References


Transient hemiparesis—a cautionary tale: coexistence of pheochromocytoma and intracranial aneurysm

Sir: The following salutary case history may prove of interest to your readers. A 40-year-old right-handed caucasian male experienced sudden onset of tinnitus one evening. The following morning, he noted marked weakness and numbness of his left arm and leg and was admitted to hospital. On examination, he had a left hemiparesis, sparing the face, and impaired sensation to pin prick and light touch over his left arm and leg. He was hypertensive (BP 190/100 mm Hg) and treated with hydralazine and a beta blocker. The hemiparesis completely resolved after three days. The patient’s haematocrit, serum urea and electrolytes, serum cholesterol, fasting lipids and blood glucose were normal. Whilst the results of urinalysis for catecholamines were awaited, additional investigations revealed normal skull and abdominal X-rays; a normal intravenous urogram and a normal head computerised tomogram (CT scan). Lumbar puncture yielded clear cerebrospinal fluid (csf) with no red cells, less than 1 white cell and 30 mg% of protein. A right carotid angiogram demonstrated an aneurysm of the right middle cerebral artery but was otherwise normal. Surgical obliteration of the aneurysm was recommended. The patient requested transfer to a centre nearer his home prior to surgery and he was, therefore, referred to our unit. The diagnosis at this stage was (1) hypertension (2) prolonged reversible ischaemic neurological deficit (3) aneurysm of the right middle cerebral artery.

On arrival, the patient had no abnormal neurological signs. After withdrawing his anti-hypertensive therapy, he remained normotensive. In view of the clinical picture, a complete series of intracranial angiograms including the neck vessels was requested. The patient was cooperative and angiography was performed under mild sedation. However, attempted femoral catheterisation was associated with the onset of hypertension (BP 270/120 mm Hg) and the examination was aborted. It was thought that anxiety was responsible for the hypertensive episode although the patient denied this. His blood pressure returned to normal as soon as he returned to the ward. A repeated attempt at angiography under heavier sedation (Pethidine 100 mg, Droperidol 6 mg) was again aborted because of a hypertensive episode (BP 250/130 mm Hg). Urinalysis for catecholamines was requested since those performed at the referring hospital were still unavailable. Direct questioning did not reveal any history of headaches, sweating or palpitations. It was decided to perform angiography under general anaesthesia. Soon after anaesthetic induction, a hypertensive crisis occurred (BP 240/150 mm Hg) which was accompanied by sweating, salivation and ventricular extrasystoles. Prompt treatment (10 mg intravenous phentolamine) by the vigilant anaesthetist controlled this. Angiography did not reveal any further vascular abnormality. After repeated direct questioning of the patient, we obtained a history of occasional headaches, nocturnal sweating attacks and palpitations—none of which had he considered significant enough to warrant divulging to the admitting doctor. A provisional diagnosis of pheochromocytoma was made and confirmed later by the results of our urinalysis for catecholamines (18-7 \(\mu\)mol normetadrenaline/24 hr). The patient’s blood pressure was controlled with Propranolol and Phentolamine. A tumour of the right adrenal gland was demonstrated by whole body CT and this was subsequently removed. Histological examination confirmed the diagnosis of pheochromocytoma. The intracranial aneurysm was successfully clipped at a later date. There was no evidence of a previous haemorrhage from the aneurysm when it was displayed at operation.

We are not aware of a reported association between intracranial aneurysms and pheochromocytomas, although hypertension may contribute to the development of aneurysms. Usually, an intracranial aneurysm is identified as a cause of a subarachnoid haemorrhage although on occasions the presentation may be that of a posterior fossa tumour or transient cerebral ischaemia. Vascular anomalies of the posterior fossa, including a basilar artery aneurysm may present with episodic hypertension and mimic a pheochromocytoma as may posterior fossa tumours. Pheochromocytoma usually causes episodic or sustained hypertension but it may present with cortical blindness or transient neurological deficits. The mechanism responsible for the latter has not been elucidated but it is possible that arterial spasm, probably involving perforating vessels arising from the proximal middle cerebral artery, may have been responsible. The transient neurological deficit that our patient experienced. Interestingly, a recent report identified large numbers of neuropeptide Y (NPY) producing cells in pheochromocytomas and plasma NPY levels were raised in patients with pheochromocytomas. NPY has been shown to have vasoconstrictor activity and it is possible that release of this neuropeptide may have contributed to impaired cerebral perfusion in our patient but we admit that this explanation is speculative.

This case study illustrates the pitfalls which may be encountered when two coincidental dangerous pathological conditions are encountered in the same patient.
Letters

References
1 Stehbens WE. Pathology of the cerebral blood vessels. The CV Mosley Company, St Louis, 1972:374.

Hemifacial spasm associated with a mixed benign parotid tumour

Sir: Hemifacial spasm is characterised by clonic, tonic or tonicoclonic contractions of the muscles innervated by the facial nerve.1 These painless, involuntary and inexpressive contractions generally first appear in the orbicularis oculi, increase progressively in severity and then spread downward to include the entire hemiface. The contractions persist during sleep and cannot be voluntarily suppressed.1 The term “facial tic” although inappropriate1 is sometimes used.2 Paradoxical synkinesis between the orbicularis oculi and the frontalis is pathognomonic of hemifacial spasm and the incurvation of the nose, the chin dimple and the movement of the external ear are also characteristic.3 The pathophysiology of this phenomenon remains obscure. We report a case of hemifacial spasm with an unusual aetiology.

In August 1980, a 70-year-old Caucasian male presented with clonic contractions of the orbicularis oculi and the upper lip levator muscle of the left face. Treatment with carbamazepine proved ineffective and was poorly tolerated with nausea and vomiting. Two months later, the contractions involved all the muscles, including platysma, innervated by the left facial nerve. Examination showed left hemifacial spasm with painless, involuntary, persistent clonic contractions worsened by emotional upset, weariness, intentional and automatic movement, remaining when sleeping. Paradoxical synkinesis was observed between the orbicularis oris and the frontalis; the latter was practically always contracted with an elevation of the eyebrow. Neurological examination was otherwise negative. Since the patient had no history of facial palsy, the diagnosis of essential hemifacial spasm was made. There was no facial weakness. Schirmer’s test, stapedius and blink reflex were normal as were recording of orbicularis oris potential after facial nerve stimulation at the stylomastoid foramen. Electromyography recorded sudden spontaneous synkinetic bursts of activity, synchronous in the orbicularis oris and the upper lip levator muscles. They were increased by the contraction of the frontalis muscle in which there was no spontaneous activity.

Physical examination revealed a left parotid tumour. It had been diagnosed as a cyst twenty years before, and had increased in volume one month before the onset of the hemifacial spasm. The tumour was homogenously firm, smooth, freely movable in all directions, without an increase in local temperature. Palpation was slightly painful. The clinical diagnosis was that of a mixed tumour of the parotid gland. Sialography showed a rounded elongation of the canal in the inferoposterior segment of the gland, an increase in glandular volume and integrity of Stenon’s canal. Puncture revealed epithelial salivary cells with a light blue cytoplasm in a radial arrangement or isolated in a thick myxoid substance. A total parotidectomy was performed (PhP). After dissection of the subchondrial tissue, the facial trunk was discovered in a very internal position and was dissected. The superficial lobe, deformed by a large tumour, was extracted from the parotid. Ligature of the external carotid artery and the internal maxillary artery preceded the dissection and excision of the internal lobe. The facial nerve was paler than normal, with an ischaemic appearance. Anatomopathological examination of the excised tumour confirmed the diagnosis of a “benign mixed tumour” without malignant change. Postoperatively, the hemifacial spasm immediately diminished and disappeared within 8 days. Six months later, the patient had no complaints but a few rare clonic Jerks of the orbicularis oris and of the orbicularis oculi as well as a tonic contraction of the left frontalis were observed. Electromyography was normal.

Hemifacial spasm can be post-paralytic or essential,4 in which case it is either symptomatic or cryptogenic. Cryptogenic hemifacial spasm was diagnosed most frequently as in 43 of 59 cases of Alajouanine and Thuret.5 Long considered specific to adults,6 it may exceptionally be found in children.4 Classically, essential hemifacial spasm, when symptomatic, implicated an inflammatory8,10 or “space occupying” lesion in the cerebello-pontine angle. Malformations of the atlanto-occipital joint,11 and Parkinson’s disease9 have also been described as possible aetiologies. In 1962, Gardner12 emphasised the role of vascular malformations or positional abnormalities which were observed in 13 of the 19 patients he had operated on. Since then, other authors9,13-14 have confirmed the role of enlarged-arteries, arterial or arteriovenous aneurysms, venous malposition, even a persistent embryonic artery, all of the vertebrobasilar system. The association of an arterial malformation has been used to explain the hemifacial spasm observed in malformations of the atlanto-occipital joint.14 The frequency of a vascular aetiology (46 cases of 47 patients of Jannetta15) has led some authors to propose surgical exploration of the cerebello-pontine angle even if there was no evident vascular cause.20 Exploration, however, has not definitively proved the validity of the traditional hypotheses evoked to explain hemifacial spasm: simple compression, nerve ischaemia, aberrant regeneration, or false synapse formation (“ephaphes”) between fibres near a lesion which is often secondary to a vascular compression. The site of compression probably is at the emergence of the facial nerve from the pons where glia gives way to the myelin sheath.16 The mechanism in hemifacial spasm of otological origin also could be due to the false synapses. More recently, it has been postulated that a peripheral axonal injury could “unmask and augment automatic, associated, and reflexive movements already present in the facial neural network.”21

Hemifacial spasm due to parotid lesions is extremely rare and is not mentioned as a complication of parotid tumours.23,24 To our knowledge, only one case of parotid actinomyositis and one case of a mixed