showed a somaesthetic transfer defect. A dichotic listening test revealed auditory suppression (left 14/120, right 68/120; $p < 0.0001$). Pure tone audiometry was within normal limits and showed no difference in threshold between right and left ears. Using a tachistoscope, visual stimuli were presented for 70 ms in each visual hemifield, beside a yellow spot 3° in visual angle. The presence of left hemialexia of Japanese kana was identified (left 16/30, right 25/30, $p = 0.025$). Some graphically simple kanji are learned by the end of the first grade of elementary school, and many of them are pictographic (for example, “口” for the mouth). Reading of such Japanese kanji revealed no significant differences between left and right (left 20/30, right 23/30, $p = 0.57$). The following callosal disconnection syndromes were absent: left visual anomia (left 10/10, right 10/10, $p = 1.0$); left agraphia of kanji (left 44/46, right 46/46, $p = 0.49$) and kana (left 43/46, right 46/46, $p = 0.24$); left tactile anomia (left 20/20, right 20/20, $p = 1.0$); and right constructional apraxia, which was examined using the block design test of WAIS-R (Wechsler Adult Intelligence Scale-Revised) (left 8/10, right 9/10, $p = 1.0$).

Using the tachistoscope, we also assessed hemispheric superiority for the naming of musical symbols. Thirty musical symbols (G, F, and C clefs, up and down bow, accent marks, repeat marks, 16th-, 8th-, quarter-, half-, and whole-notes and rest, a couple of eight notes, sharp, double sharp, flat, double flat, natural, $\sharp$, segno, tenuto, flageolet) were randomly ordered and briefly presented in each visual hemifield. Time and dynamic symbols were not included because they were composed of actual letters, the reading of which involves the left hemisphere. The patient showed left hemianomia for musical symbols (three times examined: left 22, 20, 20/30; right 28, 27, 28/30, total left 62/90, right 83/90, $p = 0.0001$). He mistook up bow, accent, 16th-note and rest, 8th-note and half rest, double sharp, natural, segno, and flageolet in the left visual hemifield, and, in the right, double sharp and flageolet. It was possible that the patient could not represent verbal answers even if his right hemisphere was able to understand the meaning of those musical symbols, because of the disconnection between two cerebral hemispheres. In order to clarify this, a way of answering that did not require verbal processing was recommended. We undertook the same hemispheric superiority task for naming of musical symbols described above. Only 13 symbols were chosen this time (up and down bow, 16th-, 8th-, quarter-, half, and whole-notes and rest, sharp, flat), because the other symbols are hard to express in a non-verbal way. Each symbol was presented twice (26 trials in all). The patient was asked to answer by pointing to a symbolic representation of the musical symbol with the hand that was ipsilateral to the represented visual hemifield. These representations consisted of figures which did not contain any words. For example, for the up and down bow, an upward and a downward arrow was used. For the quarter tone or rest, four columns with four divided rooms were shown, each column having one to four painted rooms. On this test, the patient showed no significant differences between the two visual hemifields (left 21/26 and 22/26; right 24/26 and 24/26; total left 43/52, right 48/52; $p = 0.23$).

The results of our examination showed left hemisphere dominance in naming musical

symbols. This result is in agreement with reports showing dominance of the left hemisphere in musicians when dealing with musical stimuli.⁶ Owing to the callosal lesion, the right hemisphere had lost access to the contralateral speech control centres, so reading musical symbols aloud was impaired. It is widely accepted that visual information is delivered through the splenium of corpus callosum. The callosal lesion in our patient affected the anterior half of the splenium of the corpus callosum, suggesting that information contained in musical symbols is mediated there. Our patient also showed left hemialexia of kana, but not of kanji. The kanji used in our experiment convey pictographic meaning, whereas kana are purely symbolic. Musical scores, except for time and dynamic symbols, are also symbolic. It seems reasonable to conclude that similar neuro-pathological mechanisms are at work in the patient's alexia of both kana and musical symbols.

M Satoh, K Furukawa

Department of Neurology, Mie University School of Medicine, Mie 514-8507, Japan

K Takeda

Department of Neurology, Japanese Red Cross Medical Centre, Tokyo 150-8935, Japan

S Kuzuhara

Department of Neurology, Mie University School of Medicine, Mie 514-8507, Japan

Correspondence to: Dr Masayuki Satoh, Department of Neurology, Mie University School of Medicine, Tsu, Mie 514-8507, Japan; masayuki-sato@seijui-hp.jp

doi: 10.1136/jnnp.2005.068692

Competing interests: none declared

References

- 1 **Sadie S**, editor. *Grove's concise dictionary of music*. London: Macmillan, 1994.
- 2 **Brust JCM**. Music and language; musical alexia and agraphia. *Brain* 1980;103:367-92.
- 3 **Kawamura M**, Midorikawa A, Kezuka M. Cerebral localization of the center for reading and writing music. *Neuroreport* 2000;11:3299-303.
- 4 **Grisson B**. Une etude sur les alterations musicales au cours des lesions hemispheriques [These]. Paris, 1972. [Cited by Benton AL. The amusias. In: Critchley M, Henson RA, editors, *Music and the brain*. London: Heinemann, 1977:378-97.]
- 5 **Annett M**. The bimodal distribution of right, mixed and left handedness. *Q J Exp Psychol* 1967;19:327-33.
- 6 **Evers S**, Dannert J, Roedding D, et al. The cerebral haemodynamics of music perception: a transcranial Doppler sonography study. *Brain* 1999;122:75-85.

Leucocytoclastic vasculitis neuropathy diagnosed by biopsy of normal appearing skin

Leucocytoclastic vasculitis (LCV) is a clinicopathological entity that preferentially involves capillaries or small vessels rather than the medium sized or large arteries typical of polyarteritis nodosa. Its histopathological features are characterised by the presence of perivascular polymorphonuclear leucocytes with fragmented nuclear debris (leucocytoclasia).¹ It may be limited to skin lesions such as erythematous macules, purpuric papules, and haemorrhagic vesiculobullous lesions. Although LCV often involves organ systems other than the skin, it is rarely

associated with neurological complications, unlike polyarteritis nodosa or Churg-Strauss syndrome. We report a patient who presented with leucocytoclastic vasculitic neuropathy without skin lesions, and discuss the value of doing a combined biopsy of the skin, nerves, and muscle in this patient for the detection of vasculitis.

A 44 year old man was admitted to our hospital because of progressive asymmetrical paraesthesiae in all four limbs. Eight months before admission, a tingling sensation had begun in the second finger of his left hand, but it disappeared spontaneously within one month. Over the next seven months, he experienced dysaesthesia which spread gradually on the lateral sides of his feet, and later to his left hand, and he developed drop foot on the right. Two weeks before admission, he had dysaesthesia in his right hand and left thigh. There was no weight loss, fever, general malaise, alcoholism, toxin exposure, or nutritional deficiency. He had no history of asthma, only seasonal allergic sinusitis in recent years.

On admission, he was afebrile. A general physical examination was normal. No skin erythema or purpura was present. On neurological examination, he had a pattern of multiple mononeuropathy. No cranial nerve dysfunction was found. There were peroneal nerve palsy on the right and ulnar nerve palsy on both sides. On the MRC scale, muscle strength was graded as 1/5 for the right tibialis anterior, and 4/5 for the bilateral abductor digiti minimi and dorsal interosseous muscles. The strength of the other muscles was normal. Tendon reflexes were normal, and plantar responses were flexor. Sensory examination showed mild impairment of pain and touch sensations in the bilateral ulnar nerve and right peroneal nerve territories.

Laboratory tests showed a white cell count of 6900/ μ l with 3% eosinophils, and 0.0 mg/dl serum C reactive protein. Haemoglobin, urea, and serum creatinine values were all normal. Serological markers for hepatitis B virus, hepatitis C virus, Epstein-Barr virus, cytomegalovirus, and syphilis were negative. Rheumatoid factors, antinuclear antibody, and antineutrophil cytoplasmic antibody were all negative. Cerebrospinal fluid analysis showed a normal cell count (2 lymphocytes/ mm^3) and normal protein (42 mg/dl). Nerve conduction studies showed asymmetrically reduced amplitudes of compound muscle action potentials and sensory nerve action potentials, particularly in the clinically affected nerves, indicative of multiple mononeuropathy and axonal degeneration.

Combined biopsies were performed on the left sural nerve, peroneus brevis muscle, and skin at the ankle. The skin was taken as a full thickness biopsy from the incision site. Three or more sections were examined for each specimen. Sural nerve biopsy findings showed reduced myelinated fibre density (3587/ mm^2), but no significant vessel changes, inflammatory cells, and findings suggestive of demyelination. The skin biopsy showed vasculitis, mainly involving capillaries in the dermis, with leucocyte infiltration and fragmented nuclear debris (fig 1), indicative of LCV. Findings in the peroneus brevis muscle biopsy were normal.

Intravenous methylprednisolone (1 g/day for three consecutive days) was given in two series, followed by oral methylprednisolone (80 mg on alternate days). After the start of corticosteroid treatment, the symptoms gradually lessened. There was good recovery of movement of the right foot two months later

Figure 1 Microscopy findings for the leg skin biopsy. Vasculitis involving capillaries or postcapillary venules in the dermis is associated with leucocyte infiltration and fragmented nuclear debris (haematoxylin-eosin staining; bar = 20 μ m).

(4/5 on the MRC scale), but leg paraesthesiae and muscle weakness in the ulnar nerve territory persisted.

Comment

This patient's illness was characterised by steroid responsive multiple mononeuropathy presumably caused by LCV without skin manifestations. Connective tissue disease, drugs, malignancies, and viral infections have been reported as possible causative agents of LCV, but our patient had no clinical or serological evidence of a collagen disease or any other condition. In a study of 82 patients with LCV, peripheral neuropathy was confirmed by nerve conduction studies in only four; no nerve biopsies were done.¹ There have been few reports of LCV neuropathy confirmed by pathological examination of nerves. We previously reported a patient with LCV neuropathy associated with Epstein-Barr virus infection who had generalised skin eruptions, in whom the skin biopsy showed leukocytoclastic vasculitis.² LCV is often associated with skin lesions, but the present case is unique in that the patient did not have obvious skin manifestations, but had asymptomatic cutaneous vasculitis. In a study of hepatitis C virus related neuropathy, three of 30 patients had LCV in the muscles or nerves. One of the three did not have skin eruptions, and whether the other two did was not made clear.³ A combined biopsy, rather than a simple nerve or muscle biopsy, may be important for obtaining diagnostic evidence of LCV neuropathy.

Combined muscle and nerve biopsy for patients with vasculitis is reported to be a successful method of diagnosis. Said *et al*⁴ used superficial peroneal nerve/peroneus brevis muscle biopsies on a large series of patients with vasculitic neuropathy and reported that the muscle biopsy was diagnostic of arteritis in 80% and the nerve biopsy

in 55% of the patients. Collins *et al*⁵ reported that 90% of nerve and 50% of muscle biopsies in their study group showed pathologically definite vasculitis. Combined muscle and nerve biopsy appears to have high sensitivity, but this was not true for our patient, who had no evidence of vasculitis in his muscle and nerve specimens. Vasculitis could not have been diagnosed only by sural nerve/peroneus brevis muscle biopsy, although our patient developed LCV without skin lesions. A biopsy of normal appearing skin at the nerve biopsy site in our patient proved crucial in the diagnosis of vasculitis. We suggest that LCV neuropathy may occur more commonly than expected and that the use of combined biopsy of skin, nerve, and muscle increases the likelihood of detecting vasculitis, even when patients have no obvious skin lesions.

Y Yamanaka, A Hiraga, K Arai, M Mori,
K Kanai, T Hattori, S Kuwabara

Department of Neurology, Chiba University School of Medicine, Chiba, Japan

T Oide, T Tanizawa

Department of Pathology, Chiba University Hospital

Correspondence to: Dr Satoshi Kuwabara,
Department of Neurology (D3), Chiba University
Graduate School of Medicine, 1-8-1 Inohana, Chuo-
ku, Chiba 260-8670, Japan;
kuwabara-s@faculty.chiba-u.jp

doi: 10.1136/jnnp.2005.077321

Competing interests: none declared

References

- 1 Ekenstam EA, Callen JP. Cutaneous leukocytoclastic vasculitis. *Arch Dermatol* 1984;**120**:484-9.
- 2 Kanai K, Kuwabara S, Mori M, *et al*. Leukocytoclastic-vasculitic neuropathy associated with chronic Epstein-Barr virus infection. *Muscle Nerve* 2003;**27**:113-16.
- 3 Authier FJ, Bassez G, Payan C, *et al*. Detection of genomic viral RNA in nerve and muscle of patients with HCV neuropathy. *Neurology* 2003;**60**:808-12.
- 4 Said G, Lacroix-Claudio C, Fujimura H, *et al*. The peripheral neuropathy of necrotizing arteritis: a clinicopathological study. *Ann Neurol* 1988;**23**:461-5.
- 5 Collins MP, Mendell JR, Periquet MI, *et al*. Superficial peroneal nerve/peroneus brevis muscle biopsy in vasculitic neuropathy. *Neurology* 2000;**55**:636-43.

Pulmonary arteriovenous fistulae thrombosis responsible for recurrent stroke

Pathophysiological mechanisms of ischaemic strokes occurring in patients with Rendu-Osler-Weber (ROW) disease remain unclear. We report on the case of a patient with ROW disease and pulmonary arteriovenous fistula thrombosis who suffered from recurrent strokes.

Case report

In January 2005, a 38 year old woman was referred for the first time to our hospital for an ischaemic stroke fifteen days after undergoing a medically induced abortion. She reported recurrent epistaxis during her two pregnancies (1993 and 1994) and two familial cases of Rendu-Osler-Weber (ROW) disease. She also had experienced two episodes of ischaemic stroke (January 1996 and December 1997). Stroke workup had only revealed a patent foramen ovale with atrial septal aneurysm, which had been surgically closed after the second stroke. She had been receiving aspirin since closure of the foramen ovale.

Eight years after the procedure, she presented with a new ischaemic stroke in the right middle cerebral artery territory, confirmed by brain diffusion weighted sequence magnetic resonance imaging. Physical examination was otherwise unremarkable. Cervical and transcranial ultrasonography, 12-lead ECG, 24-hour ECG monitoring, and chest radiography were normal. Routine blood tests and thrombophilia study (including tests for protein C, protein S, antithrombin III, factor V Leiden, factor II, hyperhomocysteinaemia, and antiphospholipid antibodies) did not show any abnormality. Contrast transoesophageal echocardiography (C-TOE) revealed no atrial residual shunt, but a massive appearance of contrast bubbles in the left atrium from pulmonary veins. Chest 16-slice multidetector computed tomographic (CT) angiography (Sensation 16, Siemens) with injection of 90 ml of contrast medium (Xenetix 350) at a rate of 4 ml/s using the "bolus tracking" technique was carried out. Helical CT data were acquired with a pitch of 0.75, 160 mAs, and 120 kV, and images were reconstructed with a slice thickness of 1 mm and interval of 0.5 mm. Multi-planar reconstruction and maximum intensity projection images showed multiple pulmonary arteriovenous fistulae (PAVFs), consistent with ROW disease: two PAVFs in the right lower and apical lobes and two in the lingual and the left lower lobe. A thrombus was found in the PAVF located in the paracardiac segment of the right lower lobe (fig 1). Lower limbs and inferior vena cava ultrasonography did not reveal any venous thrombosis. Warfarin was started, and a new chest CT done one month later showed complete resolution of the intra-fistula thrombus. Catheter embolisation of all PAVFs was then successfully undertaken.