Oxford Textbook of Headache Syndromes

Michel Ferrari, Andrew Charles, David Dodick, Fumihiko Sakai, and Joost Haan

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Author(s): Randolph W. Evans

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Introduction

Alice in Wonderland syndrome, blip syndrome, cardiac cephalalgia, epicrania fugax, exploding head syndrome, Harlequin syndrome, lacrimal headache, neck–tongue syndrome, and red ear syndrome are among the fascinating array of rare headache disorders. James W. Lance, Juan A. Pareja, and colleagues have first described or named six of them. Certainly, there are patients with additional rare headaches just waiting to be described by astute observers.

Some rare headache disorders, including Alice in Wonderland syndrome, blip syndrome, cardiac cephalalgia, epicrania fugax, exploding head syndrome, Harlequin syndrome, lacrimal neuralgia, neck–tongue syndrome, and red ear syndrome

In case of a rare headache disorder, an appropriate diagnosis and treatment can be most reassuring to the patient.

Alice in Wonderland syndrome

History

In 1952, Caro W. Lippman described seven migraineurs who had unusual distortions of body image (1). The descriptions are illustrative: ‘Occasionally the patient has an attack where she feels small, about 1 foot 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.’ One of the patients who felt short and wide while she walked referred to her abnormal sensation as her Tweedledum or Tweedledee feeling (two characters from Lewis Carroll’s 1871 book, *Through the Looking-Glass, and What Alice Found There* (2)). Lippman concluded, ‘*Alice in Wonderland* [full title, *Alice’s Adventures in Wonderland* (3)] contains a record of these and many other similar hallucinations. Lewis Carroll (Charles Lutwidge Dodgson), who wrote “Alice,” was himself a sufferer from classic migraine headaches.’

In 1955, Todd, in giving the syndrome its name, presented six new cases and described a syndrome of distortions of the size, mass, or shape of the patient’s own body or its position in space often associated with depersonalization and derealization (4). Distortions in the perceived passage of time were also described in some patients. Todd discussed the many causes in addition to migraine. Since then, many authors have used Alice in Wonderland syndrome (AIWS) for the visual illusions and distortions of how others appear rather than illusions of one’s own body as in Todd’s original description.

Alice’s Adventures in Wonderland was published in England in 1864 by 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 (5,6,7).

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 (Figure **28.1**): ‘ “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.)’

Figure 28.1.
Alice stretched tall.

Illustration by Sir John Tenniel, 1865.

Clinical features and aetiology

AIWS syndrome is a rare migraine aura usually 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 (8,9,10). The syndrome can occur at any age but is more common in children. A 1-year prospective observational study of young people aged 8–18 years found that AIWS can occur before the onset of headaches, may go unrecognized, and may be more common than previously realized (11). The symptoms are attributed to the non-dominant posterior parietal lobule.

In a review of 81 cases, the cases were categorized as somaesthetic (n = 7; 9%), visual (n = 61; 75%), or both (n = 13; 16%) (12). Epstein–Barr virus infection was commonly identified (n = 39; 48%) followed by migraine (n = 11; 14%). Other possible associations included other infectious disorders (varicella, H1N1 influenza, coxsackievirusB1, scarlet fever, typhoid fever, and, not included in the review, an association with Lyme neuroborreliosis (13)), acute Zika virus infection (14), and mycoplasma infection (15), toxic encephalopathy, major depression, epileptic seizures, medications (cough syrup with dihydrocodeine phosphate and DL-methylephedrine hydrochloride (16), topiramate, and aripiprazole (17)), a right medial temporal lobe stroke, and a right

temporo-parietal cavernoma (18). Six of the migraine cases were somaesthetic and two had somaesthetic and visual symptoms.

In a series of 20 paediatric cases, the average age was 9.5 ± 3.8 years (range 4–16 years) (19). Ninety per cent had micropsias and/or macropsias, 85% distortion of the form of the objects, 80% displacement of objects, 45% disturbances of body image, 45% acceleration of time, and 30% a sensation of unreality. Ninety-five per cent of the children had many episodes a day; these episodes lasted less than 3 minutes in 90% of them. Neuroimaging was normal in all cases. Migraine was considered the cause in eight and Epstein–Barr virus infection in five. The majority of cases had spontaneous resolution without recurrence. Another paediatric series of nine children followed-up for a mean of 4.6 years also showed only occasional recurrence, in two (20). There is a case report of a 15-year-old female with AIWS following acute Zika virus infection (14).

There is a single case report of a 17-year-old male with a history of abdominal migraine since the age of 10 years who developed AIWS (21). All of his symptoms improved after treatment with valproate.

Topiramate has been reported as causing AIWS 1 week after starting 25 mg daily (titrating up to 100 mg daily over 4 weeks) in a 31-year-old woman with chronic migraine (22,23). She described episodes of her entire body feeling either too big or too small and everything else either too small or too big (or two episodes of feeling too big and then too small) all lasting 5–10 minutes, followed by a mild headache behind the eyes lasting 30–45 minutes without medication. The symptoms resolved 1 month after stopping topiramate.

A 17-year-old female with episodic migraine developed distortions of body image where her head would grow bigger and the rest of her body would shrink or her hand would increase in size and become heavier while the rest of the arm would become smaller only when she did not directly fall asleep after taking topiramate 75 mg at bedtime (24). The symptoms resolved on 50 mg at bedtime and reappeared on 75 mg at bedtime, and then ceased when she stopped the drug except for one episode 3 months later. An electroencephalogram and magnetic resonance imaging (MRI) of the brain were normal.

Routine neuroimaging studies in migraineurs with the syndrome are normal. Not surprisingly, there are no placebo-controlled studies on treatment of what appears to be a rare and self-limited migraine variant.

Prognosis

Of the 15 patients with follow-up with AIWS seen by a paediatric neuro-ophthalmologist over 20 years, 20% had a few more events, which eventually stopped after the initial diagnosis, 40% had no more events, and 40% were still having symptoms at follow-up (25). Twenty-seven per

cent developed migraines and 7% developed seizures following the diagnosis of AIWS.

In a follow-up study over 30 years of 28 patients with paediatric migraine precursors, more than one-quarter still experienced distortions of time and nearly 20% still reported distortions of space (26).

Blip syndrome

History

Lance had unusual sensations himself for about 6 months, which he recognized were not associated with his cardiac extrasystoles, which he named ‘blip’ syndrome (27). He then reported 12 additional cases (including eight women; three physicians) ranging in age from 33 to 75 years with symptoms present for periods of 2 months up to 5 years, with a typical frequency of episodes of 1–4 per month (five had two or more per day and one had 12–15 per day on some occasions) with each lasting a split second up to 2 seconds (28). Interestingly, four of 12 were migraineurs and none had seizure disorders.

Clinical features

Patients described sensations including ‘a short circuit in the brain’, their mind ‘going blank for a second’ with a pressure in the forehead and a ‘feeling of losing balance’, ‘impending loss of consciousness’, and ‘a wave going through’. Testing on some of the patients, including electroencephalograms (EEGs), computed tom scans, electrocardiograms, and carotid ultrasounds were normal.

Case report

In 2013, I evaluated a 47-year-old man with a history of ‘little small short circuits’, which had been occurring for 7 months and which usually occurred 2–3 times per day up to 10 times per hour. He described a blip in his head that he did not see or feel but sensed for a half a second. The symptom could occur when sitting, standing, or walking but not when lying down. He had no alteration of consciousness, vertigo, paraesthesias, weakness, trouble speaking, associated headache, or other neurological symptom except for a brief feeling of slight imbalance.

There was a history of episodic migraine without aura since childhood and hyperlipidaemia. Neurological examination was normal.

He had seen another neurologist. MRI of the brain with and without contrast and magnetic resonance angiography of the brain were negative. Evaluation by an ear-nose-and-throat physician, including audiogram and electronystagmography, were normal. EEG was normal. Cardiac evaluation was normal.

Aetiology

The aetiology of the episodes is not certain. Lance compared blip syndrome to other benign disorders such as déjà vu, night starts, and exploding head syndrome (see ‘Exploding head syndrome’).

Cardiac cephalalgia

Clinical features

Cardiac ischaemia may rarely cause a unilateral or bilateral headache brought on by exercise and relieved by rest. This is called ‘cardiac cephalalgia’, or ‘anginal headache’ (29,30,31). Headaches may occur alone or be accompanied by chest pain. In cases of unstable angina, headaches may also occur at rest (32). Even a thunderclap headache may accompany the chest pain (33).

Thirty-six well-documented cases of cardiac cephalalgia were reported in the literature up to 2013: 58.3% were males, usually over the age of 50 years (but 22% were younger than 50—the youngest 35) (34). Anti-anginal medications (nitrates) caused headaches in 56% of cases (35). Thirty per cent had associated symptoms such as photophobia, phonophobia, osmophobia, and nausea (36). Chest pain, pain in the left arm, sometimes radiating to the mandible or epigastric region, was present in 50% of cases. Cardiac cephalalgia was the only manifestation of angina in 27% of cases. Five more cases have been subsequently reported (37).

Cardiac cephalalgia should be distinguished from migraine as the use of triptans or dihydroergotamine, in general, is contraindicated in cardiac ischaemia and might even be harmful. Appropriate cardiac testing will make the diagnosis once suspected. The headaches resolve with revascularization or conservative treatment.

Migrainous thoracalgia, a diagnosis of exclusion, is a migraine accompanied by an aura of chest pain and arm paraesthesias which can occur with or with headache (38).

Pathophysiology

Angina is generally believed to be 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 (39). Two other possible mechanisms of headache have been suggested (25): (i) a reduction of cardiac output and an increase in right

atrial pressure during myocardial ischaemia can be associated with reduction in venous return, which increases intracranial pressure producing headache; and (ii) release of chemical mediators resulting from myocardial ischaemia (serotonin, bradykinin, histamine, and substance P) may stimulate nociceptive intracranial receptors and produce headache.

Epicrania fugax

History

Pareja et al. (40) described 10 patients with a novel syndrome in 2008, which they named ‘epicrania fugax’, with over 100 cases reported (41,42, 43). Eight cases are associated with nummular headache (44).

Clinical features

Epicrania fugax is characterized by paroxysmal pain through the surface of one side of the head in a linear or zig-zag trajectory, which may move forward or backward and is not in the distribution of one single nerve. So the pain may go between the posterior scalp and the ipsilateral forehead, eye, and nose. The pain, which typically has an electrical quality, is of moderate or severe intensity and lasts one to a few seconds. The frequency ranges from a few attacks per year or less to numerous attacks per day. There may be associated ipsilateral cranial autonomic symptoms such as conjunctival injection, lacrimation, or rhinorrhoea. The pain may shift sides. Between attacks, some patients have a persistent mild pain or tenderness in the area where the pain originates.

Neurological examination was normal in all patients except for local hypersensitivity at the area where the pain originates in a few patients. Diagnostic testing, including neuroimaging and erythrocyte sedimentation rates, was normal.

Five patients have been reported with similar pain starting in the lower face (V2 or V3) and radiating upwards with a linear trajectory of moderate-to-severe intensity with a stabbing or electrical quality lasting one to a few seconds (45).

Management

Gabapentin and lamotrigine have been reported as providing partial or complete relief for some patients. A few patients have benefited from pregabalin, levetiracetam, and carbamazepine. Amitriptyline, indomethacin, occipital nerve blocks, and trochlear injections have also been occasionally effective (33,34,46).

Exploding head syndrome

History

Exploding head syndrome (EHS) was first named by John M.S. Pearce in 1988 when he reported on 10 patients (47). Robert Armstrong-Jones provided the first description as ‘snapping of the brain’ in 1920 (48), although Silas Weir Mitchell may have previously described the disorder in 1890 (49).

Clinical features

EHS is characterized by a momentary loud noise that patients usually experience during the early stages of sleep (50). Patients describe a sudden onset of ‘an explosion in the head’, enormous roar, bomb-like explosion, or lightning crack that awakens them from sleep. This is usually followed by a feeling of intense fear, terror and/or palpitations. However, there is no headache or pain associated with the noise.

Symptoms can arise from any stage of sleep, but primarily during stages 1 and 2 (51). One study of nine patients indicated that symptoms correlated with an alert state or awakening on polysomnographic recordings (52). Attacks can occasionally occur as patients are awakening following arousal and onset back to stage 1 sleep. The frequency is highly variable with a range of 2–4 attacks followed by prolonged or lifetime remission to seven attacks nightly for several nightly each week.

Fear, terror, palpitations, or a forceful heartbeat were reported as occurring after the loud noise in 47/50 patients (38). Ten per cent of patients described an associated flash of light and 6% reported a curious sensation as if they had stopped breathing and had to make a deliberate effort to breathe again—‘an uncomfortable gasp’ (38). Occasionally, brief myoclonic jerks of the extremities or the entire body may follow (53). Psychological stress and being tired may be triggers (38,41). Three patients of 50 reported a positive family history (38).

EHS may be a migraine aura. Kallweit et al. (54) reported a 54-year-old man with attacks of EHS followed by an exacerbation of his chronic migraine after each attack. Evans (55) reported a 26-year-old woman with a history of migraine without aura with multiple episodes of EHS followed by brief sleep paralysis and then one of her typical migraine headaches. The exact cause of EHS, however, is unknown (56). Rossi et al. (57) reported a middle-aged man with EHS as an aura symptoms of migraine with brainstem aura.

Epidemiology

EHS can occur at any age, but is more common in patients older than 50 years of age, with a median age of 54 years (range 12–84 years) and a female-to-male ratio of 3:2 (58,59). The prevalence is unknown as, anecdotally, patients may not report their symptoms. No large-scale prevalence studies have been performed and EHS has been believed to be rare. However, in a study of 211 US college students, 18% had a lifetime

prevalence and 16.6% had recurrent cases, not more common in females (60).

Evaluation

The diagnosis is made by the clinical history. A sleep study does not assist in diagnosis and it is not certain whether the study changes management if the patient is found to have obstructive sleep apnoea (see 'Management') (51). The neurological exam is normal. Imaging studies are not necessary, although some patients may wish to be reassured that they do not have a tumour or aneurysm.

Management

After explanation and reassurance, most patients do not require medication. For those with frequent or disturbing symptoms, there are anecdotal reports of benefit of treatment with clomipramine (41), nifedipine (61), flunarizine (45), topiramate (62), amitriptyline (58), and the use of an oral appliance for a patient with obstructive sleep apnoea (48).

Harlequin syndrome

History

Harlequin syndrome was first described by Lance and colleagues in 1988 (63). The syndrome is named after Arleccino (Harlequin) who was a character in the travelling improvisational theatre, which originated in Venice in the sixteenth century, Comedia Dell'Arte (64). Members wore Harlequin masks with blackening of one side (Figure **28.2**), which was a similar appearance of the sweating half of the face that was demonstrated with application of alizarin powder (Figure **28.3**).

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Figure 28.2
Harlequin mask.

Reproduced from *Practical Neurology*, 5, Lance JW, Harlequin syndrome, pp. 176–177. Copyright (2005) BMJ Publishing Group Ltd.

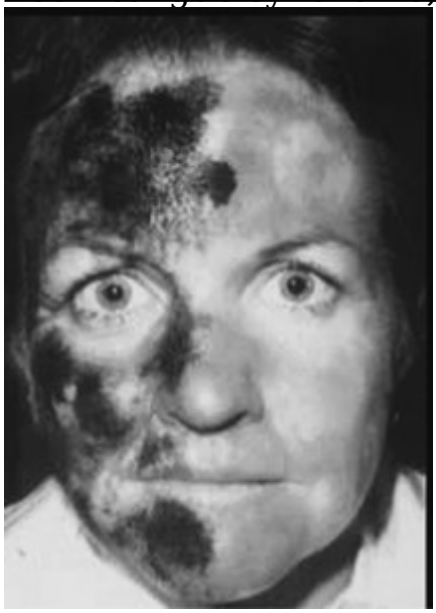

Figure 28.3
Harlequin syndrome.

Sweating on the right half of the face delineated by the application of alizarin powder.

Reproduced from *Journal of Neurology, Neurosurgery & Psychiatry*, 51, Lance JW, Drummond PD, Gandevia SC, Morris JG, Harlequin syndrome: the sudden onset of unilateral flushing and sweating, pp. 635–642. Copyright (1988) by permission of BMJ Publishing Group Ltd. DOI: 10.1136/pgmj.2009.080473.

Clinical features

Harlequin syndrome presents with unilateral erythema or redness and hyperhidrosis of the face and, less commonly, the ipsilateral arm and upper chest (65), and is believed to be due to a normal or exaggerated response to the contralateral interruption of the sympathetic nerve fibres, resulting in a vasomotor deficit of the ipsilateral side and often an exaggerated vasodilatory response on the contralateral side during thermal (exposure to heat or exercise) or emotional stimulation. There is one case report where the leg was also involved (66).

Aetiology

Abnormalities should be excluded at the level of the first or several thoracic roots such as mediastinal and pulmonary masses (67). Harlequin syndrome has also been reported as a sequelae of internal jugular catheterization, peri-operative local anaesthesia, sympathectomy to treat severe hyperhidrosis (68), toxic goitre (69), spontaneous cervical carotid

artery dissection (70), and implantation of intrathecal pumps (71), obstetric epidural anaesthesia (72), upper lobectomy (73), excision of a neck schwannoma (74), and a neuroblastoma (75). In most cases, no cause is found.

Management

Usually no treatment is required and patients are reassured by an explanation of a cause. A contralateral thoracic sympathectomy could be performed to restore symmetry if desired by the patient (54), although most patients do not choose any treatment.

Lacrimal neuralgia

Anatomy

The ophthalmic division of the trigeminal nerve divides into the frontal nerve (which divides into the supratrochlear and supraorbital nerves), the nasociliary nerve, and the lacrimal nerve. The lacrimal nerve runs along the upper border of the lateral rectus muscle in the orbit and splits into two branches, the lateral branch (which supplies the lacrimal gland) and the medial branch (sensory innervation to the lateral aspect of the upper eyelid and adjacent area of the temple) (Figure **28.4**).

Figure 28.4
Skin area supplied by the left lacrimal nerve (shaded area).

The lacrimal nerve gives sensory innervation to the lateral upper eyelid and a small cutaneous area adjacent to the external canthus.

History

Pareja and Cuadrado reported the first two cases of lacrimal neuralgia in 2013 (76).

Patient 1

A 66-year-old woman had a history of constant moderate-to-severe sore and burning pain, which was worse with lateral eye movements, in the lateral aspect of her left superior eyelid and adjacent area of the temple with onset at age 64 years, without prior trauma or other relevant disease. Neurological examination showed decreased sensation in the area supplied by the left lacrimal nerve and tenderness on palpation between the globe and the external edge of the left orbit. Ophthalmological examination was normal.

MRI of the brain and orbits and blood tests, including thyroid tests, an erythrocyte sedimentation rate, and immunological screening, were normal. Her emotional state was not affected on testing. A left lacrimal nerve block with 2% lidocaine produced complete improvement for around 4 hours. She had absolute relief with pregabalin 150 mg daily. After 9 months, pregabalin was stopped. The same symptoms recurred after 4 months, and she had the same absolute relief when pregabalin was resumed.

Patient 2

A 33-year-old woman had a history of constant moderate-to-severe pressure or stabbing pain in a small area adjacent to the lateral canthus of her left eye since the age of 25 years, with an unremarkable medical history. The symptomatic area was tender to light touch. Combing her hair on the left side or chewing could occasionally trigger paroxysmal exacerbations.

Neurological examination showed superficial hypoaesthesia, hyperaesthesia, and allodynia in the left lacrimal nerve distribution. The supero-external angle of the left orbit was hypersensitive to palpation. Ophthalmological and psychiatric evaluations were normal.

A MRI of the brain and orbits was normal. Blood tests, including thyroid tests, an erythrocyte sedimentation rate, and immunological screening, were normal. A lumbar puncture was normal. Four lacrimal nerve blocks with 2% lidocaine resulted in complete relief lasting up to 6 hours. Oral indomethacin, ibuprofen, gabapentin, flunarizine, carbamazepine, oxcarbazepine, topiramate, amitriptyline, duloxetine, mirtazapine and tramadol did not help. Lidocaine patches and capsaicin cream in the symptomatic area were of no benefit. Pulsed radiofrequency of the lacrimal nerve, the Gasserian ganglion, the sphenopalatine ganglion and the ophthalmic nerve did not help. Pregabalin 400 mg daily provided partial but substantial relief.

Three additional cases with negative testing have responded to lacrimal nerve blocks (77).

A secondary case of lacrimal neuralgia has been reported in a woman who developed similar pain attacks lasting 1–2 minutes after left cataract surgery, which was relieved by an anaesthetic block at the emergence of the lacrimal nerve (78), and another in a 53-year-old man with attacks triggered by argon laser photodynamic therapy and intravitreal injection of aflibercept relieved by lacrimal blocks (79).

Summary

Lacrimal neuralgia is a newly reported cause of orbital and peri-orbital pain which, so far, seems to only be partially or completely responsive to pregabalin or lacrimal nerve blocks. The features and response to treatment of future cases will be of interest.

Neck–tongue syndrome

History

James Cyriax reported two cases of neck–tongue syndrome in 1962 (80). Lance and Anthony named the syndrome when they reported four additional cases in 1980 (81). This is a rare disorder with a prevalence in the Vågå study of 0.2% (82) and over 50 cases reported in the literature· (80,83,84,85).

Clinical features

Neck–tongue syndrome is characterized by acute, unilateral occipital pain lasting a few seconds to 1 minute and numbness of the ipsilateral tongue lasting seconds to 5 minutes precipitated by sudden movement, usually rotation, of the neck to either side. Less frequently, dysarthria, dysphagia, tongue paralysis, or tongue movements may occur (86,87,88,89,90). Intermittent and then constant tongue paraesthesias have been reported (82). Rarely, symptoms may switch sides (78).

Pathophysiology

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 fibres from the tongue originating from the lingual nerve to the hypoglossal nerve to the C2 root (Figure **28.5**) (76, 91,92). Contraction of the accessory atlantoaxial ligament (Arnold’s ligament) during rotation might irritate the second cervical nerve root, as well as the hypoglossal nerve at its exit from the foramen magnum in some cases (93).

Figure 28.5
Lateral view of the right atlantoaxial joint; the atlas has rotated to the right.

The small arrow shows the inferior articular process impinging the C2 spinal nerve and ventral ramus.

Reproduced from *Headache*, 40, Evans RW, Lance JW, Transient headache with numbness of half the tongue, pp. 692–693. Copyright (2000) with permission from John Wiley and Sons.

Aetiology

Neck–tongue syndrome can be idiopathic without obvious abnormalities. A benign, familial form of neck–tongue syndrome is described without anatomical abnormality, which resolves spontaneously during adolescence. About 25% of cases have pathology of the occipito-atlantoaxial joints. Secondary causes of neck–tongue syndrome include head and neck trauma, Chiari 1 malformation, congenital anomalies of the cervical spine, ankylosing spondylitis, degenerative spondylosis, rheumatoid arthritis, tuberculous atlantoaxial osteoarthritis, and cervical acute transverse myelopathy (94,95,96,97). A prolonged slouching sitting posture has been proposed as a cause (98).

Management

The most effective treatment is not known (29). Non-steroidal anti-inflammatory drugs, muscle relaxants, medications for neuropathic pain (amitriptyline, gabapentin, and carbamazepine), and steroids have been reported to be helpful in single cases. Other treatments reported include

cervical collars, analgesics, manipulation, injections of local anaesthetic, nerve resection, and cervical fusion.

Red ear syndrome

History and clinical features

Since Lance first described red ear syndrome in 1994 (99), more than 80 cases have been reported in children and adults (100,101,102,103). 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 seconds to hours (constant in two cases). The average age for idiopathic cases is 35 years with 62% females and for secondary cases 50 years with 70% females.

In individuals, one ear, alternating ears, or occasionally both ears simultaneously can be involved in attacks that can occur rarely or up to 20 times daily. The redness can occur without pain. The pain may radiate to the cheek, forehead, a strip behind or below the mandible, behind the ear, occiput, and the ipsilateral upper posterior neck. Attacks may be spontaneous or precipitated (in 31% of idiopathic cases and 63% of secondary cases) by touching the ear, drinking, coughing, chewing, sneezing, neck movement, exercise, stress, or exposure to heat or cold.

Anatomy

To understand secondary causes, it is helpful to recall the sensory supply of the ear, which includes C2 and C3, and cranial nerves V, VII, IX, and X. The anterosuperior ear lobe is supplied by the auriculotemporal nerve (from V3) and the inferoposterior ear lobe is supplied by the greater auricular nerve (C2 and C3). The blood supply to the ear comes from an anastomosis between branches of the middle temporal and posterior auricular arteries, part of the external carotid circulation innervated by the trigeminal nerve.

Aetiology

RES can be idiopathic or occur in association with migraine (during or between headache episodes), trigeminal autonomic cephalgias, 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.

Pathophysiology

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. Al-Din et al. (104) suggest that primary

and secondary cases may be due to activation of the trigeminal-autonomic reflex.

Management

A variety of treatments have been tried with variable success, including gabapentin, amitriptyline, indomethacin, flunarizine, nimodipine, ibuprofen, and indomethacin (105). Local anaesthestic block or section of the third cervical root might be helpful. Some cases are resistant to treatment.

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