follows:

A. Sensation of cramp, tension or pain in the neck, radiating to the back of the head or to the whole head and fulfilling criteria C and D.
B. Abnormal movements or defective posture of neck or head due to muscular hyperactivity.
C. Evidence that pain is attributed to muscular hyperactivity based on at least one of the following:
  1. Demonstration of clinical signs that implicate a source of pain in the hyperactive muscle (e.g., pain is precipitated or exacerbated by muscle contraction, movements, sustained posture, or external pressure)
  2. Simultaneous onset of pain and muscular hyperactivity
D. Pain resolves within 3 months after successful treatment of the causative disorder.

The common central symptom of focal dystonia is the abnormal movement or defective position of the affected parts of the body. The underlying muscular hyperactivity may be tonic and therefore give rise to a *slow, smooth movement* or a *fixed defective position*; it may be *phasal*, thereby causing *jerky*, *repetitive movements* (*myoclonic dystonia*), or it may have a *rhythmic movement* character, which frequently makes it difficult to distinguish from various forms of tremor (18,19,23,24,27,57).

Craniocervical dystonias can be classified on the basis of the musculature affected. Both isolated dystonias of individual muscle areas served by cranial nerves and combinations of various types of focal dystonia may occur.

Headaches are a frequent (70%), and sometimes (35%) the principal, complaint in focal dystonia (25). Unilateral and occipital pain are the most prevalent (45). The pain of cervical dystonia with rotatory components is usually in the back of the head and to the site of head rotation. Cervical dystonia is not associated with pain in the region of the anterior cervical musculature. Pain of cervical dystonia is multifactional. Generally, it stems from pericranial muscle origins or attachments. Painful muscles are those straining consciously or by reflex to correct the abnormal, dystonia-induced head position. Also, involvement of small deep cervical muscle either by the dystonia itself or by abnormal position may contribute to pain. Regardless, there is no correlation between the intensity of the pain and the degree of abnormality of head position (15). Furthermore, headaches in cases of cervical dystonia are due to factors such as overstrained fibers in the dystonic muscles, aseptic inflammation of the muscle/tendon attachments, stretching of the autochthonous cervical spine musculature and the cervical spine ligaments, and/or irritation of nerve structures by the defective position of the cervical spine and its joints (Figs. 121-6 and 121-7).

FIGURE 121-6. Complex form of a painful cervical dystonia in a 78-year-old woman with permanent and severe cervicogenic headache. From Reichel G. Botulinumtoxin in der Therapie schmerzhafter fokaler und segmentaler Dystonien. In: Göbel H, ed. *Botulinum toxin A in der speziellen Schmerztherapie.* Uni-Med-Verlag: Bremen, 2004, with permission.

MANAGEMENT

The beneficial effect of botulinum toxin on pain in cervical dystonia was first reported in 1985 (52), and more recently reviewed (37). Success rates of 80 to 100% are reported in open label studies (37), but the range is 40 to 88% in double-blind trials (29,39,53). In practice, virtually all patients report an improvement in dystonia-induced pain (16).

Botulinum toxin best improves the pain of cervical dystonia followed by muscular hypertrophy, defective position and restriction of movement, and dystonic tremor capitis. Pain alleviation frequently occurs before, or sometimes without, improvement in motor dysfunction (45).

A correct choice of muscles for injection and the right botulinum toxin dose per muscle are key to therapeutic success. Total dose of botulinum may not be as relevant to success (44). Also, it may not be necessary to inject botulinum toxin into the painful muscles if the clinical picture and the imaging findings show that these are not dystonic (31,38).

Botulinum toxin therapy generally produces good results for 3 months. If the effect is insufficient, a fresh injection should not be given until after at least 6 weeks.

P1: KWW/KKL P2: KWW/HCN QC: KWW/FLX T1: KWW
GRBT050-121 Olesen- 2057G GRBT050-Olesen-v6.cls August 18, 2005 23:22

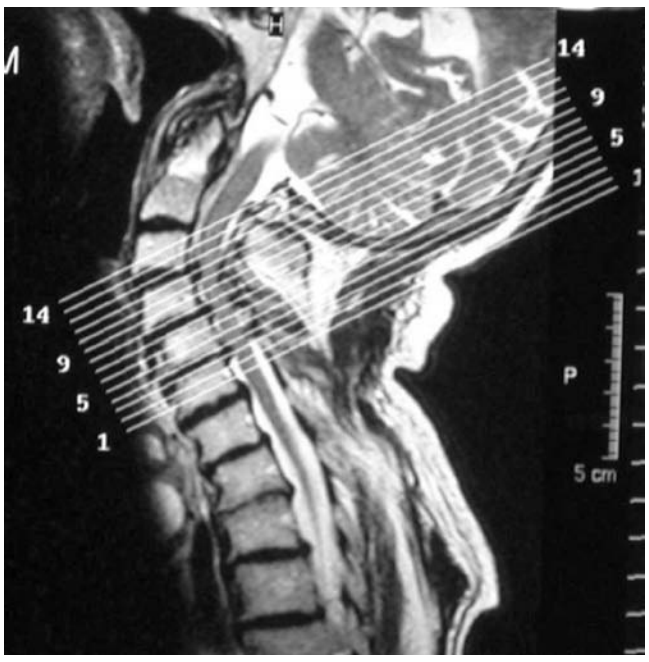

FIGURE 121-7. Magnetic resonance imaging of a patient with painful cervical dystonia and cervicogenic headache showing defective posture of neck and head due to muscular hyperactivity. From Reichel G. Botulinumtoxin in der Therapie schmerzhafter fokaler und segmentaler Dystonien. In: Göbel H, ed. *Botulinum toxin A in der speziellen Schmerztherapie.* Uni-Med-Verlag: Bremen, 2004, with permission.

Surgical resection of the peripheral nerve can be considered when botulinum toxin fails, but the long-term effects of such a procedure on pain have not been studied systematically. Patients who have undergone an operation frequently complain of nonexistent or only temporary effects on the pain and the dystonic movements.

SYNDROMES NOT FULLY ACCEPTED

The third occipital nerve, which is the superficial medial branch of the C3 dorsal ramus, supplies the C3 dermatome, part of the semispinalis capitis muscle, and the C2–C3 zygapophyseal joint. Bogduk and Marsiand shocked this nerve in a group of patients who complained of occipital headaches radiating frontally along with at least one feature suggestive of a cervical origin for the pain (e.g., history of neck injury, triggering of pain by neck movement [6]) Relief was observed in two thirds of patients. The authors later postulated that disease of the C2–C3 zygapophyseal joints could produce headaches with pain transmitted by the third occipital nerve (7), but there was no radiologic evidence of any such disease. An alternative explanation is that the nerve blocks relieved headache in a nonspecific fashion by interrupting normal afferent impulses that, along with other impulses, were helping to reduce the stimulation threshold of the trigeminal system in the upper cervical cord. This alternative hypothesis emphasizes that relief of pain following nerve block does not mean necessarily that the pain emanates from a structure supplied by that nerve.

Modern research no longer supports the existence of the posterior cervical sympathetic syndrome of Barré (3) or of the "migraine cervicale" syndrome of Bartschi-Rochaix (4).

REFERENCES

1. Aprill C, Axinn MJ, Bogduk N. Occipital headaches stemming from the lateral atlanto-axial (C1-2) joint. *Cephalalgia* 2002;22(1):15–22.
2. Bansevicius D, Pareja JA, Sjaastad O. "Skin roll" ('pinch and roll') test: skinfold thickness and tenderness. *Headache* 1997;37:281–285.
3. Barré JA. Sur un syndrome sympathique cervical posterjeur et sa cause frequent, I'arthrite cervicale. *Rev Neurol* 1926;33:1246–1248.
4. Bartschi-Rochaix W. Headache of cervical origin. In: Vinken PJ, Bruyn GW, eds. *Handbook of clinical neurology, Vol 5, Headache*. Amsterdam: North-Holland, 1968:192–203.
5. Berry H. Psychological aspects of chronic neck pain following hyperextension–flexion strains of the neck. In: Morley TP, ed. *Current controversies in neurosurgery*. Philadelphia: WB Saunders, 1976:51–61.
6. Bogduk N, Marsiand A. On the concept of third occipital headache. *J Neurol Neurosurg Psychiatry* 1986;49:775–780.
7. Bogduk N, Marsland A. The cervical zygapophyseal joints as a source of neck pain. *Spine* 1988;13:610–617.
8. Bogduk N. The neck. *Baillieres Best Pract Res Clin Rheumatol* 1999; 13(2):261–285.
9. Bogduk N. C2 ganglion can be injured by compression between the posterior arch of the atlas and the lamina of C2. *Spine* 1999; 24(3):308–309.
10. Bogduk N. Cervicogenic headache: anatomic basis and pathophysiologic mechanisms. *Curr Pain Headache Rep* 2001;5(4):382–386.
11. Bogduk N. Diagnostic nerve blocks in chronic pain. *Best Pract Res Clin Anaesthesiol* 2002;16(4):565–578.
12. Bogduk N. The anatomy and pathophysiology of neck pain. *Phys Med Rehabil Clin N Am* 2003;14(3):455–472, v.
13. Bogduk N, Lord SM. Cervical spine disorders. *Curr Opin Rheumatol* 1998;10(2):110–115.
14. Bono G, Antonaci F, Ghirmai S, et al. The clinical profile of cervicogenic headache as it emerges from a study based on the early diagnostic criteria. *Funct Neurol* 1998;13:75–77.
15. Chawda SJ, Munchau A, Johnson D, et al. Pattern of premature degenerative changes of the cervical spine in patients with spasmodic torticollis and the impact on the outcome of selective peripheral denervation. *J Neurol Neurosurg Psychiatry* 2000;68(4):465–471.
16. Dressler D. *Botulinum toxin therapy*. Stuttgart: Thieme, 2000.
17. Edmeads J. The cervical spine and headache. *Neurology* 1988;38: 1874–1878.
18. Erbguth FJ. Pain in cervical dystonia. In: Jost WH, ed. *Botulinum toxin in painful diseases*. Karger Basel 2003:54–70.
19. Erbguth F. *Das Krankheitsbild der zervikalen Dystonie (Torticollis spasmodicus)*. Regensburg: Roderer Verlag, 1996.
20. Fahlgren H. Retropharyngeal tendinitis: three probable cases with an unusually low epicenter. *Cephalalgia* 1988;8:105–110.
21. Fahlgren R. Retropharyngeal tendinitis. *Cephalalgia* 1986;6:169–174.
22. Göbel H. *Die Kopfschmerzen. Ursachen, Mechanismen, Diagnostik und Therapie in der Praxis*. Berlin: Springer-Verlag, 2004:1–901.
23. Göbel H, Heinze A, Heinze-Kuhn K, et al. Botulinum toxin A in the treatment of headache syndromes and pericranial pain syndromes. *Pain* 2001;91(3):195–199.
24. Göbel H, Heinze A, Heinze-Kuhn K, et al. Evidence-based medicine: botulinum toxin A in migraine and tension-type headache. *J Neurol* 2001;248[Suppl 1]:34–38.

P1: KWW/KKL P2: KWW/HCN QC: KWW/FLX T1: KWW
GRBT050-121 Olesen- 2057G GRBT050-Olesen-v6.cls August 18, 2005 23:22

25. Göbel H, ed. *Botulinum Toxin A in der speziellen Schmerztherapie*. Uni-Med-Verlag: Bremen, 2004.
26. Haldeman S, Dagenais S. Cervicogenic headaches: a critical review. *Spine J* 2001;1(1):31–46.
27. Hallett M, Glocker FX, Deuschl G. Mechanism of action of botulinum toxin. *Ann Neurol* 1994;36:449–450.
28. Hawkins GW. Flexion and extension injuries of the cervicocapital joints. *Clin Orthop* 1962;24:22–33.
29. Headache Classification Committee of the International Headache Society. Classification and diagnostic criteria for headache disorders, cranial neuralgias and facial pain. *Cephalalgia* 1988;8[Suppl 7]: 1–96.
30. Headache Classification Committee of the International Headache Society. International classification of headache disorders, 2nd edition. *Cephalalgia* 2004;24[Suppl 1]:9–160.
31. Hobson DE, Gladish DF. Botulinum toxin injection for cervicogenic headache. *Headache* 1997;37:253–255.
32. Jaeger B. Are "cervicogenic" headaches due to myofascial pain and cervical spine dysfunction? *Cephalalgia* 1989;9:157–164.
33. Kerr FWL, Olafson RA. Trigeminal and cervical volleys. *Arch Neurol* 1961;5:171–178.
34. Kerr FWL. A mechanism to account for frontal headaches in cases of posterior fossa tumors. *J Neurosurg* 1961;18:605–609.
35. Khurana R. Headache spectrum in Arnold-Chiari malformation. *Headache* 1991;31:151–155.
36. Leone M, D'Amico D, Grazzi L, et al. Cervicogenic headache: a critical review of the current diagnostic criteria. *Pain* 1998;78(1):1–5.
37. Lew MF. Review of the FDA-approved uses of botulinum toxins, including data suggesting efficacy in pain reduction. *Clin J Pain* 2002;18[6 Suppl]:142–146.
38. Lobbezoo F, Tanguay R, Thon MT, et al. Pain perception in idiopathic cervical dystonia (spasmodic torticollis). *Pain* 1996;67:483–491.
39. Lorentz IT, Subramaniam SS, Yiannikas C. Treatment of idiopathic spasmodic torticollis with botulinum toxin A: a double-blind study on twenty-three patients. *Mov Disord* 1991;6:145–150.
40. Maigne R. *Douleur d'origine vertebrale et traitment par manipulation*. 3rd ed. Paris: Expansion Scientifique, 1977.
41. McRae DL. Bony ahnormalities at the craniospinal junction. *Clin Neurosurg* 1969;16:356–375.
42. Peterson D, Austin G, Dayes L. Headaches associated with discogenic disease of the cervical spine. *Bull Los Angeles Neurol Soc* 1975;40:96–100.
43. Pfaffenrath V, Dandekar R, Mayer E, et al. Cervicogenic headache: results of computer-based measurements of cervical spine mobility in 15 patients. *Cephalalgia* 1988;8:45–48.
44. Poewe W, Deuschl G, Nebe A, et al. What is the optimal dose of botulinum toxin A in the treatment of cervical dystonia? Results of a double blind, placebo controlled dose ranging study using Dysport. *J Neurol Neurosurg Psychiatry* 1998;64:13–17.
45. Reichel G. Botulinumtoxin in der Therapie schmerzhafter fokaler und segmentaler Dystonien. In: Göbel H, ed. *Botulinum toxin A in der speziellen Schmerztherapie*. Uni-Med-Verlag: Bremen, 2004.
46. Sjaastad O, Bovim G. Cervicogenic headache: the differentiation from common migraine. An overview. *Funct Neurol* 1991;6:93–100.
47. Sjaastad O, Fredriksen TA, Pfaffenrath V. Cervicogenic headache: diagnostic criteria. *Headache* 1990;30:725–726.
48. Sjaastad O, Fredriksen TA, Stolt-Nielsen A, et al. Cervicogenic headache: a clinical review with special emphasis on therapy. *Funct Neurol* 1997;12:305–317.
49. Sjaastad O, Salvesen R, Jansen J, et al. Cervicogenic headache: a critical view on pathogenesis. *Funct Neurol* 1998;13:71–74.
50. Sjaastad O. Reliability of cervicogenic headache diagnosis. *Cephalalgia* 1999;19(9):767–768.
51. Soyka D. Zervikogener Kopfschmerz. *Nervenheilkunde* 1990;9:265–267.
52. Tsui JKC, Eisen A, Mak E. A pilot study on the use of botulinum toxin in spasmodic torticollis. *Neurol Sci* 1985;12:314–316.
53. Tsui JKC, Eisen A, Stoessl AJ, et al. Double blind study of botulinum toxin in spasmodic torticollis. *Lancet* 1968;II:245–247.
54. Vincent MB, Luna RA. Cervicogenic headache: a comparison with migraine and tension-type headache. *Cephalalgia* 1999;19[Suppl 25]: 11–16.
55. Vincent MB, Luna RA, Scandiuzzi D, et al. Greater occipital nerve blockade in cervicogenic headache. *Arq Neuropsiquiatr* 1998;56(4):720–725.
56. Voorhies RM, Sundaresan N. Tumors of the skull. In: Wilkins RH, Rengachary SS, eds. *Neurosurgery*. New York: McGraw-Hill, 1985: 984–1001.
57. Tarsy D, First ER. Painful cervical dystonia: clinical features and response to treatment with botulinum toxin. *Mov Disord* 1999;14(6):1043–1045.
58. Weiss RD, Stern BJ, Goldberg J. Post-traumatic migraine: chronic migraine precipitated by minor head or neck trauma. *Headache* 1991; 31:451–456.
59. Wilk V. What of cervicogenic headaches? *Aust Fam Physician* 1998; 27(10):877–878.
60. Zwart JA. Neck mobility in different headache disorders. *Headache* 1997;37:6–11.