syndromes. Functional imaging studies have suggested abnormal hypothalamic activity. Patients are more often smokers with a higher than average alcohol consumption.

Clinical features

Cluster headache is strikingly periodic, featuring runs of identical headaches beginning at the same time for weeks at a stretch (the 'cluster'). Patients may experience either one or several attacks within a 24-hour period, and typically are awoken from sleep by symptoms ('alarm clock headache'). Cluster headache causes severe, unilateral periorbital pain with autonomic features, such as ipsilateral tearing, nasal congestion and conjunctival injection (occasionally with the other features of a Horner syndrome). The pain, though severe, is characteristically brief (30–90 minutes). In contrast to the behaviour of those with migraine, patients are highly agitated during the headache phase. The cluster period is typically a few weeks, followed by remission for months to years, but a small proportion do not experience remission.

Management

Acute attacks can usually be halted by subcutaneous injections of sumatriptan or inhalation of 100% oxygen. The brevity of the attack probably prevents other migraine therapies from being effective. Migraine prophylaxis is often ineffective too but attacks can be prevented in some patients by verapamil, sodium valproate, or short courses of oral glucocorticoids. Patients with severe debilitating clusters can be helped with lithium therapy, although this requires monitoring.

Trigeminal neuralgia

This is characterised by unilateral lancinating facial pain, most commonly involving the second and/or third divisions of the trigeminal nerve territory, usually in patients over the age of 50 years.

Pathophysiology

For most, trigeminal neuralgia remains an idiopathic condition but there is a suggestion that it may be due to an irritative lesion involving the trigeminal root zone, in some cases an aberrant loop of artery. Other compressive lesions, usually benign, are occasionally found. Trigeminal neuralgia associated with multiple sclerosis may result from a plaque of demyelination in the brainstem.

Clinical features

The pain is repetitive, severe and very brief (seconds or less). It may be triggered by touch, a cold wind or eating. Physical signs are usually absent, although the spasms may make the patient wince and sit silently (tic douloureux). There is a tendency for the condition to remit and relapse over many years. Rarely, there may be combined features of trigeminal neuralgia and cluster headache ('cluster–tic').

Management

The pain often responds to carbamazepine. It is wise to start with a low dose and increase gradually, according to effect. In patients who cannot tolerate carbamazepine, oxcarbazepine, gabapentin, pregabalin, amitriptyline or glucocorticoids may be effective alternatives, but if medication is ineffective or poorly tolerated, surgical treatment should be considered. Decompression of the vascular loop encroaching on the trigeminal root is often performed and may lead to pain relief in some cases. Otherwise, localised injection of alcohol or phenol into a peripheral branch of the nerve may be effective.

Headaches associated with specific activities

These usually affect men in their thirties and forties. Patients develop a sudden, severe headache with exertion, including sexual activity. There is usually no vomiting or neck stiffness, and the headache lasts less than 10–15 minutes, though a less severe dullness may persist for some hours. Subarachnoid haemorrhage needs to be excluded by CT and/or CSF examination (see Fig. 29.13) after a first event. The pathogenesis of these headaches is unknown. Although frightening, attacks are usually brief and patients may need only reassurance and simple analgesia for the residual headache. The syndrome may recur, and prevention may be necessary with propranolol or indometacin.

Other headache syndromes

A number of rare headache syndromes produce pains about the eye similar to cluster headaches (Box 28.24). These include chronic paroxysmal hemicrania and SUNCT (short-lasting unilateral neuralgiform headaches with conjunctival injection and tearing). The recognition of these syndromes is useful because they often respond to specific treatments such as indometacin.

28.24 Paroxysmal headaches

Type	Character of pain	Duration	Location	Comment
Ice pick	Stabbing	Very brief (split-second)	Variable, usually temporoparietal	Benign, more common in migraine
Ice cream	Sharp, severe	30–120 secs	Bitemporal/occipital	Obvious trigger by cold stimuli
Exertional/sexual activity	Bursting, thunderclap	Severe for mins, then less severe for hours	Generalised	Subarachnoid haemorrhage needs to be excluded
Cough	Bursting	Secs to mins	Occipital or generalised	Intracranial pathology needs to be excluded (especially craniocervical junction)
Cluster headache (migrainous neuralgia)	Severe unilateral, with ptosis, tearing, conjunctival injection, unilateral nasal congestion	30–90 mins 1–3 times per day	Periorbital	Usually in men, occurring in clusters over weeks/months
Chronic paroxysmal hemicrania	Severe unilateral with cluster headache-like autonomic features (see above)	5–20 mins, frequently through day	Periorbital/temporal	Usually in women, responds to indometacin
SUNCT*	Severe, sharp, triggered by touch or neck movements	15–120 secs, repetitive through day	Periorbital	May respond to carbamazepine

***S**hort-lasting, **u**nilateral, **n**euralgiform headache with **c**onjunctival injection, **t**earing, rhinorrhoea and forehead sweating.