Hemifacial spasm associated with a mixed benign parotid tumour

Sir: Hemifacial spasm is characterised by clonic, tonic or tonico-clonic contractions of the muscles innervated by the facial nerve.3 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 hemisphere. 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 inervation 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 sub-perichondrial 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. Anatomopathologic 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,1 in which case it is either symptomatic or cryptogenic. Cryptogenic hemifacial spasm was diagnosed most frequently in both cases of Alaouaine and Thurel.1 Long considered specific to adults,7 it may exceptionally be found in children.8 Classically, essential hemifacial spasm, when symptomatic, implicated an inflammatory9 or "space occupying" lesion in the cerebello-pontine angle. Malformation of the atlanto-occipital joint,10 11 and Parkes' disease with ataxia,12 13 have also been described as possible aetiologies. In 1962, Gardner14 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 authors15 16 have confirmed the role of enlarged-arteries, arterial or arteriovenous aneurysms, venous malposition, even a persistent embryonic artery, all of the verteobasilar system. The association of an arterial malformation has been used to explain the hemifacial spasm observed in malformations of the atlanto-occipital joint.14 They suggested a frequency of a vascular aetiology (46 cases of 47 patients of Jannetta18) has led some authors to propose surgical exploration of the cerebello-pontine angle even if there was no evident vascular cause.20 Explora- tion, 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 ("ephapses") 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.21 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 neuronal network.22

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 actino-mycosis7 and one case of a mixed...
Recurrent intracranial haemorrhage due to Behcet disease

Sir: Vascular lesions are one of the common complications of Behcet disease. Most of the vascular lesions are considered to result in an occlusive process or aneurysm formation in the large vessels. There has been limited evidence for the intracranial site of these vascular lesions. According to the extensive study by Maeda and Nakao, however, 9% of patients with Behcet disease died of cerebrovascular disease, which is the third leading direct cause of death following Behcet disease itself (41-6%) and heart disease (11-2%). This suggests that cerebrovascular disease may occur in a significant number of the patients with Behcet disease. I report a patient with a history of Behcet disease and hypertension who had repeated massive intracranial haemorrhages three times.

A 50-year-old Japanese man was admitted complaining of severe headache and vomiting. His past history included a recurrent ocular disturbance, aphthous stomatitis and genital ulcer since he was 18 years old. He was diagnosed as having Behcet disease by an ophthalmologist when he was 42 years old. At that time he also had hypertension. He became totally blind at the age of 44 years. He noticed a mild right hemiparesis when he was 46 years old, and a mild dysarthria followed two years later. When the patient was 49 years old, he was hospitalised with a diagnosis of arachnoid cyst in the left frontal lobe which was confirmed by operation. He was discharged with a mild right hemiparesis and dysarthria. Upon the patient's admission, the patient was responsive to verbal commands, and had marked cerebellar ataxia in the left extremities. He was totally blind but there was neither obvious aphthous stomatitis nor genital ulcer. The blood pressure was 240/120 mm Hg. The CT scan revealed a massive haemorrhage in the left cerebellar hemisphere. The laboratory examination demonstrated a haemoglobin of 11-2 g/dl; RBC 4,440,000/cu mm; WBC 15,095/cu mm; platelets 230,000/cu mm; haematocrit 40%. Fibrinogen was 226 mg/dl and FDP was 4 μg/ml. Bleeding time was 1 ' 15; Clotting time was 10' 30'. The maximum rate of platelet aggregation induced by ADP (1 μM) was 80%. There was no evidence of a haemorrhagic diathesis in the laboratory data. Blood sugar was 140 mg/dl. Sodium was 143 mEq/l; Potassium 26 mEq/l; Chlor 107 mEq/l. The protein value was 7-7 g/l. The ECG was within...