parotid tumour²⁷ have been reported. In these two reports, as well as in our own patient, hemifacial spasm might have been due to a direct compression with local ischaemia as suggested by the appearance of the facial nerve at surgery. The role of functional reorganisation in the facial nucleus, suggested by Ferguson²² and emphasised by Martinelli,²⁸ for hemifacial spasm after injury of a peripheral branch of the facial nerve cannot be excluded, nor can false synapses between motor fibres.

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References

- Meige H. Les convulsions de la face. *Rev Neurol (Paris)* 1910;21:437-43.
- Harrison MS. The facial tics. *Laryngol Otol* 1976;90:561-70.
- Babinski J. Hémispasme facial périphérique. *Rev Neurol (Paris)* 1905;13:151-8.
- Jesell M. Hémispasme facial périphérique. Contribution clinique et E.M.G. à l'étude des hémispasmes faciaux et au diagnostic différentiel des contractures et hypercinnés faciales. *Thèse Med Paris* 1965.
- Alajouanine T, Thurel R. *Les Spasmes de la Face et leur Traitement*. Paris: Masson, 1936.
- Ehni G, Woltman HW. Hemifacial spasm: Review of 106 cases. *Arch Neurol Psychiat* 1945;53:205-11.
- Gardner WJ. Cross talk—the paradoxical transmission of a nerve impulse. *Arch Neurol* 1966;14:149-56.
- Langston JW, Tharp BR. Infantile hemifacial spasm. *Arch Neurol* 1976;33:302-3.
- Laine E. Hémispasme facial guéri par intervention sur la fosse postérieure. *Rev Neurol (Paris)* 1948;80:38-40.
- Laine E, Graux P, Fontan M. A propos de deux nouvelles observations d'hémispasme facial guéri par intervention sur une arachnoïdite de l'angle ponto-cérébelleux. *Rev Neurol (Paris)* 1951;84:82-4.
- Arnould G, Tridon P, Laxenaire M, Barrucand D. Hémispasme facial et malformation de la charnière occipito-vertébrale. *Rev Otoneuroophthalmol* 1962;34:1-5.
- Boudouresques J, Roger J, Vigouroux RA, Gouin M, Duport P, Daniel F. Manifestations neurologiques multiples (dont un hémispasme facial) au cours d'une maladie de Paget avec impression basilaire. *Marseille Med* 1969;106:67-9.
- Gardner WJ, Sava GA. Hemifacial spasm. A reversible pathophysiologic state. *J Neurosurg* 1962;19:240-7.
- Carella A, Caruso G, Lamberti P. Hemifacial Spasm due to elongation and Ectasia of the Distal Segment of the vertebral artery. *Neuroradiology* 1973;6:233-6.
- Nappi G, Moglia A, Poloni M, Arrigo A. Hemifacial Spasm Associated with dolichomegavertebrobasilar anomaly. *Eur Neurol* 1977;15:94-101.
- Jannetta PJ, Abbasy M, Maroon JC, Ramos FM, Albin MS. Etiology and definitive microsurgical treatment of hemifacial spasm. Operative techniques and results in 47 patients. *J Neurosurg* 1977;47:321-8.
- Maroon JC. Hemifacial spasm: a vascular cause. *Arch Neurol* 1978;35:481-3.
- Rohmer F, Heldt N, Coquillat G, Thiebaut JB. Hémispasme facial par compression vasculaire. *Rev Otoneuroophthalmol* 1979;51:243-5.
- Pierry A, Cameron M. Clonic hemifacial spasm from posterior arterio venous malformation. *J Neurol Neurosurg Psychiatry* 1979;42:670-2.
- Fabinyi GCA, Adams CBT. Hemifacial spasm: treatment by posterior fossa surgery. *J Neurol Neurosurg Psychiatry* 1978;41:829-33.
- Woltman HW, Williams HL, Lambert EH. An attempt to relieve hemifacial spasm by neurolysis of the facial nerve: a report of two cases of hemifacial spasm with reflections on the nature of the spasm, the contracture and mass movement. *Proc Mayo Clin* 1951;26:236-40.
- Ferguson JH. Hemifacial spasm and the facial nucleus. *Ann Neurol* 1978;4:97-103.
- Redon H. *Chirurgie des Glandes Salivaires*. Paris: Masson, 1955, 275.
- Krolls SO, Boyers RC. Mixed tumors of salivary glands. Long-term follow-up. *Cancer* 1972;30:276-81.
- Thackray AC, Lucas RB. Tumors of the major salivary glands. In: *Atlas of Tumor Pathology*, IInd series. Washington: Armed Forces Institute of Pathology 1974; fascicle 10.
- Gandon J, Trotoux J, Peynegre R, Andre J, Brasnu D. Bilan de 158 parotidectomies et problèmes histologiques posés par les tumeurs mixtes des glandes salivaires. *Ann Oto-Laryngol (Paris)* 1979;96:261-80.
- Nussbaum M. Hemifacial Spasm associated with benign Parotid Tumor. *Ann Otol* 1977;86:73-4.
- Martinelli P, Gabellini AS, Lugaresi E. Facial nucleus involvement in post-paralytic hemifacial spasm. *J Neurol Neurosurg Psychiatry* 1983;46:586-7.

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Recurrent intracranial haemorrhage in 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 Nakae,¹ 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 38 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 48 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 this present 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 2.9 mEq/l; Chlor 107 mEq/l. The protein value was 7.7 g/dl. The ECG was within

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normal limits. The EEG showed an irregular and diffuse alpha rhythm (9–11 Hz) mixture with sporadic theta activity mainly in the left frontal region. A marked hypertension had persisted in spite of therapy. On the 7th day of admission, the patient suddenly became unresponsive to verbal commands (semicoma) presenting a decorticate posture of his right extremities. A massive left basal ganglionic haematoma was demonstrated on CT. Thereafter the patient was in semicoma and there was no obvious recovery. On 53rd day of admission, the patient developed a generalised convulsive seizure. Immediately after the seizure he displayed a bilateral decerebrate posture and he became completely unresponsive to any painful stimuli. Then CT scan revealed a massive haematoma in the right basal ganglion region. There was no remarkable recovery in his consciousness after the third attack of cerebral haemorrhage. The patient developed complications of broncopneumonia and severe gastrointestinal bleeding. The patient died on 65th day of admission.

Postmortem examination showed the brain weight to be 1390 g. Old subarachnoid haemorrhage was seen at the base. There was a large old haematoma in the left basal ganglia. In the left basal ganglia, there was a relatively fresh large haematoma penetrating into the left lateral ventricle. An old large haematoma was found in the left cerebellar hemisphere. No arteriovenous malformation or visible aneurysm was found in these haematomas. There was a small secondary haemorrhage and small localised infarction in the pons. The gastrointestinal tract showed severe ulceration and erosion accompanied by haemorrhage. The right and left lungs weighed 770 and 960 g, respectively. Both lungs were oedematous with severe bronchopneumonia. There were many microemboli in the pulmonary arteries. Arteriosclerotic changes were marked also in both kidneys which showed benign nephrosclerosis accompanying with interstitial nephritis. Microscopic investigation revealed a slight lymphocytic infiltration around the small vessels in the cerebral white matter which resembled a characteristic neuropathological feature of neuro-Behcet disease. A perivascular lymphocytic cell cuffing was observed in the post-capillary venule located deep in the cerebral hemisphere. Small softening foci were scattered in the cerebral white matter and in the pons. In addition to these findings, there were marked hypertensive arterial changes in the intracerebral small

arteries: thickening of vessel wall, hyalinosis, and fibrinoid degeneration. Hypertensive arterial changes were marked in the small arteries in the basal ganglia.

The central nervous system is affected in 10–25% of the patients with Behcet disease.^{2,3} Meningoencephalitis and diffuse brainstem lesions are known to be typical forms of neuro-Behcet disease. Their characteristic neuropathological features include perivascular cell cuffing and disseminated focal softenings.³ At this admission, there were scattered small softenings deep in the cerebral hemisphere but the perivascular lymphocytic infiltration was mild and it was not in the active stage so that it could cause organic changes of the vessel wall. Five years after the diagnosis of Behcet disease, the patient experienced a mild hemiparesis and dysarthria. There is a possibility that these neurological deficits might be associated with a small infarction (lacunar stroke syndrome) of the basal ganglia, because the patient had a history of hypertension and there was a spontaneous gradual recovery of these neurological deficits. There was no arteriovenous malformation or aneurysm which could cause a massive intracranial haemorrhage. In addition, the pathological features of these intracranial haematomas resemble so-called spontaneous (hypertensive) intracerebral haemorrhage which can be seen in the hypertensive patients. Furthermore, there were marked hypertensive arterial changes including hyalinosis and fibrinoid degeneration in the intracerebral small arteries in this patient. These pathological findings indicate that this recurrent intracranial haemorrhage might be associated with hypertension. The intracranial small penetrating arteries are known to be the common sources of bleeding in hypertensive intracerebral haemorrhage.⁴ In the histopathological examinations, it has been suggested that a hyalin change in the media of small arteries and a focal fibrinoid degeneration are closely related with the occurrence of the hypertensive intracerebral haemorrhage.^{5,6} Fibrinoid necrosis is now considered to be the responsible factor for the weakness of the arterial wall in hypertensive haemorrhage.⁷ Consequently, these marked hypertensive arterial changes of the intracerebral small arteries might be responsible for the recurrent intracranial haemorrhages in this patient. On the other hand, the intracranial blood vessels have usually been normal in the previous reports on patients with Behcet disease, except for several

instances: a small perivascular haemorrhage;⁸ an occlusion of the middle cerebral artery;⁹ bilateral sigmoid sinus thrombosis accompanied with arteriovenous malformation;¹⁰ a thrombosis¹¹ or thrombophlebitis¹² of the sagittal sinus. There is no literature describing intracranial haemorrhage in patients with Behcet disease. However, Aksel¹³ reported a hyaline degeneration in the media of the intracerebral arteries in a 34-year-old patient with Behcet disease who displayed hemiparesis, dysarthria and meningeal signs. This may suggest some participation of Behcet disease in the progression of the vascular lesions of cerebrovascular disease in such patients. There are several reports of patients with systemic vasculitis who developed intracranial cerebral haemorrhage. In these patients,^{14–17} cerebral haemorrhage was not directly due to the vasculitis itself. The authors concluded that the pathogenesis of cerebral haemorrhage is associated with the side effects of anticoagulant therapy in the treatment of thrombophlebitis. Since there is very little pathological information concerning patients with Behcet disease who developed cerebrovascular disease, it seems difficult to differentiate the pathogenesis of cerebral haemorrhage in Behcet disease. As far as our patient is concerned, however, the histopathological features and the clinical history indicate that the recurrent intracranial haemorrhage is more closely associated with the hypertensive arterial changes than with the perivascular lesions of Behcet disease.

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References

- Maeda K, Nakae K. Epidemiological study on dead cases of Behcet's disease in Japan. In: Dilsen N, Konice M, Ovul C, eds. *Behcet's Disease: Proceedings of an International Symposium on Behcet's Disease*. Amsterdam-Oxford: Excerpta Medica, 1979:47–52.
- Schotland DL, Wolf SM, White HH, Dubin HV. Neurologic aspects of Behcet's disease: case report and review of the literature. *Am J Med* 1963;34:544–53.

Matters arising

The relation of essential tremor to Parkinson's disease.

Sir: We note with interest the absence of a genetic linkage between essential tremor and Parkinson's disease reported by Marttila *et al* from their epidemiological studies in a Finnish population.¹

Unfortunately the precise incidence and prevalence of essential tremor in the United Kingdom is not known as no properly constructed epidemiological studies have yet been carried out, though indirect evidence suggests that, as in the Finnish survey² essential tremor is a common disorder. In agreement with Marttila *et al* our uncontrolled observations on more than 400 patients with essential tremor seen over the last five years have not suggested a greater incidence of Parkinson's disease in either the patients themselves or their relatives than one would expect to find in the general population. However, until the prevalence of essential tremor within the UK population is established by much needed community-based epidemiological studies, the possibility of a genetic link between essential tremor and Parkinson's disease cannot be discounted entirely.

The occurrence of "an additional tremor of the essential tremor type" in patients with Parkinson's disease needs clarification. For instance Shahani and Young³ describe the presence of an action tremor of higher frequency than the typical rest tremor in 5–10% of patients with Parkinson's disease. In fact, surveys have shown higher frequency action or postural tremor to be common in Parkinson's disease co-existing with, and separate in origin from, the characteristic lower frequency tremor of rest.^{4,5} In a study of 60 patients with Parkinson's disease, symptomatic postural tremor was as common as the rest tremor and in more than 80% of patients, two corresponding peaks could be seen to co-exist in the tremor spectrum.⁵ This feature we have found pathognomonic of Parkinson's disease.^{5,6} Using techniques of waveform analysis other, separate tremors of different frequencies can also be seen in some patients.⁷ Thus, action and postural tremors are an integral part of the tremulous phenomena seen in Parkinson's disease. These tremors have behavioural characteristics in common with other postural and action tremors, including essential tremor, and may share common underlying pathophysiological mechanisms. However,

Parkinson's disease and essential tremor are distinct nosological entities, separable in terms of structural pathology and prognosis.

To avoid confusion it would be preferable if the term "tremor of essential type" was not used when referring to tremors of Parkinson's disease. The overlap in phenomenology between essential tremor and Parkinson's disease may account for the frequency with which the former is misdiagnosed as the latter.⁸

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References

- Marttila RJ, Rautakorpi I, Rinne UK. The relation of essential tremor to Parkinson's disease. *J Neurol Neurosurg Psychiatry* 1984;47:734–5.
- Sevitt I. A comparison of propranolol and benzhexol in essential tremor. *Practitioner* 1974;213:91.
- Shahani BT, Young RR. Physiological and pharmacological aids in the differential diagnosis of tremor. *J Neurol Neurosurg Psychiatry* 1976;39:772–83.
- Lance JW, Schwab RS, Peterson, EA. Action tremor and the cogwheel phenomenon in Parkinson's disease. *Brain* 1963;86:95–101.
- Findley LJ, Gresty MA, Halmagyi GM. Tremor, the cogwheel phenomenon and clonus in Parkinson's disease. *J Neurol Neurosurg Psychiatry* 1981;44:534–46.
- Gresty MA, Findley LJ. Postural and resting tremors in Parkinson's disease. In: Hassler RG, Christ JF eds, *Advances in Neurology*. Vol 40. New York, Raven Press, 1984;361–4.
- Findley LJ, Gresty MA. Tremor and rhythmic involuntary movements in Parkinson's disease. In: Findley LJ, Capildeo R eds, *Movement Disorders—Tremor*. Macmillan Press, 1984;295–304.
- Dupont E. Parkinson's disease and essential tremor: Differential diagnostic and epidemiological aspects. In: Rinne UK, Klinger M, Stamm G eds, *Parkinson's disease—Current Progress, Problems and Management*. Elsevier/North-Holland Biomedical Press, 1980;165–79.
- Shimizu T. Clinicopathological studies on Behcet's disease. In: Dilsen N, Konice M, Ovul C, eds. *Behcet's Disease: Proceedings of an International Symposium on Behcet's Disease*. Amsterdam-Oxford: Excerpta Medica, 1979:9–43.
- Fisher CM. The pathology and pathogenesis of intracerebral hemorrhage. In: Fields WS, ed. *Pathogenesis and Treatment of Cerebrovascular Disease*. Springfield: Charles C Thomas, 1961:295–317.
- Anders HE, Eicke WJ. Über Verengerungen an Gehirngefassen bei Hypertonie. *Z Ges Neurol Psychiatr* 1939;167:562–75.
- Feign I, Prose P. Fibrinoid arteritis of the brain associated with hypertension and gross cerebral hemorrhage: a form of "hyalinosis". *Trans Am Neurol Assoc* 1958;83:71–4.
- Fisher CM. The pathologic and clinical aspects of thalamic hemorrhage. *Trans Am Neurol Assoc* 1959;84:56–9.
- Scott D. Mucocutaneous-ocular syndrome (Behcet syndrome) with meningoencephalitis: report of a case with autopsy. *Acta Med Scand* 1958;161:397–400.
- Bienenstock H, Margulies ME. Behcet's syndrome: report of a case with extensive neurologic manifestations. *New Eng J Med* 1961;264:1342–5.
- Imaizumi M, Nukada T, Yaneda S, Abe H. Behcet's disease with sinus thrombosis and arteriovenous malformation in brain. *J Neurol* 1980;222:215–8.
- Ueda T, Gondo M, Kitano I, Kinoshita K, Kikuchi I. Vasculo-Behcet's disease with superior sagittal sinus thrombosis—case report. *Brain Nerve* 1982;34:349–54.
- Segasothy M. Behcet's syndrome with sagittal sinus and inferior vena caval thrombophlebitis. *Med J Malaysia* 1982;37:132–3.
- Aksel IS. Au sujet d'une forme confusionnelle-encephalitique de la maladie de Behcet. Etude anatomico-clinique et electro-encephalographique. *Rev Neurol (Paris)* 1958;99:316–22.
- Ford RG, Siekert RG. Central nervous system manifestations of periarteritis nodosa. *Neurology (Minneapolis)* 1965;15:114–22.
- Harvey AM, Shulman LE, Tumulty PA, Conley CL, Schoenrich EH. Systemic lupus erythematosus: review of the literature and clinical analysis of 138 cases. *Medicine* 1954;33:291–437.
- Johnson RT, Richardson EP. The neurologic manifestations of system lupus erythematosus. *Medicine* 1968;47:337–69.
- Edwards KR. Hemorrhagic complications of cerebral arteritis. *Arch Neurol* 1977;34:549–52.

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