Painful tic convulsif

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SUMMARY Painful tic convulsif is a syndrome restricted to paroxysmal dysfunction of the fifth and seventh cranial nerves. It occurs primarily in women over the age of 50 years and is usually associated with an ectatic vertebrobasilar artery—less frequently an arteriovenous malformation or cholesteatoma—which compresses the trigeminal and facial nerve roots in the posterior fossa. In rare instances this syndrome may be caused by disseminated sclerosis. Because of the high incidence of posterior fossa lesions in painful tic convulsif, a complete neurological evaluation including computerised transaxial tomography and vertebrobasilar angiography should be performed in every case.

The term 'painful tic convulsif' was coined by Cushing to describe the association between hemifacial spasm and ipsilateral facial pain. Since his original description in 1920, a total of 41 patients with both trigeminal neuralgia and hemifacial spasm have been reported. The largest series, 14 cases reported by Harris (1940), provides no clue as to the aetiology of this syndrome. Campbell and Keedy (1947) operated on two patients with painful tic convulsif; in both cases a cirroid aneurysm of the basilar artery compressed the fifth and seventh cranial nerves. Subsequently, Gardner (1968) reported 11 patients with the syndrome, all of whom had documented posterior fossa lesions (cirroid aneurysms, arteriovenous malformations, and cholesteatomas). Maurice-Williams (1973) has re-emphasised that the coexistence of trigeminal neuralgia and hemifacial spasm signals the presence of a posterior fossa lesion.

We recently examined a patient with painful tic convulsif and vertebrobasilar ectasia and, in consequence, undertook a systematic review of the literature. We discovered that less than half of the 41 reported cases provide sufficient clinical data for analysis and that only 10 patients with painful tic convulsif have been subjected to angiography, surgery, or postmortem examination. Moreover, although the signs and symptoms of patients suffering from it have been described, no attempt has yet been made to define the clinical syndrome or to discuss its diagnosis or the results of treatment.

Case report

A 67 year old woman was admitted to The New York Hospital with a five years' history of paroxysmal lancinating pain over the right jaw, radiating posteriorly to the ear. Paroxysms of pain were triggered by talking, chewing, or palpating the skin over the ramus of the right mandible; spontaneous paroxysms also occurred. Approximately two years after the onset of right-sided facial pain, she became aware of twitching in the right periorbital muscles, which spread during the ensuing months to involve the majority of the muscles innervated by the right facial nerve. These hemifacial spasms occurred without warning, could not be controlled by conscious effort, and were exacerbated by anxiety. There was no temporal relation between her right-sided facial pain and the attacks of facial hemispasm.

The patient complained of mild bilateral hearing loss but denied tinnitus or vertigo. There was no history of headache, visual symptoms, facial paralysis, dysphagia, dysarthria, limb weakness, incoordination, sensory disturbance, or gait disorder.

The general medical examination was unremarkable. Neurological examination revealed a right facial contracture (Fig. 1a) and spasmodic, irregular mass contractions of the right facial muscles, including the orbicularis oculi, levator anguli oris, and platysma. Both corneal reflexes were intact, and facial sensation was normal. Mild bilateral sensorineural deafness was confirmed by audiometry; Hallpike differential caloric tests were normal.

Routine laboratory investigations, including serum electrolytes, blood urea nitrogen, fasting blood sugar, VDRL, liver function tests and complete

Fig. 1 (a) Right facial contracture before facial nerve block; (b) 'Release' of facial contracture after lidocaine block at the stylomastoid foramen.

blood count, were normal. Skull radiographs with special views of the internal auditory canals were also normal, as was a technetium brain scan. Electromyography of the right facial muscles failed to reveal any evidence of denervation or of lower motor neurone dysfunction. Infiltration of the right facial nerve at the stylomastoid foramen with 1% lidocaine hydrochloride produced an immediate flaccid paralysis of the right side of the face with cessation of the spasmodic contractions and 'release' of the right facial contracture (Fig. 1b). Computerised transaxial tomography (CTT) without intravenous contrast medium was normal; an ill-defined linear density in the right cerebellopontine angle, seen only after the administration of intravenous contrast medium, was thought to represent an ectopic vessel. Retrograde right brachial arteriography demonstrated a markedly ectatic right vertebral artery which appeared to impinge upon the right facial nerve in the cerebellopontine angle (Fig. 2) and the right trigeminal nerve in the cisterna ambiens (Fig. 3).

Diphenylhydantoin and carbamazepine produced prompt relief of her facial pain but no noticeable effect on the hemifacial spasm. Two months after discharge the patient remained pain-free on carbamazepine, although her hemifacial spasm persisted as before.

Discussion

Because of the paucity of clinical data available in the literature, it is impossible to draw any conclusions

which apply to all of the reported cases of painful tic convulsif. Of the 34 patients whose sex is known, 25 are female, nearly a 3:1 predominance. Symptoms occurred with equal frequency on the right (10/19) and left (9/19) sides of the face, and Penman (1968) reported a single case of bilateral painful tic convulsif.

The onset of the syndrome may be heralded by either hemifacial spasm or trigeminal neuralgia. The average age at onset of either symptom was 54 years, an equal number of patients presenting with trigeminal neuralgia (7/13) and hemifacial spasm (6/13). The interval separating the onset of the two symptoms varied from a few months to 25 years.

The signs and symptoms of painful tic convulsif as described in the literature were limited to the fifth, seventh, and, occasionally, the eighth cranial nerves. Hemifacial spasm characteristically began in the orbicularis oculi muscle and spread gradually to involve the other muscles of facial expression on the ipsilateral side. The spasms were not under voluntary control, occasionally persisted during sleep and were exacerbated by anxiety. Clinically, the hemifacial spasm was identical with the idiopathic variety described by Ehni and Woltman (1945). Trigeminal neuralgia in patients with painful tic convulsif most frequently implicated the third division of the trigeminal nerve, less frequently the first or second divisions. Typically, there was no temporal relationship between the paroxysms of facial pain and the hemifacial spasm. Eighth nerve dysfunction, although not a consistent finding, was

Fig. 2 Subtracted right brachial arteriogram, anterior-posterior view. Note loop of right vertebral artery (large arrow) projecting into the right cerebellopontine angle. Small arrows delineate the right vertebral artery.

Fig. 3 Unsubtracted right brachial arteriogram, lateral view. Arrows delineate a redundant loop of the right vertebral artery in the right cerebellopontine angle. Note also the anomalous course of the first portion of the basilar artery.

evidenced by hearing loss, nystagmus, and/or abnormal oculovestibular responses on the affected side.

Only 10 case reports (Table) provided angiographic, surgical, or postmortem data bearing on the aetiology of painful tic convulsif. Within this group there were six cirroid aneurysms, two arteriovenous malformations, and two cholesteatomas. Two patients with hemifacial spasm and ipsilateral facial pain reported by Cushing (1920) were excluded from this series because Cushing clearly states that neither patient suffered from 'true trigeminal neuralgia'; moreover, gasserian ganglionectomy failed to relieve

their facial pain, and neither patient had a demonstrable posterior fossa lesion at postmortem examination.

Maurice-Williams (1973) described two patients with hemifacial spasm and ipsilateral trigeminal neuralgia. His first patient had clear-cut cerebellar signs and was therefore atypical, and his second patient was not subjected to angiography. Both patients were excluded from the series. Two other patients—one with a cirroid aneurysm and one with a cholesteatoma—were excluded because the only documentation of their illness was passing reference in a paper by Gardner (1968).

The facial pain of painful tic convulsif was usually relieved by diphenylhydantoin, carbamazepine, or a combination of the two. Neither of these medications relieved the hemifacial spasm, however. Ethanol injection of the gasserian ganglion temporarily relieved some patients of their trigeminal neuralgia but had no effect on their hemifacial spasm. Successful treatment of hemifacial spasm in painful tic convulsif—that is, alleviation of spasm with preservation of seventh nerve function—has been achieved only by Gardner (1968), whose operation involves saline neurolysis of the facial nerve within the posterior fossa.

Table Posterior fossa lesions associated with painful tic convulsif

Abnormality	No.	Evidence	Reference
Cirroid aneurysms			
Ectatic basilar artery	4	Surgery	Campbell and Keedy (1947) Gardner (1968)
Ectatic vertebral artery	2	Angiography	This report Gardner (1968) Gardner (1968)
Arteriovenous malformations	2	Surgery	Gardner (1968)
Cholesteatomas	2	Surgery	Revilla (1947) Gardner (1968)

Although Cushing is credited with the *princeps* description of painful tic convulsif, it was Campbell and Keedy (1947) who first drew attention to a possible common aetiology for the trigeminal neuralgia and ipsilateral hemifacial spasm. Gardner has since discussed 'symptomatic' trigeminal neuralgia, 'symptomatic' hemifacial spasm and the combined syndrome. He points out that the incidence of posterior fossa lesions is remarkably high in all three conditions and that the nature of the responsible lesion is strikingly similar. Based on the well-documented cases presented in the Table, the lesions responsible for painful tic convulsif are almost always vascular in nature, cirroid aneurysms of the vertebral or basilar arteries being the most common. These aneurysms were best described by Dandy (1944) as 'S-shaped elongations of the arteriosclerotic basilar and/or vertebral arteries, with consequent bulging that compresses the fifth or the eighth nerve or both'. Dandy operated on 11 patients with vertebral or basilar ectasia, many of whom had trigeminal neuralgia or vertigo; however, none of Dandy's patients had painful tic convulsif.

A most intriguing case of the disorder was reported by Ehni and Woltman in 1945. Their patient was a 21 year old man with right sided trigeminal neuralgia who obtained complete pain relief for four years from an alcohol injection of the right fifth nerve. Three months after the tic recurred, 'he noted a slackening of the pain, though he was not undergoing any treatment, and as the pain disappeared there began an intermittent twitching of the right eyelid, which continued without modification for 18 months'.

The onset of trigeminal neuralgia at age 21 years is distinctly unusual and suggests the associated diagnosis of multiple sclerosis. Hemifacial spasm may also usher in an attack of multiple sclerosis (Thiébaud *et al.*, 1960), and, thus, the possibility exists that the cause of painful tic convulsif in Ehni and Woltman's patient was multiple sclerosis rather than a posterior fossa lesion.

The need for angiographic examination of the posterior fossa in patients with painful tic convulsif is self-evident. Even a CTT scan after the administration of intravenous contrast material may be inadequate to demonstrate the vascular anomaly. Surprisingly, in only two cases—our case and Gardner's case 6 (1968)—has the causative lesion been identified by arteriography. In both instances an ectatic vertebral artery was found. Gardner states that it is often difficult to distinguish the basilar from the vertebral

artery at surgery when the 'routine cerebellar approach' is used. He suggests that in those cases of painful tic convulsif explored surgically without previous angiography, vertebral ectasia was not appreciated and basilar ectasia misdiagnosed.

Gardner and Dohn (1965) have suggested that trigeminal neuralgia and hemifacial spasm result from ephaptic conduction in partially demyelinated axons. Both multiple sclerosis and vertebrobasilar ectasia can produce demyelination, the latter by direct compression of nerve fibres, and both have been implicated in the pathogenesis of typical trigeminal neuralgia and hemifacial spasm. Our case and a critical review of the literature lead us to conclude that in older patients painful tic convulsif is caused primarily by a vascular anomaly in the posterior fossa—usually vertebrobasilar ectasia—and that multiple sclerosis may be responsible for the rare occurrence of this syndrome in younger patients.

References

- Campbell, E., and Keedy, C. (1947). Hemifacial spasm: A note on the etiology in two cases. *Journal of Neurosurgery*, **4**, 342–347.
- Cushing, H. (1920). The major trigeminal neuralgias and their surgical treatment. *American Journal of the Medical Sciences*, **160**, 157–184.
- Dandy, W. E. (1944). *Intracranial Arterial Aneurysms*, pp. 60–66. Haffner: New York.
- Ehni, G., and Woltman, H. W. (1945). Hemifacial spasm. *Archives of Neurology and Psychiatry (Chic.)*, **53**, 205–211.
- Gardner, W. J. (1968). Trigeminal neuralgia. *Clinical Neurosurgery*, **15**, 1–56.
- Gardner, W. J., and Dohn, D. F. (1965). Trigeminal neuralgia-hemifacial spasm—Paget's disease: significance of this association. *Brain*, **86**, 555–562.
- Harris, W. (1940). An analysis of 1433 cases of paroxysmal trigeminal neuralgia. *Brain*, **63**, 209–224.
- Maurice-Williams, R. S. (1973). Tic convulsif: the association of trigeminal neuralgia and hemifacial spasm. *Postgraduate Medical Journal*, **49**, 742–745.
- Penman, J. (1968). Trigeminal neuralgia. In *Handbook of Clinical Neurology*, vol. 5, pp. 296–322. Edited by P. J. Vinken and G. W. Bruyn. Wiley: New York.
- Revilla, A. G. (1947). Tic douloureux and its relationship to tumors of the posterior fossa. *Journal of Neurosurgery*, **4**, 233–239.
- Thiébaud, F., Isch, F., Isch-Treussard, C., and Ebtinger-Jouffroy, J. (1960). Hemispasme faciale dans la sclérose en plaques. *Revue Oto-Neuro-Ophthalmologique*, **32**, 162–166.