

Salvatore Gentile

Indications for the diagnosis and treatment of acute headaches correlated with neurological pathologies

Published online: 20 July 2005

S. Gentile (✉)
Department of Neurosciences,
Neurology 3, Headache Centre,
University of Turin,
Via Cherasco 15, I-10126 Turin, Italy
e-mail: sgentile@molinetto.piemonte.it
Tel.: +39-011-6334763
Fax: +39-011-6638510

Abstract The majority of patients consulting a doctor for headache are generally suffering from a primary headache. Nevertheless, the possibility of it being symptomatic, and sometimes life threatening, must always be in the forefront of the doctor's mind. Medical history and clinical examination first, and instrumental examinations afterwards, are the keys to a correct diagnosis, and the first steps for correct treatment.

Key words Symptomatic headache • Medical history • Clinical examination • Instrumental examinations • Therapy

Introduction

A headache may have an extraordinarily high number of causes [1]. In the majority of cases it is a primary headache. Nevertheless, 2% of the patients followed at the outpatients' clinic and 4% of those brought to the emergency room are suffering from a symptomatic headache [2, 3]. A differential diagnosis therefore becomes essential. Accurate collection of medical history and a thorough clinical examination represent the keystone to reaching a correct diagnosis. Age of onset, location, promoting or aggravating factors, associated symptoms and signs, clinical overture and evolution are the most useful characteristics to take into consideration.

Age

If a headache suddenly appears in a patient who is over 50, it must always be considered a symptomatic one in the first place, its cause possibly being a tumour, a cerebrovascular illness or giant-cell arteritis.

Location

When the pain always has the same location, neuroimaging helps to exclude the presence of a brain aneurysm or an arteriovenous malformation.

Promoting or aggravating factors

Cough or posture changes may favour the onset of a headache attack [4] if there is an underlying structural cause such as Arnold-Chiari malformation type 1, colloid cyst of the third ventricle or tumour growing in the ventricular system. The explosive headache which results is due to temporary intracranial hypertension, probably inducing a further impaction of the cerebellar tonsils in the cerebellar-foramen magnum, caused by Valsalva manoeuvre or obstruction of Monro's foramen following the intraventricular tumour's dislocation.

Exertional or sexual headache may be symptomatic of a subarachnoid haemorrhage (SAH). If a headache begins or remarkably worsens when the patient has an upright position, it may be considered pathognomonic for CSF hypotension syndrome [5].

Therefore, before diagnosing it as a primary headache correlated to a particular situation, the above causes must be excluded.

Associated signs and symptoms

Focal cerebral symptoms may be associated with a headache attack, representing the so-called "aura". Visual and sensory auras generally begin gradually and diffuse slowly: it takes almost 20 min to reach their widest distribution. Their slow diffusion helps to distinguish them from transient ischaemic attacks, which have a quicker diffusion.

Brainstem symptoms (dizziness, diplopia, ataxia, dysarthria) might be associated with basilar migraine, more frequent during childhood or adolescence. The onset of a headache in an adult must be carefully evaluated, because vertebral-basilar artery insufficiency syndrome shows more frequently in adults and has the same symptoms [6, 7].

Horner's syndrome is frequently associated with cluster headache or homolateral carotid artery dissection in the neck [8]. This possibility must be taken into consideration when a patient shows symptoms and signs suggesting a cluster headache, but the pain is continuous instead of intermittent.

Headache associated with a general malaise, high temperature and high ESR values is the typical manifestation of giant cell arteritis [9]. Jaw claudication may be associated.

Headache associated with high temperature and stiff neck may be the opening sign of meningitis or meningoencephalitis. When the meninges of the cranial basis are involved, the deficiency of some particular cranial nerve (VII, VIII, VI, III) may help to formulate the right diagnosis.

Evolution

If a headache begins acutely and very soon becomes severe, it is more likely an organic than a primary one. First, SAH must be excluded. When evaluating a migraine characterised by recent onset, the possibility of it being a migraine following a SAH must always be considered.

In a typical migraine attack, pain reaches its highest point in 30 min to two hours, while the pain caused by a SAH has a sudden onset and is severe from the very beginning. A stiff neck might be present during a migraine attack, but it is rare to find meningism; on the contrary, in a SAH meningism is part of the clinical picture, even if it sometimes appears after several hours. Presyncopal signs or a faint at the onset of the pain; a worse than ever cranial pain ("the worst attack of my life"); and the presence of neurological failure signs all indicate the presence of a SAH [10]. The rupture of an aneurysm is the most frequent cause of a SAH; to a lesser extent the cause is an arteriovenous malformation. An increase of the aneurysm or of the malformation's volume, or their leak into the subarachnoid space, which would not be enough to cause a catastrophic haemorrhage, might instead be the cause of one or more warning headaches, which might be interpreted as migraine attacks [11]. Even though it is difficult, it is important to identify as soon as possible a warning headache, because this is the only way to begin timely and successful treatment, which is essential to save the patient's life. Non-headache suffering patients, who report the recent onset of a head pain with a sudden beginning (the so-called "thunderclap headache") and already known headache suffering patients who report a worsening of the usual pain without any apparent cause must always be carefully evaluated.

If a headache does not gradually wane, but on the contrary progressively worsens over days or weeks, it could be the sign of a mass lesion [12]. The symptoms of a headache associated with a mass lesion and those of a tension-type headache are very similar and therefore these two possibilities must be considered when formulating a differential diagnosis. If a headache is the sign of increasing intracranial pressure, often it shows in the morning, worsens by sneezing or physical activity and is accompanied by vomiting (not preceded by nausea, especially in children). Headache is more often the sign of an intracranial tumour in children than in adult patients. In fact, tumours of the posterior cranial fossa are more frequent during childhood.

When to suspect a dangerous headache

Even if the neurological examination does not reveal anomalies, a doctor should suspect that a headache might

be a symptomatic one if certain clinical characteristics are present, such as:

- recent onset of a severe headache of a new kind in middle aged or older patients,
- inexplicable worsening of a pre-existing headache,
- fixed unilateral location of the pain,
- no response to treatment,
- progressive course,
- association with physical efforts,
- association with derangement, loss of consciousness, seizures, profuse vomiting.

If these clinical characteristics go together with anomalies of the neurological objective examination, the diagnosis of “symptomatic headache” can be formulated with little or no doubt.

Diagnosis

All the examinations used in neurology and neurosurgery might be requested to diagnose a headache. The indiscriminate use of medical tests has no justification and would represent a waste of money, being at the same time a source of anxiety for the patient involved.

The choice must be based on the results from the medical history and the objective examination [13, 14]. A full blood test might be useful, if an infectious disease is suspected; ESR will help to diagnose a giant cell arteritis, other blood vessel inflammations, or hidden mass lesions; head CT scan is the right choice to find the presence of a SAH; brain MRI is more sensitive and specific when the presence of tumours of the posterior cranial fossa or CSF flow pathologies are suspected; MR angiography allows a better definition in case of venous sinuses thrombosis, aneurysms, arteriovenous malformations, carotid artery

dissection (in this last case, Doppler ultrasonography of the supra-aortic blood vessels could be useful).

If a SAH is suspected, but the head CT scan is normal, or meningitis and/or encephalitis is suspected, a lumbar puncture is suggested. Cerebral angiography represents the so-called golden standard to diagnose structural lesions of cerebral circulation, especially aneurysms, vasculitis and arteriovenous malformations.

Therapy

Where symptomatic headaches are concerned, therapy will be both symptomatic and causal.

Pain therapy is based on the use of NSAIDs, analgesics and triptans. Sumatriptan can be effective in different forms of secondary headache, including those due to dissection of carotid or vertebral artery, head and neck cancer and seizures [15, 16]. Anti-oedema drugs (for wet brain or intracranial hypertension), AEDs (for seizures) and antipyretics (for high temperature) might be used when a symptomatic therapy is necessary.

Causal therapy might be both medical (e.g., antibiotics or antiviral drugs to treat meningitis or encephalitis; corticosteroid drugs to treat giant cell arteritis, etc.) and surgical (in case of a mass lesion or an aneurysm, etc.).

Conclusions

When evaluating a head pain, it must always be remembered that a headache can be called “a primary one” only if and when the possibility of it being symptomatic has been excluded without doubt.

References

1. Headache Classification Subcommittee of the International Headache Society (2004) The International Classification of Headache Disorders, 2nd edn. *Cephalalgia* 24[Suppl 1]:1–151
2. Bigal ME, Bordini GA, Speciali JG (2000) Etiology and distribution of headaches in two Brazilian primary care units. *Headache* 40:241–247
3. Steiner TJ, Fontebasso M (2002) Headache. *Br Med J* 325:881–886
4. Pascual J, Igessias F, Oterino A et al (1996) Cough, exertional, and sexual headaches: an analysis of 72 benign and symptomatic cases. *Neurology* 46:1520–1524
5. Mokri B (2003) Headaches caused by decreased intracranial pressure: diagnosis and management. *Curr Opin Neurol* 16:319–326
6. Martsen BH, Sorensen PS, Marquardsen J (1990) Transient ischemic attacks in young patients: a thromboembolic or migrainous manifestation? A ten-year follow-up of 46 patients. *J Neurol Neurosurg Psychiatry* 53:1029–1033
7. Caplan R (1991) Migraine and vertebrobasilar ischemia. *Neurology* 41:55–61

-
8. Silbert PL, Mokri B, Schievink WI (1995) Headaches and neck pain in spontaneous internal carotid and vertebral artery dissections. *Neurology* 45(8):1517–1522
 9. Caselli RJ, Hunder GC, Whisnant JP (1988) Neurologic disease in giant cell (temporal) arteritis. *Neurology* 38:352–359
 10. Vermeulen M (1996) Subarachnoid hemorrhage: diagnosis and treatment. *J Neurol* 243:496–501
 11. Leblanc R (1987) The minor leak preceding subarachnoid hemorrhage. *J Neurosurg* 66:35–39
 12. Lavyne MH, Patterson RH (1987) Headache and brain tumor. In: Dalessio DJ (ed) *Wolff's headache and other head pain*. Oxford University Press, New York, pp 343–349
 13. Evans RW (1996) Diagnostic testing for the evaluation of headache. *Neurol Clin* 14(1):1–26
 14. Newman LC, Lipton RB (1998) Emergency department evaluation of headache. *Neurol Clin* 16(2):285–303
 15. Leira EC, Cruz-Flores S, Leacock RO, Abdulrauf SI (2001) Sumatriptan can alleviate headaches due to carotid artery dissection. *Headache* 41:590–591
 16. Manfredi PL, Shenoy S, Payne R (2000) Sumatriptan for headache caused by head and neck cancer. *Headache* 40:758–760