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Short report

The association of hemifacial spasm and facial pain

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SUMMARY The recognition of an association between trigeminal neuralgia and ipsilateral hemifacial spasm has been delayed by confusion over the nomenclature of the two conditions. Three patients are presented who had facial pain associated with hemifacial spasm. The findings on investigation of these patients, and an analysis of the literature, suggests that the combination is almost inevitably associated with pathological processes in the posterior fossa, particularly anomalous, ectatic or aneurysmal blood vessels.

The readiness to attribute abnormal facial movements when accompanying trigeminal neuralgia to a non-specific reaction to a painful stimulus delayed the recognition of an association between trigeminal neuralgia and hemifacial spasm. Gowers[1] described the reflex muscle spasms occurring during the paroxysms of pain as tic convulsif, but Cushing[2] appreciated that tic convulsif was an alternative title for hemifacial spasm and suggested the term painful tic convulsif[3] for those patients with a combination of facial pain and hemifacial spasm. Soon afterwards, Harris[4] described patients in whom the association of hemifacial spasm and trigeminal neuralgia was beyond doubt. Confusion still exists in the literature over the use of the term tic convulsif, exemplified by one recent report which again applied it to the combination of facial pain and hemifacial spasm.[5]

We have encountered three patients who described facial pain ipsilateral to the side of their hemifacial spasm. The opportunity has been taken to appraise the literature of the condition and to discuss its pathogenesis.

Case reports

Patient 1, aged 63 years, was admitted to hospital in 1973. She gave an 18 month history of left hemifacial spasm, and a 12 year history of left deafness. For 2 months she had had attacks of sharp, left sided facial pain superimposed on a background of persistent discomfort. Examination revealed nystagmus, reduced pinprick on the left side of the face, left hemifacial spasm and a severe left perceptive deafness. Investigation established the presence of a left cerebellopontine angle mass. At operation, an extensive cholesteatoma was found, surrounding the fifth to the eleventh cranial nerves. Sub-total removal of the tumour was achieved with immediate resolution of the hemifacial spasm. Her clinical state deteriorated subsequently with evidence of increasing hydrocephalus and electrolyte imbalance. Despite ventriculoatrial shunting, she continued to deteriorate and died one month after surgery.

Patient 2, aged 57 years, was admitted to hospital in 1983. She had had left hemifacial spasm for 16 years. For 2 years, she had had paroxysmal facial pain affecting the ophthalmic and maxillary divisions of the left trigeminal nerve. The pains occurred up to four times a day. The development of a sensitivity reaction within 5 days of starting carbamazepine had prevented her from continuing with the drug. Examination confirmed a severe left hemifacial spasm accompanied by left facial weakness. There was a moderate left sensorineural deafness. Neuro-otological assessment indicated a bilateral high tone perceptive deafness, more marked on the left. Caloric responses showed a mild left canal paresis. Measurement of brain stem evoked responses was attempted but interference from the hemifacial spasm hindered analysis of the findings. CT scan was negative. At operation, the left seventh and eighth nerves were compressed by the posterior inferior cerebellar artery. The trigeminal nerve was compressed by a branch of the superior cerebellar artery. Both were dissected free. Following surgery, the hemifacial spasm and trigeminal neuralgia remitted and remained absent over a 3 year follow up period.

Patient 3, aged 52 years, seen in 1987, gave a three year history of trigeminal neuralgia affecting the maxillary division of the left trigeminal nerve. Over the same period, he had had left hemifacial spasm. There was no facial weakness. CT demonstrated ectasia of the left vertebral and basilar arteries, both of which looped to the left (fig). At operation, the trigeminal nerve was found to be compressed at the root-entry zone by an artery indenting it inferiorly. The seventh nerve was compressed by the ectatic vertebral artery. Both arteries were dissected away and held clear by cotton gauze. Post-operatively the hemifacial spasm and facial pain remit-

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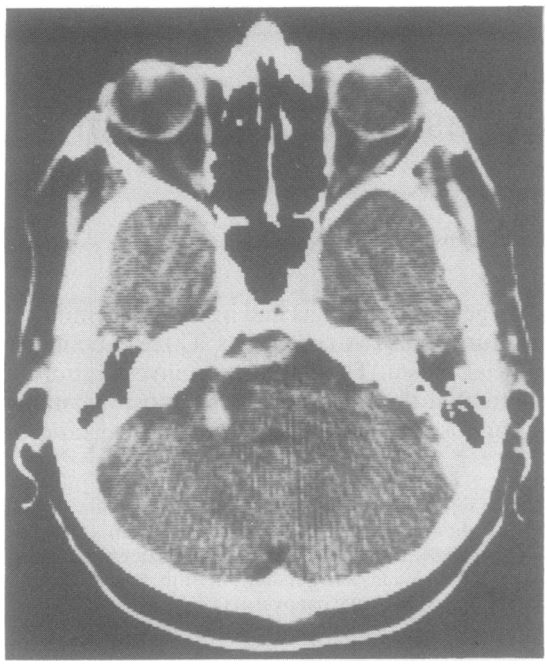

Fig *CT scan, after contrast, showing an ectatic and looped left vertebral artery.*

ted and had not returned during a 12 month follow up period.

Discussion

The term tic douloureux was originally introduced to describe the affliction of those patients with trigeminal neuralgia whose paroxysms of facial pain were accompanied by brief muscle contractions. Gowers used the term tic convulsif to describe these non-specific movements despite the fact that the term was already in use, in the French and German literature at least, to describe painless hemifacial spasm.[6 7]

Romberg reported a patient with episodic left sided facial pain, often triggered by shaving, who displayed convulsive movements of the face occurring independently of the painful paroxysms.[6] Post-mortem examination revealed thickening of the left vertebral artery, several dilatations of the basilar artery and a left cavernous aneurysm. Cushing described three patients with a combination of facial pain and hemifacial spasm,[3] though he was not confident that the pain was that of trigeminal neuralgia since at least one of the cases had failed to respond to trigeminal section. He believed that his patients were describing geniculate neuralgia perhaps influenced by the then recent description of paroxysmal aural pain relieved by removal of the geniculate ganglion.[8] If geniculate neuralgia exists, it is characterised by paroxysmal aural pain associated with a trigger zone.[9] Cushing's cases hardly fulfilled these criteria, two of them experiencing pain in all three divisions of the trigeminal nerve. Modern authors have often been lax in defining geniculate neuralgia, including patients with dull persistent pain centred on the ear.[5 10] A combination of true trigeminal neuralgia and hemifacial spasm was encountered by Harris and Wright[4] in at least 12 cases. Ehni and Woltman, reviewing over 100 cases of hemifacial spasm, described three with trigeminal neuralgia ipsilateral to the spasm.[11] No consistent time relationship was found between the onset of the pain and the facial movement.

Gardner commented on a number of similarities between trigeminal neuralgia and hemifacial spasm.[12] In his experience, the symptoms, when occurring in combination, were always ipsilateral. Relief of one, by alcohol injection of the relevant ganglion, failed to influence the other. He suggested that the conditions might be triggered by similar pathological mechanisms. Romberg[6] believed that a lesion at the base of the brain, by stimulating the seventh nerve, could trigger attacks of hemifacial spasm. Subsequently, Schultze[7] reported a male with painless hemifacial spasm of a few months duration in whom necropsy revealed a left vertebral aneurysm displacing the seventh and eighth cranial nerves. Ramsay Hunt's[9] collection of cases of hemifacial spasm included examples triggered by small tumours or aneurysms in proximity to the facial nerve. Cases due to tumour were subsequently reported by Weisenburg[13] and Cushing.[2] Among 19 patients, Gardner and Sava[14] found seven in whom the facial nerve was compressed by the internal auditory or anterior inferior cerebellar artery. Of the 47 cases recorded by Jannetta *et al*,[15] 46 had evidence of vascular compression or distortion of the root exit zone of the facial nerve.

The recognition of a corresponding association between trigeminal neuralgia and compression of the fifth cranial nerve was established beyond doubt by Dandy who showed that a proportion of patients with trigeminal neuralgia had vascular compression of the fifth cranial nerve in the posterior fossa.[16] A third of his cases showed distortion of the trigeminal nerve by the superior cerebellar artery. More recently, Jannetta[17] and Apfelbaum[18] have shown that vascular compression of the fifth cranial nerve is present in most patients with trigeminal neuralgia.

Limited pathological data exist for those patients with a history of combined hemifacial spasm and trigeminal neuralgia. Post-mortem examination was performed in only one of Cushing's cases and revealed no abnormality.[3] Both the patients described by Campbell and Keedy[19] had trigeminal neuralgia. In one the pain had been followed, in the other preceded, by ipsilateral hemifacial spasm. At operation, both

patients were found to have a cirsoid aneurysm of the basilar artery. Gardner[12] encountered four examples of this clinical combination. Two had cirsoid basilar aneurysms, and two an arteriovenous malformation occupying the cerebello-pontine angle. One of the patients reported by Kempe and Smith[20] had a combination of hemifacial spasm and a paroxysmal ear and throat pain considered to represent glossopharyngeal neuralgia. At necropsy a large anomalous vessel, possibly a primitive acoustic artery, was found to be stretching the seventh and eighth cranial nerves.

All the three patients reported here had left hemifacial spasm. In two, the spasm was associated with ipsilateral trigeminal neuralgia, the third patient having a less specific, though ipsilateral, facial pain. The pathological basis for the clinical picture differed in each patient. One had a cholesteatoma, one ectatic vertebral and basilar arteries and the third compression of the fifth, seventh and eighth cranial nerves by the superior and posterior inferior cerebellar arteries.

Hemifacial spasm can exist in isolation, or in association with facial pain. In some patients, the pain has no specific features, in others it has the characteristics of trigeminal, glossopharyngeal or geniculate neuralgia. Exploration of the posterior fossa almost inevitably reveals compression or distortion of the relevant cranial nerve. Rarely, this is due to tumour, more often the consequence of an anomalous, ectatic or frankly aneurysmal vessel. In the absence of an alternative, the term suggested by Cushing, painful tic convulsif, should be used for the clinical syndrome of hemifacial spasm and trigeminal neuralgia.

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