lateral ventricles. Although slow growing and rarely large enough to create neurological symptoms, they can occasionally cause seizures, headaches, visual field defects, or gait disturbances. However, there are no references in the literature to ependymal cysts related to psychotic symptoms. Those documented include confusion, disturbances of memory, and mental deterioration, mental slowness and poor concentration, irritability and a personality disturbance improved by the extirpation of a cyst exerting a mass effect in the right sylvian fissure. Some reports have indicated that the psychiatric symptoms related to other intracranial cysts, such as arachnoid or colloid cysts, have disappeared after extirpation of the cyst. However, almost all of the mentioned cases were accompanied by some neurological symptoms, and these cysts showed pronounced mass effects. The present case is the first report of an ependymal cyst in a patient without neurological symptoms, but with severe psychiatric symptoms (depressed mood, agitation, depersonalisation, and how the cyst resection resulted in a considerable improvement, see Pearfield’s1)), cannot be resolved in the context of the cervical sympathetic paralytic paradigm. Such an inverse temporal pattern between autonomic features and pain in cluster headache suggests another unrelated disease. Importantly, attacks of cluster headache that occurred between the ages of 25 to 33 years in this patient were not associated with Horner’s syndrome, and, the index pain attacks were identical; it would be useful to know if any subsequent cluster headache attacks are so associated with the usual temporal profile. The quest for a definitive diagnosis of the underlying cause of Horner’s syndrome is often fruitless. A theoretical possibility of viral infection, especially of herpes simplex virus—often found in the trigeminal, superior cervical sympathetic, and cranial parasympathetic ganglia—has been advanced; painful periodic activation of the virus may often occur without skin lesions. In a systemic influenza-like illness with accompanying viral vestibular neuronitis,1 it is entirely conceivable that a fortuitous concurrent self limited right sided viral superior cervical sympathetic ganglionitis was also associated. Virological studies might lend insight in otherwise inexplicable Horner’s syndrome. A pupillometric pattern consistent with a sympathetic third neuron pattern seems to be unusual in chronic lacrimation and cranial parasympathetic symptoms; a more frequent occasionally seen in only about 15% of patients.1 Influences other than sympathoparalytic also operate in producing the ptosis and miosis of cluster headache. Eyelid oedema in cluster headache and chronic paroxysmal hemicrania indicate that clinically significant ocular adnexal inflammation develops in both conditions, probably through antidromic ophthalmic nerve discharge. Eyelid oedema may close the eye mechanically—a form of pseudo or apparent ptosis.1 Such non-paralytic purely mechanical miosis may occur in cluster headache without miosis.1 Isolated inflammatory oedema of the upper eyelid in cluster headache (or chronic paroxysmal hemicrania) would seem itself to periodically exacerbate mechanical ptosis, as was manifest in this patient. Furthermore, intraocular pressure rises have been shown in both cluster headache (statistically insignificant) and chronic paroxysmal hemicrania; a biologically remarkable 298% increase in intraocular pressure was, however, shown on the symptomatic side in one subject with cluster headache.1 Intracocular prostaglandin or substance P release causes miosis and increase in intraocular pressure,1 constituting a form of ocularosympathetic reflex. Mechanical stimulation of the ophthalmic nerve through variations of intraocular pressure, both with and without stellate ganglionectomy, causes miosis through an antidromic reflex.8 Additionally, the interpretation of pupillary dilatation lag—tardy dilatation being generally assumed to denote ocular sympathetic deficit—must be guarded in conditions associated with pain because pain associated central sympathetic tone is likely to nullify such requirements. A well developed degree of central sympathetic tone.8 Importantly, the miosis of Horner’s syndrome is never maximal and is usually slight. Conversely, pain or emotional states or reduced intraocular pressure in both pupillary dilatation through the psychosensory reflex as well as neurogenic sympathetic lid retraction. These synergistic as well as antagonistic influences, besides residual ocular sympathetic deficit that outlasts the pain, dissociate pupillary miosis/dilatation lag and ptosis from each other as well as from the severity of individual cluster headache attacks. Intriguingly, in chronic paroxysmal hemicrania, HTN occurs without miosis. The development of ptosis with or without miosis in both cluster headache and chronic paroxysmal hemicrania cannot be simply assumed to reflect pure ocular sympathetic deficiency. The lack of salivation in disorders characterised by lacrimation and nasal congestion/rhinorhoea as well as the inconsistency of periorbital findings and postoperative results in connection with procedures directed on parasympathetic structures remain unexplained. Diffuse antidromic trigeminal nerve excitation9 also cannot explain the lack of salivation. The development of headaches associated with components of both migraine and cluster headache after gasserian ganglial ablation10 attenuates the possible role of activation of the nucleus salivatory.11 Lacrimal (ocular adnexal) gland and nasal innervation is associated with the branches of the ophthalmic nerve. Given an intracranial source of generation of trigeminal neural discharge through fluctuations of intracranial pressure lacrimation and nasal congestion/rhinorhoea in both cluster headache and chronic paroxysmal hemicrania may represent the effects of trigeminal neural discharge through sympathetic inhibition of the peripheral/local orthodromic-antidromic reflex driven phenomena. This concept obviates the need to invoke a theoretically unacceptable “selective” cranial parasympathetic barrage. Lacrimation and nasal congestion/rhinorhoea are not features of glaucoma in general. The rapidity of rise of intracranial pressure in cluster headache and chronic paroxysmal hemicrania less than 30 seconds in chronic paroxysmal hemicrania—might be critical to the triggering of an aberrant antidromic ophthalmic division trigeminal discharge that results in “automatic” manifestations.1

MATTERS ARISING

Painless Horner’s syndrome in cluster headache

Dissociation between autonomic dysfunction and pain during cluster headache, with the painless Horner’s syndrome preceding the headache attacks by a considerable interval (see Pearfield’s1), cannot be resolved in the context of the cervical sympathetic paralytic paradigm. Such an inverse temporal pattern between autonomic features and pain in cluster headache suggests another unrelated disease. Importantly, attacks of cluster headache that occurred between the ages of 25 to 33 years in this patient were not associated with Horner’s syndrome, and, the index pain attacks were identical; it would be useful to know if any subsequent cluster headache attacks are so associated with the usual temporal profile. The quest for a definitive diagnosis of the underlying cause of Horner’s syndrome is often fruitless.1 A theoretical possibility of viral infection, especially of herpes simplex virus—often found in the trigeminal, superior cervical sympathetic, and cranial parasympathetic ganglia—has been advanced; painful periodic activation of the virus may often occur without skin lesions.1 In a systemic influenza-like illness with accompanying viral vestibular neuronitis,1 it is entirely conceivable that a fortuitous concurrent self limited right sided viral superior cervical sympathetic ganglionitis was also associated. Virological studies might lend insight in otherwise inexplicable Horner’s syndrome. A pupillometric pattern consistent with a sympathetic third neuron pattern seems to be unusual in chronic lacrimation and cranial parasympathetic symptoms; a more frequent occasionally seen in only about 15% of patients.1 Influences other than sympathoparalytic also operate in producing the ptosis and miosis of cluster headache. Eyelid oedema in cluster headache and chronic paroxysmal hemicrania indicate that clinically significant ocular adnexal inflammation develops in both conditions, probably through antidromic ophthalmic nerve discharge. Eyelid oedema may close the eye mechanically—a form of pseudo or apparent ptosis.1 Such non-paralytic purely mechanical miosis may occur in cluster headache without miosis.1 Isolated inflammatory oedema of the upper eyelid in cluster headache (or chronic paroxysmal hemicrania) would seem itself to periodically exacerbate mechanical ptosis, as was manifest in this patient. Furthermore, intraocular pressure rises have been shown in both cluster headache (statistically insignificant) and chronic paroxysmal hemicrania; a biologically remarkable 298% increase in intraocular pressure was, however, shown on the symptomatic side in one subject with cluster headache.1 Intracocular

Arousal, relaxation, and autonomic tone.


Peatfield replies:

I wholly agree with Gupta that independent mechanisms for the headache and Horner's syndrome are required to explain the sequence of events in my patient. It is certainly possible the Horner's syndrome is entirely coincidental although the correlation suggests otherwise. Unfortunately, follow up information on my patient is not available.

I must confess to unwillingness to invoke a viral infection in parasympathetic ganglia to explain the pathogenesis of typical cluster headache, which even in this patient proved to be a spontaneously reversible periodic syndrome responding to corticosteroids among a wide variety of pharmacologically active agents.

Dr Gupta's second suggestion, that a purely mechanical process can be related to a rise in intracranial pressure, is certainly ingenious but I find it difficult to accept that a sudden rise in intracranial pressure sufficient to set up a trigeminal nerve mediated axon reflex would not first be painful.

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Non-invasive carotid imaging

We agree with most of the comments made by Sellar in his comprehensive review of imaging techniques for displaying blood vessels of the head and neck.1 However, by contrast with Sellar, we would argue that conventional angiography is no longer an essential component of the preoperative work up for prophylactic internal carotid endarterectomy; the combination of ultrasound and magnetic resonance angiography (MRA) is satisfactory for the vast majority of patients. In coming to this conclusion, we considered two key issues.

Firstly, to what risk do we expose patients with symptomatic carotid occlusive disease when we perform invasive, intra-arterial contrast angiography? Secondly, do non-invasive techniques reliably identify appropriate patients? (using currently available ECST criteria) this would be those patients with > 70% stenosis of the symptomatic internal carotid artery.

In assessing the risk of angiography, Sellar . . . suspect(s) that the complication rate remains at around 1% in most centres . . . , but provides no separate prospective evidence to support this figure. The risks of conventional angiography clearly vary according to which group of patients are studied. Patients with symptomatic carotid stenosis seem to be at greater risk than other groups and several studies have shown that the risk increases with increasing degrees of stenosis. This is particularly important because, in many centres, patients with mild disease are referred for angiography4 by non-invasive techniques such as ultrasound.

We therefore need to know the risks of angiography for the group of patients with moderate or severe stenosis, as these are the patients who will be exposed to the test. Few studies have looked specifically at this group.

In a prospective study of 200 such patients in our centre, Davies and Humphrey found a 4% stroke rate in 72 hours postangiography, with 2% death or serious disability.2 This figure intuitively seems high. However, it is not dissimilar to other studies, provided the appropriate patients are considered. In a study from Edinburgh, Hankey et al.3 reported an overall stroke rate of 2% with a permanent deficit rate of 1%.3 From their paper it is possible to calculate the postangiographic stroke rate in those patients with > 50% stenosis.

The failure, then, is an angiographic stroke risk of 5.5% and a permanent deficit risk of 3.1%. The benefits of carotid endarterectomy depend on a low surgical complication rate, as shown by the ECST and NASCET studies. Neither of these studies included the angiographic risk in their analyses. In borderline decisions, the risk of angiography, added to the surgical risk, could outweigh the benefits of endarterectomy.

The accuracy of non-invasive techniques is rightly assessed by comparison with invasive intra-arterial angiography, and Sellar is correct to state that we do not do this. This, in our opinion, is a gap in the current evaluation.

The necessity for angiography, however, is more clearly defined for those patients in whom the non-invasive investigations disagree significantly (an infrequent occurrence). In a situation where non-invasive methods clearly indicate the presence of a significant stenosis we do not think it is advisable to expose a patient to the risks of invasive angiography.

The concern raised by Sellar regarding the signal gaps that appear on magnetic resonance angiograms beyond a tight stenosis has not proved to be a problem in our experience. A non-invasive angiography is the issue of co-existent carotid siphon disease, which cannot be picked up reliably by duplex ultrasound. Our experience has been that significant siphon disease is a relatively uncommon finding. Furthermore, we are not convinced that its presence should alter the decision to perform endarterectomy, a similar conclusion to studies specifically looking at this question.

In our unit we now use a combination of ultrasound and MRA for preoperative assessment of patients, and, like others, have found this highly successful. We reserve conventional angiography for those patients in whom the non-invasive investigations disagree significantly (an infrequent occurrence). In a situation where non-invasive methods clearly indicate the presence of a significant stenosis we do not think it is advisable to expose a patient to the risks of invasive angiography.

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Painless Horner's syndrome in cluster headache.

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