Headache syndromes can be associated with focal neurological symptoms or signs. Good knowledge of primary headaches, a detailed history and a thorough clinical examination are prerequisites for their differential diagnosis. The neurological symptoms produced by the migraine aura are the most characteristic and recognisable. However, structural lesions, such as vascular malformations, can produce similar symptoms to migraine with aura, which highlights that paraclinical investigations are necessary in most patients with headache and focal neurological symptoms. In this review, we provide an overview of the differential diagnosis of the most common headache disorders with focal neurological symptoms or signs to refresh the practising neurologist’s differential diagnostic knowledge for the clinical situation and to aid the teaching of neurology residents.


Headache is a symptom with various causes. In some disorders, headache is associated with focal neurological signs or symptoms. If this happens, one has to distinguish between a primary headache (eg, migraine) and a symptomatic headache secondary to an underlying infectious, inflammatory, vascular, neoplastic, or epileptic disorder. This differential diagnosis may be difficult in some patients. Above all, its prerequisite is a precise knowledge of the clinical spectrum and characteristics of primary headache disorders. Any clinical presentation that deviates even slightly from the classical features of primary headaches warrants a thorough work-up in order to search for a secondary cause of headache. We will focus on the most common disorders that cause headache and focal neurological symptoms or signs (panel 1). In this review, we refer to the latest edition of the Classification and Diagnostic Criteria for Headache Disorders, Cranial Neuralgias and Facial Pain. Our goal is to refresh the practising neurologist’s differential diagnostic knowledge for the clinical situation and to aid the teaching of neurology residents.

The following general rules of thumb (figure 1) are useful when confronted with a headache syndrome in the clinical situation. Long-lasting headaches (eg, over weeks or months) associated with neurological abnormalities are typically caused by structural brain lesions. If headaches are episodic in nature, neurological signs or symptoms may precede or accompany and outlast the headache. The disorder is likely to be benign (ie, not secondary) if the symptoms occur before the headache; and it is likely to be caused by a structural lesion if symptoms appear during and after the headache, or if they outlast it. The rare exception to the latter is hemiplegic migraine.

After migraine with aura, the most common headache disorders associated with focal neurological signs (in this case, ipsilateral autonomic signs) are cluster headache and the related trigeminal autonomic cephalalgias.

Migraine
Migraine is a multifaceted disorder, of which the head pain is only one component. It is a primary CNS disorder, but in some instances it may occur for the first time in close temporal relation to a secondary headache. Migraine is a paroxysmal disorder characterised by attacks, which are separated by symptom-free intervals. Similar to epilepsy, migraine is characterised by the repetition of attacks rather

Panel 1. Summary of the disorders described in this review and their classification in the International Classification of Headache Disorders

<table>
<thead>
<tr>
<th>Disorder</th>
<th>Code</th>
</tr>
</thead>
<tbody>
<tr>
<td>Migraine with aura (code 1.2) and ophthalmoplegic migraine (code 13.17)</td>
<td></td>
</tr>
<tr>
<td>Cluster headache and other trigeminal-autonomic cephalgias (code 3)</td>
<td></td>
</tr>
<tr>
<td>Ischaemic stroke and transient ischaemic attacks (code 6.1)</td>
<td></td>
</tr>
<tr>
<td>Intracerebral haemorrhage (code 6.2.1)</td>
<td></td>
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<tr>
<td>Subarachnoid hemorrhage (code 6.2.2)</td>
<td></td>
</tr>
<tr>
<td>Unruptured vascular malformations (code 6.3)</td>
<td></td>
</tr>
<tr>
<td>Arteritis (code 6.4)</td>
<td></td>
</tr>
<tr>
<td>Carotid or vertebral artery pain (code 6.5)</td>
<td></td>
</tr>
<tr>
<td>Cerebral venous thrombosis (code 6.6)</td>
<td></td>
</tr>
<tr>
<td>High cerebrospinal fluid pressure (code 7.1)</td>
<td></td>
</tr>
<tr>
<td>Low cerebrospinal fluid pressure (code 7.2)</td>
<td></td>
</tr>
<tr>
<td>Intracranial neoplasms (code 7.4)</td>
<td></td>
</tr>
<tr>
<td>Syndrome of transient headache and neurologic deficits with cerebrospinal fluid lymphocytosis (“HaNDL”, code 7.8)</td>
<td></td>
</tr>
<tr>
<td>Trigeminal neuralgia (tic douloureux; code 13.2)</td>
<td></td>
</tr>
<tr>
<td>Tolosa-Hunt syndrome (code 13.16)</td>
<td></td>
</tr>
<tr>
<td>Acute herpes zoster (code 13.15.1) &amp; post-herpetic neuralgia (code 13.15.2)</td>
<td></td>
</tr>
</tbody>
</table>

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Diagnosis of focal headache

**Case report 1**

This 25-year-old woman visits her doctor because of disabling migraine attacks since the age of 20 years. Each attack starts with visual disturbances: a small spot in one hemifield first becomes fuzzy, then it enlarges over a few minutes interfering with (for example) reading. The greyish visual field defect is surrounded by “worm-like” white luminous strings and occupies the whole hemifield about after 10 min (to the right in some attacks, to the left in others). Then, about a third of the way through the attacks she feels tingling and numbness in the hand on the side of the visual field defect; this may “jump” to the ipsilateral labial commissure and rarely spread over the whole arm. These neurological symptoms disappear gradually after 20–30 min and are replaced by a hemicrania—typically located on the side opposite to the field defect. The headache is of moderate intensity, associated with anorexia and mild sensitivity to light; if untreated, it lasts for 4–6 h. Attacks occur irregularly (average of six to eight per year) and tend to cluster on successive days in the same week. The patient does not report any trigger factors. Given the typical pattern of the symptoms, there is no indication for further investigation.

**Migraine with aura (1.2)**

Visual disturbances are the most common aura symptoms, occurring in about 99% of patients. Sensory symptoms and motor or speech disturbances seldom occur without pre-existing visual symptoms. Migrainous aura phenomena are thought to be caused by spreading depression and can be distinguished from those produced by a seizure or by a transient ischaemic attack (TIA) by their progressive onset, their spreading over time, and their quality (table). In patient care this distinction is a common problem and clinicians commonly rely on phenomenology, when making their diagnosis.

Compared with migraine without aura, migraine with aura is characterised by headache of low intensity and short duration. The headache that follows the aura may phenotypically resemble tension-type headache (code 2) and recently migrainous-like auras preceding typical cluster-headache attacks were described in rare patients. Basilar type migraine (code 1.2.6) is a controversial entity, which is described as migraine with aura where symptoms can be attributed to dysfunction in the brainstem or in both hemispheres and where no motor weakness is present. In addition to a fully reversible visual, sensory, or speech aura without motor weakness, two or more aura symptoms have to be of the following type: dysarthria, vertigo, tinnitus, decreased hearing, double vision, ataxia, decreased level of consciousness, simultaneous bilateral visual symptoms in both the temporal and nasal field of both eyes, and simultaneous bilateral paraesthesias. The differential diagnosis of such phenomena include vascular and other pathologies, the
or sporadic (code 1.2.5) forms. This rare subtype of pleocytosis (code 7.8).

Temporary neurological symptoms and lymphocytic lumbar puncture to rule out pseudomigraine with sporadic form, will require neuroimaging as well as a (which codes the

Complications of diagnosis
Persistent aura without infarction (1.5.3) and migrainous infarction (code 1.5.4) complicate the diagnosis of migraine with aura. In the former, one or more aura symptoms persist for more than 24 h and it is important to exclude migrainous infarction by MRI including diffusion-weighted imaging. Migrainous infarction is characterised by neurological deficit during a migraine attack that is typical of those previously experienced by a patient; the deficit must last for more than 60 min and be associated with an ischaemic infarction in the relevant area as shown by neuroimaging techniques. Most importantly, other causes of infarction have to be ruled out.14

Ophthalmoplegic "migraine" (code 13.17) is a very rare disorder that typically starts in childhood. It is characterised by repeated attacks of headache with migrainous features, but with typically long duration of a week or more, associated with a palsy of one or more of the cranial nerves involved in eye movements. Intracranial mass lesions have to be excluded for its diagnosis. Several MRI studies have shown reversible thickening or contrast enhancement of the cisternal portion of the oculomotor nerve.22 This finding suggests that the disorder is caused by an inflammatory oculomotor neuritis13 and that "ophthalmoplegic migraine" is a misnomer.

Cluster headache and other trigeminal-autonomic cephalalgias
The most typical feature of cluster headache is the temporal clustering of attacks during several weeks separated by remissions of at least 14 days, but generally of several months (episodic cluster headache, code 3.1.1.). In chronic cluster headache (code 3.1.2) remissions of at least 1 month are absent for more than 1 year.

Cluster headache is a primary neurovascular headache that is unilateral but differs from migraine. Characteristics distinguishing cluster headache from migraine are short duration (typically 30–45 min), presence of autonomic symptoms on the painful side, and the extreme intensity of the pain. Most patients are agitated and pace the floor, which is clearly different from people with migraine who generally lie down and seek rest during the attack.14 Cluster headache attacks generally happen in the evening or during sleep. Cluster headache has to be distinguished from trigeminal neuralgia. In the International Headache Society's classification, the trigeminal autonomic cephalalgias (TACs) are grouped with cluster headache. They share the clinical hallmark of unilateral headache with prominent ipsilateral cranial parasympathetic autonomic features.

Cluster headache is a rare disorder with prevalence below 0.1%;16 about 10% of patients have the chronic form.17 The onset of the first attack is typically between 20 years and 40 years of age; about 70% of patients are men18 and, for unknown reasons, there is an association with smoking. Genetic influences seem to be less pronounced than in migraine, but familial disorder with an autosomal dominant inheritance pattern does exist.19 Recent studies suggest an involvement of the posterior ventral hypothalamic grey matter in the generation of attacks,20 although the cavernous sinus may play a part during the attack itself.21 Cluster-like headaches, as well as other TACs, can be caused by organic lesions—the most common of which are pituitary adenomas.22

Other headaches classified in the group of the TACs, episodic paroxysmal hemicrania (code 3.2.1), and chronic paroxysmal hemicrania (code 3.2.2) differ from cluster headache by a shorter duration, a higher frequency of attacks, and a greater prevalence in women than in men.23 The so called SUNCT (short-lasting unilateral neuralgiform headache with conjunctival injection and tearing) syndrome (code 3.3) is characterised by the shortest attack duration (10–200 s) and pronounced autonomic signs. If such a syndrome is suspected, a lesion in the posterior fossa must be ruled out, as this might mimic the primary form.24–26 Hemicrania continua (code 3.4) is a non-remitting form of

Differential diagnosis of focal paroxysmal neurological symptoms

<table>
<thead>
<tr>
<th>Feature</th>
<th>TIA</th>
<th>Epilepsy</th>
<th>Migraine</th>
</tr>
</thead>
<tbody>
<tr>
<td>Onset</td>
<td>Sudden</td>
<td>Sudden</td>
<td>Progressive</td>
</tr>
<tr>
<td>Progression rate</td>
<td>None</td>
<td>Fast</td>
<td>Slow</td>
</tr>
<tr>
<td>Different symptoms</td>
<td>Simultaneous</td>
<td>In succession</td>
<td>In succession</td>
</tr>
<tr>
<td>Type of visual symptoms</td>
<td>Negative</td>
<td>Positive coloured</td>
<td>Negative or positive, uncoloured</td>
</tr>
<tr>
<td>Territory</td>
<td>Vascular</td>
<td>Cortical</td>
<td>Cortical</td>
</tr>
<tr>
<td>Duration</td>
<td>Short (10–15 min)</td>
<td>Short (min)</td>
<td>Long (30–60 min)</td>
</tr>
</tbody>
</table>

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TAC. It shares with paroxysmal hemicrania an absolute responsiveness to indometacin, which is part of their diagnostic criteria.

**Headache associated with vascular disorders**

All headaches in this group have symptoms or signs of a vascular disorder, appear as a new symptom or as part of a clinical picture previously unknown in the patient, and are in close temporal relation to the onset of the vascular disorder.

**Acute ischaemic cerebrovascular disease (code 6.1)**

The importance of headache as a symptom of ischaemic cerebrovascular disease is still neglected by many physicians. However, comprehensive reviews have shown several associations. The incidence of headache accompanying transient ischaemic attacks (TIAs) or strokes varies from 15–65% between studies (average 30%). Headache seems to be more likely in patients with posterior circulation ischaemia. 2) Headache precedes the event in about 10% of patients with haemorrhage or ischaemic attack. 3) The headache is typically on the side of the affected artery or frontal when the carotid or the posterior cerebral arteries are involved. In basilar or vertebral artery stenosis or occlusion the headache is typically occipital and bilateral. 4) In ischaemic cerebrovascular disease, headaches vary between continuous and throbbing, and most are of moderate intensity. 5) Headache at the onset of the ischaemic stroke does not help to distinguish embolic from atherothrombotic stroke. Headache is probably less common in lacunar infarction.

Whether or not migraine is an independent risk factor for ischaemic stroke is still debated. Several surveys indicate that this is only true in young women with migraine with aura, and that in these patients the risk is amplified by use of the contraceptive pill and by smoking.

**Non-traumatic and traumatic intracranial haemorrhage**

As a diagnostic element, headache is far more useful in haemorrhagic than in ischaemic stroke, because it is more commonly the presenting symptom. The overall incidence of headache as a major symptom of intracerebral haemorrhages (code 6.2.1) ranges from 36–66%. In all the published series there is a proportion of non-comatose, non-aphasic patients who do not have headaches—ranging from 10% in the basal ganglia to 30% in lobar haemorrhages.

In patients with head trauma, who recover consciousness and subsequently deteriorate, headache is a common and useful indicator of the late development of an acute epidural (code 5.5.1) or a subdural (code 5.5.2) haematoma. This type of headache may be similar to that caused by high intracranial pressure. Subdural haematomas can sometimes produce a characteristic paroxysmal type of headache that comes and goes irregularly throughout the day, lasts only a couple of minutes, and is generally accompanied by sympathetic overactivity. Most are frontal, but when the subdural haematoma is in the posterior fossa, the headache is likely to be occipital. In the post-traumatic situation, occipital headache associated with neck stiffness may indicate the onset of cerebellar pressure coning caused by a supratentorial subdural haematoma.

**Subarachnoid haemorrhage (code 6.2.2)**

Headache caused by subarachnoid haemorrhage (SAH) is typically abrupt in onset (split-second headache)—which is its key feature—and incapacitating in severity. When not maximum from the beginning, time from onset to greatest pain intensity is less than 60 min for ruptured aneurysm and less than 12 h if caused by an arteriovenous malformation. The headache is diffuse and commonly occipital radiating to the neck. It can be accompanied by blunting of consciousness, vomiting, and stiff neck. CT scan (which may be normal in 10% of patients) and CSF examination can confirm the diagnosis of subarachnoid haemorrhage (case report 2).

**Unruptured vascular malformation (code 6.3)**

About 25% of patients with an intracranial aneurysm report unusual headaches before a rupture occurs, partly of a “thunderclap” type. These are generally called “sentinel” or

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**Case report 2**

A 50 year-old woman suddenly presented with a posterior headache that was rapidly followed by a scintillating scotoma in her left visual hemifield and paresthesias of the left hand. Neurological symptoms lasted for 45 min, but the headache remained intense for several hours and was accompanied by nausea and vomiting. The patient has no personal or family history of migraine. A CT scan shows blood in the subarachnoid space in the parieto-occipital regions predominantly on the right side (figure 3).

**Figure 3. CT scan (horizontal section) showing a subarachnoid haemorrhage over the right parietal cortex.**
"premonitory" headaches and are thought to be caused by the aneurysm intermittently leaking. Not every severe headache of abrupt onset is a symptom of an intracranial aneurysm. Most patients struck by thunderclap headache, who have normal CT scan and lumbar puncture results, do not subsequently present with subarachnoid haemorrhage. There is no reliable difference in the clinical phenotype between "primary thunderclap headache" and the headache caused by subarachnoid haemorrhage.

Consequently, the following guidelines are helpful in patients with an abrupt "worst headache of my life", and a normal neurological examination: first take a CT scan; if normal, do a lumbar puncture. If the lumbar puncture is normal, the patient can be reassured that the headache is benign (but it may recur in some patients). If any feature of the CT scan or lumbar puncture is abnormal, a conventional angiography is required. The role of magnetic resonance angiography is not yet established.

Arteriovenous malformations (code 6.3.2), which account for 6% of all subarachnoid haemorrhages, commonly cause focal seizures or neurological deficits. Although the relation of migraine and other headaches to unruptured arteriovenous malformations is poorly studied, there are several case reports of them mimicking attacks of migraine with aura. Possible diagnostic clues are strictly unilateral symptom localisation, a lack of family history for migraine, absence of visual aura symptoms, and atypical auras (case report 3).

Case report 3
A 40-year-old man had headache attacks since age 17 years. Attacks started with scintillations in his left visual field accompanied by paraesthesia and numbness of the entire left side of the body. These neurological symptoms were lock-sided and followed after a few minutes by a right hemianopia of moderate intensity. Headaches lasted several hours, the sensory symptoms outlasted the headaches by 1–2 h. The frequency of attacks varied over the years by two to ten a year and there was no family history of migraine. He was diagnosed as having migraine with visual aura. Neuroradiography disclosed a right parieto-occipital angioma (figure 4) that was surgically treated. The patient has not reported any attack since.

**Figure 4.** Angiographic demonstration of a parieto-occipital arteriovenous malformation mimicking migraine with aura. Reproduced with permission from the American Medical Association.

**Arteritis (code 6.4)**
Giant-cell arteritis (temporal arteritis or Horton’s disease; code 6.4.1) is any type of new persisting headache and one or more of the following: swollen and tender scalp arteries with high erythrocyte sedimentation rate or C-reactive protein; a close temporal relation with possible other symptoms and signs of giant-cell arteritis; and major improvement or disappearance of headache within 3 days of steroid therapy. The presence of typical histopathological features on temporal artery biopsy is no longer a mandatory diagnostic requirement.

**Carotid or vertebral artery pain (code 6.5)**
Ipsilateral headache or cervical pain may be the only manifestation of carotid or vertebral dissection (code 6.5.1), or it can accompany focal neurological symptoms (case report 4). This is of particular importance, as generally, the headaches occur early and precede ischaemia, the most feared complication of a dissection. Most headaches are ipsilateral to the dissected artery and severe.

**Cerebral venous thrombosis (code 6.6)**
Headache is the most common, and typically first, symptom in cerebral venous thrombosis. It is generally diffuse and subacute. Its intensity is highly variable. Associated neurological signs (focal deficit or seizures), or raised intracranial pressure, are present in most patients. Headache can occasionally be the only symptom of cerebral venous thrombosis, which is another reason why persistent new-onset headache should prompt appropriate investigations.

**Headache attributed to nonvascular intracranial disorder**
Within this group of headaches, we will focus on the more common disorders, the diagnosis of which is difficult at a stage when headache is the only symptom such as in benign intracranial hypertension, post-lumbar puncture headache, and headache associated with brain tumour.
High CSF pressure (code 7.1)

Idiopathic intracranial hypertension (code 7.1.1), also called pseudotumour cerebri or benign intracranial hypertension, may mimic chronic tension-type headache: it is generalised, non-throbbing, and sometimes of low or moderate intensity. It is increased by coughing or straining. The following symptoms are characteristic for the diagnosis of idiopathic intracranial hypertension: predominance in young, obese women (93%); most severe headache ever experienced by the patient (93%); nausea (57%); vomiting (30%); orbital pain (43%); transient visual obscuration (71%); diplopia (38%); and visual loss (31%). Papilloedema, without neuroradiological abnormalities (except for possible "empty sella"), is pathognomonic for this disorder. CSF cytology is normal, but protein content may be low. Axial CT may show narrow, slit-like ventricles.

Low CSF pressure (code 7.2)

The clinical hallmark of low CSF pressure headache is that the pain is aggravated by orthostasis and disappears in a supine position. The time lag after change of position can be up to 15 min. The headache may be frontal, occipital, or diffuse and is typically severe, dull, or throbbing. Other symptoms include nausea, vertigo, and tinnitus, and shaking of the head aggravates pain. Results of physical examination are generally normal. Sixth-nerve palsy may occur and is reversible in most patients. Normal spinal fluid pressure ranges from 0–30 mm H2O in the lateral supine position.

The most common cause of low CSF pressure is lumbar puncture (case report 5). According to a recent study, the incidence of headache after lumbar puncture (code 7.2.1) may reach 37%. Headache after lumbar puncture is more common in patients with a history of primary headaches.

There are many other causes of low-pressure-headache syndrome—such as post-traumatic, postoperative, or spontaneous (or idiopathic) CSF leakages (code 7.2.3) or systemic illnesses such as dehydration, diabetic coma, hyperpyoecia, or uraemia. Pachymeningeal contrast enhancement on MRI is a hallmark of intracranial hypotension (figure 5). Dilatation of epidural veins and CSF leakage into the epidural space can be seen with MRI in patients with "spontaneous" low CSF pressure.

Case report 5

This 37-year-old man had a lumbar puncture because of recurrent paraesthesias in his left arm with fluctuating sensory loss. Multiple sclerosis was suspected. He developed severe postural headache associated with nausea and photophobia. 3 days later, he complained of diplopia and presented with bilateral lateral rectus paresis. The diagnosis of intracranial hypotension caused by CSF leakage after lumbar puncture was made and he was treated with an epidural blood patch in the lumbar region. The headache resolved within 1 week. The bilateral VI nerve palsy remitted gradually over 4 weeks.

Intracranial neoplasm (code 7.4)

Headache occurs at presentation in 36–50% of adults with brain tumours and develops in the course of the disease in 60%. Headache is typically generalised, of the dull, deep, aching type, and intermittent at first. Simple analgesics relieve many of these headaches. If there is variation during the 24 h cycle, the headache is typically worse in the early morning hours. This variation is more prominent with rapidly growing tumours than with those of slower growth.

In 30–80% of patients the headache overlies the projection of the tumour to the nearest skull surface. Some general rules about headache as an aid to tumour-localisation in patients with brain tumour have been proposed by Dalessio. 1) In about a third of all patients, headache overlies the tumour. 2) If the tumour is above the tentorium, the pain is commonly at the vertex or in the frontal region. 3) If the tumour is below the tentorium, the pain is occipital and cervical muscle spasms may be present. 4) Headache is nearly always present in patients with posterior fossa tumours. 5) If the tumour is midline, it may increase with coughing, straining, or sudden head movement. 6) If the tumour is hemispheric, the pain is typically felt on the same side of the head. 7) If the tumour is chiasmal, at the sella, the pain may be felt on the top of the head.

In specialised headache or pain clinics, brain tumours account for less than 1% of patients. There is significant overlap in the clinical characteristics of headaches caused by brain tumours and migraine-type or tension-type headaches. The following clues indicate that a thorough neuroradiological examination is indicated: any headache of recent onset; previously existing headaches that have changed in character; a lock-sided headache not resembling the primary unilateral headaches; morning or nocturnal headaches associated with vomiting in a patient who has no history of migraine.

Headache with neurological deficits and CSF lymphocytosis (“HaNDL”) (code 7.8)

This syndrome is of unknown cause. It may mimic migraine with prolonged aura with the notable exception that there is lymphocytosis on CSF examination. The episode may start...
with visual symptoms, but one-sided paraesthesias and weakness are more common. The headache may precede the neurological symptoms; it may be unilateral or bilateral and accompanied by nausea, vomiting, and sensoriphobia. Although the headache lasts for several days, the neurological deficits take a few weeks to disappear completely. The episodes may recur several times in the same individual and are thought to be benign.10,57

Cranial neuralgia, nerve trunk pain, and deafferentation pain

**Trigeminal neuralgia (tic douloureux; code 13.1)**

Classical trigeminal neuralgia (code 13.1.1) is characterised by very short (a few seconds up to 2 min) attacks of intense, electric-shock-like pain. Most trigeminal neuralgia starts in the second (or third) division of the trigeminal nerve. More women are affected than men (ratio three to two), and most patients are older than 50 years. Pain occurs spontaneously or is triggered by stimuli such as washing, shaving, chewing, teeth brushing, or speaking. Painful episodes start and end abruptly and may recur dozens of times or more in a day but interfere little with sleep. Remissions of variable duration are described. The pain often induces reflex spasms of facial muscles on the affected side, hence the name "tic douloureux".

On MRI or during surgical treatment of trigeminal neuralgia, the trigeminal nerve root commonly looks compressed or in intimate contact with a vessel. Vascular decompression (Janetta’s procedure) is a highly effective treatment of drug-resistant trigeminal neuralgia, which is, in many patients, a secondary pain disorder. For this reason, the term “idiopathic” trigeminal neuralgia is replaced by “classic” trigeminal neuralgia in the revised classification.

Trigeminal neuralgia with non-vascular causes—such as acoustic neurinomas, brainstem infarctions, or multiple sclerosis—is called symptomatic, trigeminal neuralgia (code 13.1.2). The main differences from the classic form are a younger age at onset, persisting ache between paroxysms, and signs of sensory impairment in the distribution of the corresponding trigeminal division.

**Tolosa-Hunt syndrome (code 13.16)**

This syndrome is characterised by episodic orbital pain with paralysis of the third, fourth, or sixth cranial nerves that resolves spontaneously after days or weeks, but may relapse and remit.

There are several other causes of painful ophthalmoplegia (panel 2). Extensive clinical assessment is needed to exclude inflammatory, infectious, vascular, or neoplastic causes (panel 3).59

**Herpes zoster (code 13.15.1) and postherpetic neuralgia (code 13.15.2)**

In 10–15% of patients with the virus, herpes zoster affects the trigeminal ganglion, with particular affinity for the ophthalmic division (80% of patients). Palsy of the third, fourth, or sixth cranial nerve is sometimes observed. Herpes zoster may also involve the geniculate ganglion, with an eruption in the external auditory meatus (Ramsay-Hunt’s zone) which is associated with facial nerve palsy in many patients.

Postherpetic neuralgia (code 13.15.2) is a chronic pain that commonly develops during the acute phase of infection and persists more than 6 months. It is a common sequel of infection with herpes zoster and affects up to 50% of patients, particularly the elderly.60,61 Incidence is reduced by antiviral treatment at the acute phase. The typical neuropathic pain is felt in the area formerly involved by the infection; it is...
constant, moderate to severe, and many patients describe it as burning. Paraesthesias and hypaesthesia are common.

Conclusion

Many headache syndromes are associated with focal neurological symptoms or signs. Good knowledge of the clinical phenotypes of primary headaches, a detailed history, and a thorough clinical examination are prerequisites for their differential diagnosis. There are well-defined clinically relevant diagnostic hints on the headache as well as focal neurological signs or symptoms for some of the disorders (panel 4). The neurological symptoms produced by the migrainous aura are probably the most characteristic and recognisable. However, structural lesions—such as vascular malformations—can cause symptoms similar to migraine with aura, which highlights that in most patients with headache and focal neurological symptoms paraclinical investigations are necessary.

References


