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Case Studies of Uncommon Headaches

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The following nine cases present uncommon but fascinating primary and secondary headache disorders. If you are not already familiar with or have not seen these disorders, you will be prepared when someone presents or a colleague asks you about an unusual case.

Case 1. Noises in the night

A 43-year-old woman was seen with a 5-month history of a noise in her head. On an almost nightly basis, as she was falling asleep, she would hear a loud noise like "electrical current running" lasting a second. Sometimes her whole body would shake for a second afterwards. Very occasionally, she would have an associated flash of light. Frequently, a second episode of the loud noise occurred shortly after the first. She then could fall asleep without any problem.

Her medical history was positive only for hypertension controlled with medication. Neurologic examination was normal. Diagnostic testing was not performed [1].

Questions

What is the diagnosis? Which type of headaches can awaken patients from sleep?

Discussion

These episodes are characteristic of exploding head syndrome, a disorder named by Pearce in 1988 [2]. Episodes of exploding head syndrome, which occur on falling asleep or, less often, on awakening, awaken people from

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sleep with a sensation of a loud bang in the head, like an explosion. Ten percent of cases are associated with the perception of a flash of light. Five percent of patients report a curious sensation as if they had stopped breathing and had to make a deliberate effort to breathe again. The episodes have a variable frequency and onset at any age, although the most common is middle age and older. The episodes take place in healthy individuals during any stage of sleep without evidence of epileptogenic discharges. The basis of this syndrome may be a delay in the reduction of activity in selected areas of the brainstem reticular formation as the patient passes from wakefulness to sleep. Symptoms typically resolve with time and with reassurance that the disorder is benign.

Secondary causes of nocturnal headaches include drug withdrawal, temporal arteritis, sleep apnea, nocturnal hypertension-headache syndrome, oxygen desaturation, pheochromocytomas, primary and secondary neoplasms, communicating hydrocephalus, subdural hematomas, subacute angle-closure glaucoma, and vascular lesions [3]. Migraine, cluster, hypnic, and chronic paroxysmal hemicrania are primary headaches that can cause awakening from sleep. Migraine typically has associated symptoms and uncommonly occurs only during sleep. Cluster headaches have autonomic symptoms and may occur during the day and during sleep. Chronic paroxysmal hemicrania occurs during the day and at night, lasts for less than 30 minutes, and occurs 10 to 30 times a day.

Case 2. Headache causing awakening from sleep

> A 56-year-old man who had a history of well-controlled hypertension and episodic tension-type headache was awoken from sleep 3 nights per month for the past 2 years between 1:00 AM and 2:30 AM with a moderately severe bifrontal pressing headache, which persisted for 1 to 2 hours untreated and 10 minutes if treated with a caffeine-containing over-the-counter analgesic [4].

Questions

What is the diagnosis? Which other treatments might be effective?

Discussion

Hypnic headache is a rare disorder, first reported by Raskin in 1988 [5], that occurs more often in the elderly (but with a range of 36 to 83 years of age and a single case of a 9 year old [6]) with a female predominance [7]. The headaches occur only during sleep and awaken the sufferer at a consistent time. Nausea is infrequent, and autonomic symptoms are rare. The headaches can be unilateral or bilateral, throbbing or nonthrobbing, and mild to severe in intensity. During the headaches, patients typically prefer to sit up or stand, as lying supine may intensify the pain. The headaches can last 15 minutes to 3 hours and can occur frequently, as often as nightly,

for many years. Spontaneous resolution is uncommon. There are two case reports of secondary hypnic headache: one patient who had obstructive sleep apnea with resolution of headaches with use of continuous positive airway pressure [8] and a second who had a posterior fossa meningioma with resolution of headaches after removal [9].

Medications reported as effective include caffeine (one or two cups of caffeinated coffee or a 40- to 60-mg caffeine tablet before bedtime), lithium carbonate (300 mg at bedtime), indomethacin, atenolol, melatonin, cyclobenzaprine, verapamil, pizotifen, gabapentin, clonidine, acetylsalicyclic acid, acetaminophen, ergotamine derivatives, acetazolamide, prednisone, and flunarizine (not available in the United States). The diagnosis is one of exclusion.

Case 3. Numb tongue

This is a 15-year-old boy who had a history of migraines since approximately age 4. The migraines occur approximately once every 6 months and are described as a severe bifrontal throbbing associated with nausea, light and noise sensitivity, and sometimes vomiting but no aura. The headaches last approximately a half day, and he often goes to bed during the attack. Acetaminophen is of mild help.

For the past 6 months, he has developed a new type of headache, occurring approximately once every 2 weeks. These headaches are triggered by activity, such as throwing a football hard, hitting an overhead ball in tennis, or chasing his dog around in a circle, but not by weightlifting or straining with a bowel movement. He describes a sharp pain in the mid nuchal-occipital region associated with numbness of the left side of the tongue, all lasting approximately 20 seconds.

Medical history is negative except for congenital bilateral hearing loss. Family history is remarkable for migraines in his father and congenital hearing loss. Neurologic examination is normal, except for profound hearing loss bilaterally. MRI scan of the brain shows a large retrocerebellar arachnoid cyst without hydrocephalus. The cerebellar tonsils are above the foramen magnum [10].

Question

What is the cause of the transient episodes of nuchal-occipital pain and numbness of the left side of the tongue?

Discussion

This teenager has neck-tongue syndrome first described by Lance and Anthony in 1980 [11]. The arachnoid cyst is an incidental finding. Neck-tongue syndrome is an uncommon disorder characterized by acute unilateral occipital pain and numbness of the ipsilateral tongue lasting seconds to 1 minute and precipitated by sudden movement, usually rotation, of the head. The

symptoms are the result of transient subluxation of the atlantoaxial joint that stretches the joint capsule and the C2 ventral ramus, which contains proprioceptive fibers from the tongue originating from the lingual nerve to the hypoglossal nerve to the C2 root (Fig. 1). Although neck-tongue syndrome can occur without obvious abnormalities, associated disorders include degenerative spondylosis, ankylosing spondylitis, psoriatic arthritis, and genetically determined laxity of ligaments of joint capsules. A benign, familial form of neck-tongue syndrome is described without anatomic abnormality, which resolves spontaneously during adolescence [12].

Case 4. I feel really big, no make that really small

A 31-year-old woman has a 10-year history of mild headaches occurring approximately once in a month. For the past 6 months, the headaches have increased in frequency and severity, occurring 5 days per week, and are described as a pressure around her eyes with nausea and light sensitivity typically of moderate intensity but severe once a week lasting many hours. There was no aura. There is no family history of migraine. Neurologic examination was normal. A MRI scan of the brain was normal. She was started on topiramate (25 mg daily) to be increased by 25 mg per week to 100 mg per day.

When seen at follow-up 1 month later, the headaches were mild and had decreased to 1 or 2 per week. She reported, however, having five episodes during the prior 3 weeks lasting 2 to 3 minutes and followed by a mild pressure headache behind her eyes lasting 30 to 45 minutes without medication. With three of the episodes, she felt like her entire body was

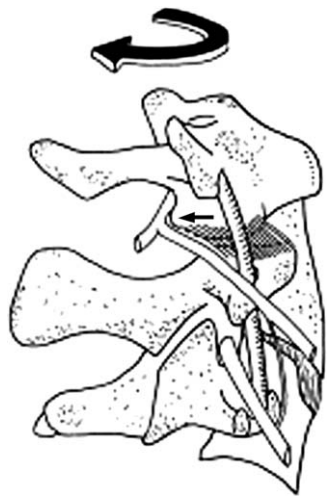

Fig. 1. A lateral view of a right atlantoaxial joint in which the atlas has rotated to the right. (*From* Evans RW, Lance JW. Transient headache with numbness of half of the tongue. Headache 2000;40:692–3; with permission.)

too big and everything else was too small. With two of the episodes, she had a feeling that her entire body was too small and everything else too big. During the episodes, however, everything actually looked normal and she was aware her abnormal feeling was not real. An EEG was normal.

At follow-up 5 weeks later, she reported eight episodes all lasting approximately 5 to 10 minutes. During four of the episodes she reported feeling too small and with two, too big. With two episodes, she felt too big for approximately 5 minutes and then too small for approximately 5 minutes. All of the episodes were followed by the mild headache lasting approximately 1 hour [13].

Questions

What is the diagnosis? What is the derivation of the name of this disorder?

Discussion

''Alice in Wonderland'' syndrome is a rare migraine aura where patients experience distortion in body image characterized by enlargement, diminution, or distortion of part of or the whole body, which they know is not real. The syndrome can occur at any age but is more common in children. The cause may be migrainous ischemia of the nondominant posterior parietal lobule. Although most common with migraine, it also is reported after viral encephalitis (especially after Epstein-Barr virus) and as an epileptic phenomenon.

Although previously not reported as a side effect of medication use, case four's ''Alice in Wonderland'' syndrome may be associated with topiramate use [14]. Her visual symptoms started 1 week after starting topiramate and resolved 1 month after stopping topiramate (2.5 months after initiation). After follow up 2 years and 9 months later, she had no further episodes. This association seems plausible with my recent report [14] of two migraneurs with reversible palinpopsia associated with topiramate use. Both sufferers had resolution of the palinposia on discontinuation of topiramate and the same visual symptoms again when topiramate was restarted.

Lippman describes seven migraineurs who had unusual distortions of body image in 1952 [15]. The descriptions of four patients are illustrative: ''Occasionally the patient has an attack where she feels small, about 1 ft high.'' Another patient had the sensation of ''her left ear ballooning out six inches or more.'' A third patient described his sensations: ''the body is as if someone had drawn a vertical line separating the two halves. The right half seems to be twice the size of the left half.'' And a fourth noted, ''I feel that my body is growing larger and larger until it seems to occupy the whole room.''

In 1955, Todd gave the syndrome its name [16] from the book, *Alice's Adventures in Wonderland*, published in England in 1864 by Charles

Lutwidge Dodgson under the pseudonym of Lewis Carroll (the Latinization of Lutwidge Charles). Dodgson was a Professor of Mathematics at Oxford University and a migraineur. There is speculation that he might have had the syndrome [17,18].

In the first chapter of the book, Alice jumps down a rabbit hole and lands in a hallway where she finds a bottle, which she drinks from, causing her to shrink: "'I must be shutting up like a telescope.' And so it was indeed: she was now only 10 in high …'' Later, she eats a piece of cake that makes her grow (Fig. 2): "'Curiouser and couriouser!' cried Alice…; 'now I'm opening out like the largest telescope that ever was! Good-bye, feet!' (for when she looked down at her feet, they seemed to be almost out of sight, they were getting so far off.)'' Neurologic topics related to another character introduced in the first chapter, the White Rabbit, are beyond the scope of this discussion.

Neuroimaging studies in migraineurs with the syndrome are normal. Patients who have frequent Alice in Wonderland auras may benefit from migraine preventive medications.

Fig. 2. Alice stretched tall. (Illustration by Sir John Tenniel, 1865).

Other rare visual hallucinations, distortions, and illusions that are reported in migraine include the following: zoopsia (visual hallucinations containing complex objects, such as people and animals); achromatopsia (no perception of color); prosopagnosia (inability to recognize faces); visual agnosia (inability to recognize objects); akinetopsia (loss of ability to perceive visual motion); metamorphopsia (distortion of the shapes of objects); micropsia (objects appear too small); macropsia (objects appear too small); teleopsia (objects seem too far away); lilliputianism (people appear too small); multiple images; persistent positive visual phenomena (diffuse small particles, such as TV static or dots, in the entire visual field lasting months to years); palinopsia (the persistence or recurrence of visual images after the exciting stimulus object isn removed); cerebral polyopia (the perception of multiple images); and tilted and upside-down vision [19].

Case 5. My scalp hurts

A 45-year-old woman presents with a 3-month history of a scalp pain. She describes a burning, stinging, itching, and sore pain of the midposterior frontal and anterior parietal scalp in an elliptic distribution extending across both sides with a diameter of approximately 5 cm. The pain is present intermittently daily, lasting hours at a time with an intensity of 5/10. At times, the area is sensitive when she brushes her hair. Ibuprofen may reduce the discomfort. She has seen two dermatologists who found normal skin examinations. There is no history of migraine or other headaches. There is a medical history of hypertension. Neurologic examination was normal with no abnormality of scalp sensation.

MRI of the brain was normal. Blood work was normal, including the following: erythocyte sedimentation rate, 12 mm per hour; rheumatoid factor, 11 IU/mL; antinuclear antibody (ANA) screen, negative; serum protein electrophoresis, normal; serum immunofixation, showing no monoclonal immunoglobin; Sjögren's antibodies A and B, negative; vitamin B_{12} level, 743 pg/mL; and thyrotropin, 1.5. The patient was placed on gabapentin (100 mg by mouth three times a day), which she took as needed with a reduction in the level of pain to a 2/10 [20].

Question

What is the diagnosis?

Discussion

After seeing this patient, I had no idea of the diagnosis. Several hours later, I was reading an article by Pareja and colleagues [21] during lunch and immediately recognized her problem. Nummular headache (a coin-shaped cephalalgia) is a rare, chronic, mild to moderate, pressure-like pain in a rounded or elliptic scalp area (most often the parietal region, in particular its most convex portion, although any region of the head may be affected) of approximately 1 to 6 cm in diameter first described by Pareja

and colleagues in 2002 [22,23]. The location usually is single and unilateral, not changing in size or shape with time, but it can be midline and bilateral as in this case [24]. Typically, the pain is continuous and persists for days to months with exacerbations described as lancinating pains lasting for several seconds or minutes up to a few hours. The affected area may show a variable combination of hypoesthesia, dysesthesia, paresthesia, or tenderness. Spontaneous remissions may occur but the pain usually recurs.

Diagnostic testing, including CT, MRI of the brain, and blood work, is normal. Mild cases typically require no treatment. Patients who have more intense pain might benefit from naproxen or gabapentin. Although the cause is not known, the disorder is benign and might be the result of a localized terminal branch neuralgia of a pericranial nerve.

Case 6. My ear is red, hot, and burning

A 54-year-old white woman was seen who had a 10-year history of episodes of a burning sensation of the left ear. The episodes are preceded by nausea and a hot feeling for approximately 15 seconds and then the left ear becomes visibly red for an average of approximately 1 hour, with a range of approximately 30 minutes to 2 hours. Approximately once every 2 years, she had a flurry of episodes occurring over approximately a 1-month period during which she averaged approximately five episodes, with a range of 1 to 6.

There also was an 18-year history of migraine without aura occurring approximately once a year. At the age of 36, she developed left-sided pulsatile tinnitus. A cerebral arteriogram revealed a proximal left internal carotid artery occlusion of uncertain cause after extensive testing. MRI scan at age 45 was normal. Neurologic examination was normal. A carotid ultrasound study demonstrated complete occlusion of the left internal carotid artery and a normal right [25].

Question

What is the diagnosis?

Discussion

Lance first described the red ear syndrome in 1995 [26] and also proposes the term, auriculoautonomic cephalgia. The disorder is characterized by episodic burning pain, usually in one ear lobe, associated with flushing or reddening of the ear with a duration of 5 minutes to 3 hours in children and adults [27]. In individuals, one ear, alternating ears, or occasionally both ears can be involved in attacks that can occur rarely or up to 4 per day. The redness can occur without pain. Frequent episodes might be reduced with preventive use of gabapentin.

The syndrome can be idiopathic or occur in association with migraine (during or between headache episodes) [27,28], thalamic syndrome, atypical

glossopharyngeal and trigeminal neuralgia, upper cervical spine pathology (cervical arachnoiditis, cervical spondylosis, traction injury, Chiari malformation, or herpes zoster of the upper cervical roots), and dysfunction of the temporomandibular joint [29–31]. Lance postulates that the cause might be an antidromic discharge of nerve impulses in the third cervical root and greater auricular nerve in response to some local pain-producing lesion in the upper neck or trigeminal areas of innervation. In this case, the red ear syndrome probably is associated with migraine and the carotid occlusion an incidental finding.

Case 7. My mouth is burning

A 49-year-old woman was referred by her primary care physician with a 1.5-year history of daily constant burning or numbness of the entirety of her tongue and the back of her throat. She also complains that the inside of her mouth is sensitive. She has had a dry mouth for the past year. She had seen an ear, nose, and throat physician, gastroenterologist, and dentist. Artificial saliva has not been helpful. She has tried a variety of pain pills without any help. She tried Mycostatin at the onset without any benefit. She has been treated with triamcinolone dental paste without any benefit. She does not have any dentures.

There is a medical history of hyperlipidemia on colesevelam (Welchol) and mild depression on buproprion (Wellbutrin). Oropharyngeal and neurologic examinations were normal. Serum zinc, ferritin, and vitamin B_{12} levels were normal. Complete blood count and glycosylated hemoglobin was normal. Sjögren antibodies were negative [32].

Questions

What is the diagnosis? Which treatments are available?

Discussion

Burning mouth syndrome is characterized by a burning, tingling, hot, scalded, or numb sensation in the oral cavity in patients who have a clinically normal oral mucosal examination [33]. Synonyms include glossodynia, glossopyrosis, glossalgia, stomatodynia, stomatopyrosis, sore tongue and mouth, burning tongue, oral or lingual paresthesia, and oral dysesthesia. This pain occurs most commonly on the anterior two thirds and tip of the tongue but also may occur on the upper alveolar region, palate, lips, and lower alveolar region. Less commonly, the buccal muscosa, floor of the mouth, and the throat are affected. The pain may be constant or absent in the morning and progress during the day or be intermittent with symptom-free intervals. The prevalence in the general population is 3.7% with a 7:1 female-to-male ratio, usually in a middle-aged and elderly population, with a mean age of 60 years. Burning mouth syndrome, thus, is not an uncommon disorder but is one that may be uncommonly seen and recognized by neurologists.

The diagnosis is one of exclusion. Although approximately one third may have a psychiatric disorder, often depression, anxiety, or other causes should be considered. The following are causes: xerostomia or dry mouth, which can be the result of medications, such as tricyclic antidepressants, or systemic disease, such as Sjögren's; nutritional deficiency, such as iron, vitamin B_{12}, zinc, or B-complex vitamins; a trigeminal small fiber neuropathy [34]; allergic contact dermatitis resulting from food and oral preparation, which may be detected by patch testing; denture-related etiology; parafunctional behavior, such as clenching or grind the teeth, thrusting the tongue, or running the tongue along the teeth. Candidiasis may be a cause in up to 30% of cases and can be present with a normal examination; diabetes mellitus may be present in 5% of cases; and angiotensin-converting enzyme inhibitors (eg, enalapril, captropril, and lisinopril) can be a cause.

If an underlying cause cannot be found and treated, treatments that might be tried include empiric anticandidal agents, B-complex vitamins, tricyclic antidepressants, gabapentin, oral clonazepam, and topical clonazepam (sucking a 1-mg tablet for 3 minutes and then spitting it out 3 times a day) [35,36]. Women who are postmenopausal might benefit from estrogen-progesterone replacement therapy [37].

Case 8. Headache triggered by straining, stooping, or getting up from a sitting position

This is a 66-year-old white man who has had occasional mild headaches in the past. He presented with a 4-month history of headaches occurring 1 to 4 times per day, brought on by having a bowel movement, stooping, or getting up from a sitting position. He did not know if the headaches were triggered by coughing, because he had not coughed at all. The headaches were a bifrontal and bitemporal sharp, aching pain with a 7/10 intensity and occasionally a 9/10 intensity, with an average duration of 1 minute and a range of 30 seconds to 1 hour. Approximately 20 of the headaches had lasted more than 1 minute, with most in a range of 1 to 2 minutes. He had tried ibuprofen and acetaminophen with questionable help. For the prior 5 days, he had increased his dose of aspirin from 81 mg per day to 325 mg per day. The headache then was different with a constant bifrontotemporal pressure with an intensity of 1/10 but he had not had the brief headaches with activity exacerbation. He had a CT scan of the sinuses on June 14, 2004, with essentially negative findings.

There was a medical history of insulin-dependent diabetes with sensory neuropathy and hypertension. Neurologic examination was normal except for diminished pinprick distally of both lower extremities and absent deep tendon reflexes diffusely.

MRI scan of the brain was normal except for smooth diffuse dural enhancement around both cerebral convexities and, to a milder degree, in the posterior fossa. A lumbar puncture produced an opening pressure of 11 cm of water. Cerebrospinal fluid (CSF) analysis revealed 0 white blood

cells, 1 red blood cell/μL, a glucose of 101 mg/dL (with a serum glucose of 156 mg/dL), and a protein of 109 mg/dL. The VDRL was nonreactive. An erythrocyte sedimentation rate was [36]. ANA, rheumatoid arthritis (RA) factor, Sjögren's antibodies, Lyme antibodies, and angiotensin-converting enzyme level were negative or normal. MRI scan of the cervical, thoracic, and lumbar spine revealed degenerative changes but no evidence of extra-arachnoid fluid collections, extradural extravasation of fluid, or meningeal diverticula.

Questions

What is the diagnosis? What are the MRI findings in this disorder?

Discussion

The history could be compatible with primary cough headache as defined by the International Headache Society's second edition criteria: sudden onset, lasting from 1 second to 30 minutes, and brought on by and occurring only in association with coughing, straining, or Valsalva's maneuver [38]. Primary cough headache, however, is a diagnosis of exclusion, where the symptoms cannot be attributed to another disorder. Primary cough headache usually is bilateral and affects predominantly patients older than 40. In some cases, the onset may be after a respiratory infection with cough. The term, cough headache, also is used by many to include headaches brought on by sneezing, weightlifting, bending, stooping, or straining with a bowel movement. Weightlifting also can cause an acute bilateral nuchal-occipital or nuchal-occipital-parietal headache that can persist as a residual ache for days or weeks, which may be the result of stretching of cervical ligaments and tendons. Other secondary causes should be excluded as appropriate, such as subarachnoid hemorrhage. Although primary cough headache is associated with an increase in intracranial pressure, the exact cause of the pain is not certain. Posterior cranial fossa overcrowding may be a contributing factor [8].

Primary cough headache may be diagnosed only after structural lesions are excluded, such as posterior fossa tumor, Chiari I malformation, platybasia, basilar impression, spontaneous intracranial hypotension (SIH), pneumocephalus, middle cranial fossa or posterior fossa meningioma, medulloblastoma, pinealoma, chromophobe adenoma, midbrain cyst, and subdural hematoma (excluded by neuroimaging) [39]. Internal carotid artery stenosis and unruptured intracranial saccular aneurysms are questionable associations with unilateral cough headache. MRI of the brain without and with contrast is the preferred imaging study to exclude secondary causes.

Benign cough headaches (and less often secondary cases) may respond to indomethacin (25–50 mg 3 times a day) [9], lumbar puncture [10], methysergide [11], acetazolamide (500–2000 mg per day in divided doses) [12], and perhaps topiramate (because of its weak carbonic anhydrase inhibition). Some patients may have an abrupt recovery after extraction of abscessed teeth.

SIH syndrome often presents with a headache when upright, which is relieved by lying down, or an orthostatic headache. As SIH persists, however, chronic daily headaches may be present without orthostatic features. SIH also may present with other types of headaches, including exertional without any orthostatic features, acute thunderclap onset, parodoxic orthostatic headaches (present in recumbency and relieved when upright), intermittent headaches resulting from intermittent leaks, and the acephalgic form (no headaches). Neck or interscapular pain may precede the onset of headaches in some cases by days or weeks.

Box 1 summarizes MRI abnormalities of the brain and spine that are variably present. SIH can be present with a normal MRI with contrast of the brain and spine. MRI scan of the brain may reveal diffuse pachymeningeal (dural) enhancement with gadolinium without leptomeningeal (arachnoid

Box 1. MRI abnormalities in cerebrospinal fluid leaks

Head MRI

Diffuse pachymeningeal (dural) enhancement
Descent (''sagging'' or ''sinking'') of the brain
- Descent of cerebellar tonsils (may mimic type I Chiari)
- Obliteration of some of the subarachnoid cisterns (ie, prepontine or perichiasmatic cisterns)
- Crowding of the posterior fossa

Enlargement of the pituitary
Flattening or ''tenting'' of the optic chiasm
Subdural fluid collections (typically hygromas, infrequently hematomas)
Engorged cerebral venous sinuses
Decrease in size of the ventricles (ventricular collapse)
Increase in anteroposterior diameter of the brainstem

Spine MRI

Extra-arachnoid fluid collections (often extending across several levels)
Extradural extravasation of fluid (extending to paraspinal soft tissues)
Meningeal diverticula
Identification of level of the leak (not uncommonly)
Identification of the actual site of the leak (very uncommonly)
Spinal pachymeningeal enhancement
Engorgement of spinal epidural venous plexus

From Mokri B. Low cerebrospinal fluid pressure syndromes. Neurol Clin North Am 2004;22:55–74; with permission.

and pial) involvement and, in some cases, subdural fluid collections, which return to normal with resolution of the headache. One is reversible descent of the cerebellar tonsils below the foramen magnum (acquired Chiari I malformation), which can be the result of SIH and also the result of lumbar puncture and overdraining CSF shunts. The diffuse meningeal enhancement on MRI may be explained by dural vasodilation and a greater concentration of gadolinium in the dural microvasculature and in the interstitial fluid of the dura. (Before the characteristic picture of the postural headache and diffuse pachymeningeal enhancement on MRI was recognized, some patients underwent extensive testing, including meningeal biopsy, to exclude other conditions, such as meningeal carcinomatosis and neurosarcoidosis.) The pleocytosis and elevated protein in the CSF and the subdural fluid collections probably are the result of decreased CSF volume and hydrostatic pressure changes resulting in meningeal vasodilation and vascular leak.

A lumbar puncture usually demonstrates an opening pressure from 0 to 70 cm H_2O (and can even be negative), although the pressure can be in the normal range, especially if the procedure is performed after a period of bed rest. The CSF analysis may be normal or can demonstrate a moderate, primarily lymphocytic pleocytosis (50 cells/mm^3 are common and values may be as high as 220 cells/mm^3), the presence of red blood cells, and elevated protein levels that rarely are as high as 1000 mg/dL. CSF glucose concentration never is low.

CT myelography is more sensitive than other studies for determining the actual site of a CSF leak, because most leaks occur in the spine, especially at the thoracic level. Because the leaks can be high or low flow, early and delayed CT may be helpful. The study may demonstrate extra-arachnoid fluid, meningeal diverticula, and extradural leak of contrast into the paraspinal soft tissues.

Radioisotope cisternography using indium 111 may demonstrate an absence or paucity of activity over the cerebral convexities at 24 or 48 hours. Less commonly, parathecal activity at the approximate level of the leak may be apparent.

Case 9. Headache with walking

Grace and colleagues report the following case:

> A 59-year-old man presented to the hospital neurosurgical department with a history of severe headache experienced in all of the vertex and upper occipital area on exercise. Before this presentation there was no history of significant or persistent headache. The pain appeared immediately on walking at a normal pace and immediately was relieved on rest. It was described as bursting in quality and severe in degree. There was no history of chest pain.
>
> Resting electrocardiograph (ECG) revealed T-wave inversion in the anterolateral leads. Treadmill exercise immediately induced the same headache

in the same location. The headache was severe and associated with simultaneous further S-T depression on the ECG. The headache and the fresh S-T segment abnormality resolved immediately with rest. Coronary angiography revealed two areas of subtotal occlusion at the midpoint of the right coronary. There also was 70% stenosis of the left anterior descending coronary artery (LAD) proximal to the diagonal branch [40].

He underwent aortocoronary bypass surgery and was headache-free for 7 years. The same exertional headache recurred with resolution following angioplasty of a high-grade stenosis of the LAD. One year later, an exertional headache recurred that did not resolve despite repeat bypass surgery and was believed the result of refractory cardiac ischemia.

Questions

Which types of headaches are associated with exertion? What is the diagnosis? What is the mechanism for the referred head pain?

Discussion

Primary (benign) exertional headache, which has a lifetime prevalence of 1% [41], is brought on by and occurs only during or after physical exercise and lasts from 5 minutes to 48 hours [39]. It typically is a throbbing bilateral headache and not attributed to another cause. Reported activities include running, rowing, tennis, and swimming. One particular activity may precipitate the headaches in some individuals but not others. This headache type is prevented by avoiding excessive exertion, particularly in hot weather or at high altitude. Exercise can be a trigger for a typical migraine for some migraineurs. Secondary causes to be excluded include subarachnoid hemorrhage (SAH), pheochromocytomas, cardiac ischemia, middle cerebral artery dissection, paranasal sinusitis, intracranial neoplasms, colloid cysts of the third ventricle, and hypoplasia of the aortic arch after successful coarctation repair. An MRI of the brain with magnetic resonance angiography assists in ruling out structural or vascular lesions.

In some cases, exertional headaches may be prevented by a warm-up period. Some patients choose to avoid the particular activity. Indomethacin (25–150 mg per day) may work as a preventive, taken minutes to 1 hour before exertion. Prophylactic drugs used for migraine, such as β-blockers, may be effective for some patients.

Cardiac ischemia rarely may cause a unilateral or bilateral headache in any part of the head brought on by exercise and relieved by rest, cardiac cephalalgia, or anginal headache [42,43]. Headaches rarely may occur alone or be accompanied by chest pain. In cases of unstable angina, headaches may occur at rest [44].

Angina generally is believed the result of afferent impulses that traverse cervicothoracic sympathetic ganglia, enter the spinal cord via the first and the fifth thoracic dorsal roots, and produce the characteristic pain in the

chest or inner aspects of the arms. Cardiac vagal afferents, which mediate anginal pain in a minority of patients, join the tractus solitarius. Although the cause is not known, a potential pathway for referral of cardiac pain to the head would be convergence with craniovascular afferents [45]. Two other possible mechanisms of headache are suggested [42]. A reduction of cardiac output and an increase in right atrial pressure occur in myocardial ischemia. The associated reduction in venous return may increase intracranial pressure, which could produce headache. Second, release of chemical mediators resulting from myocardial ischemia (serotonin, bradykinin, histamine, and substance P) may stimulate nociceptive intracranial receptors and produce headache.

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