detection, mainly in the digestive tract⁴ but sometimes in lung adenocarcinoma.⁵

The present patient high CSF CA 19-9 concentration, in whom leptomeningeal carcinomatosis probably originated from lung carcinoma, raised two new issues. Firstly, this is the first report in which CSF concentrations of CA 19-9 but not those of CEA proved useful as a marker in leptomeningeal carcinomatosis. The case showed a high CSF/serum CA 19-9 ratio of 1.56 (61.4 U/ml/39.4 U/ml), indicating that the marker was produced and released by meningeal tumour cells. In addition to the evidence from calculations demonstrating local production of CA 19-9, tumour cells in CSF were shown to possess CA 19-9-producing ability by cytoplasmic immunostaining. Therefore, serial determinations of CA 19-9 concentration in CSF may be helpful in the diagnosis and clinical management of leptomeningeal carcinomatosis even when the CEA concentration in CSF is not increased.

The second point of interest is that the CEA concentration was not increased throughout the clinical course despite increases in serum, and despite CEA immunoreactivity in the cytoplasm of tumour cells in the CSF. This discrepancy indicates that CEA produced by tumour cells in the meninges was not effectively released into CSF, for unknown reasons. However, determination of both CEA and CA 19-9 concentrations in CSF may increase sensitivity and effectiveness of tumour markers in the diagnosis and monitoring of leptomeningeal carcinomatosis, as one may be raised in isolation.

YOSHIHIRO SATO

Department of Neurology, Futase Social Insurance Hospital, Iizuka 820, Japan

YOSHITAKA OHTA

MASAHIDE KAJI

Department of Pathology, St Mary's Hospital, Kurume 830, Japan

KOTARO OIZUMI

MASAHIDE KAJI

Department of Internal Medicine, Kurume University School of Medicine, Kurume 830, Japan

Correspondence to: Dr Yoshihiro Sato, Department of Neurology, Kurume University Medical Center, 155-1 Kokubumachi, Kurume 839-0863, Japan. Telephone 0081 942 22 6111; fax 0081 942 22 6533; email y-sato@ktarn.or.jp

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Hereditary motor and sensory neuropathy type 1A associated with sensorineural deafness

The most common type of hereditary motor and sensory neuropathy type 1, HMSN 1A, is caused by a duplication of the gene for peripheral myelin protein 22 (PMP 22), situ-

ated on chromosome 17p. We report on a patient with this genotype with bilateral sensorineural deafness.

A 28 year old man presented with progressive distal weakness, numbness, and progressive bilateral hearing loss. He had first noticed problems with running in his early teens and at the age of 13 had two operations to correct bilateral pes cavus. His walking tolerance gradually deteriorated to half a mile unaided. At the age of 24, he had three operations to correct thoracic scoliosis and subsequently noticed progressive weakness of his hands. Since the age of 26 he had been aware of diminished sensation in his feet and progressive bilateral hearing loss. Medical history was unremarkable and he had not been exposed to any relevant drugs or toxins. There was no history of neurological problems among three siblings, his three children or the rest of his family. There was no parental consanguinity.

General medical examination disclosed bilateral pes cavus, palpable greater auricular nerves, and evidence of previous spinal surgery. Otoscopy was normal. Audiological examination showed bilateral sensorineural deafness (-40 dB at 6 kHz in both ears). Visual acuity, fundoscopy, and all other cranial nerves were normal. There was a typical essential tremor of both hands. In the limbs there was symmetric distal wasting, worse in the lower limbs, with corresponding weakness. Deep tendon reflexes were absent. Pinprick and vibration sensation were impaired in both feet.

Laboratory investigations showed no evidence of any underlying systemic disorder. Protein concentration in CSF was 1.55 g/l. Nerve conduction studies showed markedly diminished motor conduction velocities: right median nerve motor velocity was 16 m/s; right posterior tibial nerve motor velocity was 14m/s; sensory responses were unobtainable in the right sural and superficial peroneal nerves. Right sural nerve biopsy showed a severe demyelinating neuropathy with massive onion bulb formation. Genetic evaluation of a serum sample confirmed the presence of the PMP 22 duplication on chromosome 17. Brain stem auditory evoked responses (BAERs) were performed (table). Wave I latencies were significantly delayed compared with laboratory controls. Interwave I-V latencies were only marginally increased.

A recent paper on a series of 61 patients with the PMP 22 duplication reported no cases of deafness.¹ However, deafness and asymptomatic abnormalities of BAERs has been reported in association with HMSN. In one kindred, 61 patients had a history of hearing loss in association with an autosomal dominant HMSN type 1.² There were seven cases of sensorineural deafness among the 14 family members described by Cruse *et al.*,³ who had an autosomal dominant HMSN type 1. The four patients with symptomatic deafness described by Raglan *et al.*⁴ also con-

form to recognised criteria for a diagnosis of HMSN type 1 but more detailed clinical information on these patients is lacking. Perez *et al.*⁵ suggested, in a small community study, that the incidence of deafness in a mixed group of HMSN1 and HMSN 2 could be as high as 30%. Only the 77 year old female patient of Nicholson *et al.*⁶ is definitely HMSN 1A, and it could be speculated whether at this age deafness is a coincidental symptom.

Nicholson *et al.*⁶ have recently renewed interest in auditory abnormalities in patients with HMSN 1A. Our patient conforms to the pattern they found, with isolated delayed wave I latencies suggesting a problem in the distal VIIIth nerve, in which PMP 22 expression is known to occur. This is the first case report to provide incontrovertible evidence, with nerve biopsy and genetic analysis, of bilateral sensorineural deafness in a young patient with "a full house" phenotypically and genotypically for HMSN 1A.

Neurologists and otolaryngologists should be aware of this association, which may be commoner than previously thought.

J STONE

G MORAN

T J WALLS

Regional Neurosciences Centre, Newcastle General Hospital, Westgate Rd, Newcastle, UK

Correspondence to: Dr TJ Walls, Regional Neurosciences Centre, Newcastle General Hospital, Westgate Rd, Newcastle NE4 6BE, UK. Telephone 0044 191 273 8811 ext 22460; fax 0044 191 272 4823.

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Chronic inflammatory demyelinating polyneuropathy accompanied by carcinoma

We read the article by Antonie *et al* with interest.¹ We have also reported on a patient with chronic inflammatory demyelinating polyneuropathy (CIDP) accompanied by hepatocellular carcinoma who showed improvement after intravenous methylprednisolone injection.² On the basis of this experience, we investigated 20 consecutive patients with pathologically established hepatocellular carcinoma in our hospital and found another patient having definite CIDP using the criteria of the Ad Hoc subcommittee of the American Academy of Neurology.³ This patient also had motor sensory neuropathy and showed improvement after intravenous methylprednisolone injection. Our two patients showed a motor sensory neuropathy affecting the four limbs. Their CSF contained high concentrations of protein and the study of conduction velocities and biopsied nerves showed demyelination.

Brain stem auditory evoked potentials in this patient show marked delay of wave I latencies with preservation of interwave I-V latencies compared with laboratory controls

Latency (ms)	Normal range Mean (SD)	HMSN 1A (n=1)	
		Left	Right
Wave I	1.7 (0.15)	2.22	2.37
Wave IV	5.1 (0.24)	6.30	6.33
Wave V	5.7 (0.25)	6.60	6.68
Interwave I-V	4.0 (0.23)	4.38	4.31

We did not find anti-GM1 antibodies in either patient. As discussed in Antonie *et al*¹ the association of carcinoma and primary demyelinating neuropathy has seldom been reported. However, the cases reported by Antonie *et al* as well as us suggest the possibility that weakness or clumsiness, which is sometimes seen in patients with carcinoma may be caused by CIDP, and these symptoms should not be misinterpreted as representing general weakness caused by malignancies. Further investigation with large numbers of patients may be useful in determining the mechanism.

KAZUO ABE
FUMINOBU SUGAI

Department of Neurology, Osaka University Medical
School, 2-2 Yamadaoka, Suita, Osaka 565-0871,
Japan

Correspondence to: Dr Kazuo Abe, Department of
Neurology, Osaka University Medical School, 2-2
Yamadaoka, Suita, Osaka 565-0871, Japan. Tel-
ephone 0081 6 879 3579; fax 0081 6 879 3579;
email abe@neuro.med.osaka-u.ac.jp

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Frequency, causes, and consequences of burns in patients with epilepsy

The increased incidence of burns in people with epilepsy has long been recognised. Previous surveys (via questionnaires in clinic or burns unit admissions) have identified cooking, showering, and heaters as the most common causes. The duration of epilepsy and frequency of seizures have been recognised as the greatest risk factors,¹⁻⁴ compounded by lack of awareness and education

among people with epilepsy about the risk of burns.³

The aim of this study was to determine the frequency, causes, and consequences of burns and scalds in patients with epilepsy. The population comprised patients with chronic epilepsy who were resident at the Chalfont Centre for Epilepsy (CCE). The residential part of the CCE consists of a series of houses that provide for varying degrees of independence in terms of self care. There is also a short stay tertiary referral inpatient assessment facility and a medical and nursing unit on site where any injury is documented and assessed.

The daily records of the Medical and Nursing Unit for the year June 1995-6 were examined and any record of a burn extracted and followed until it was recorded as healed. The records for the day of the burn were then examined to determine the cause of the burn and whether it was seizure related.

The residential houses were divided into three groups according to the level of self care of the patients within each. Dependency was largely governed by physical infirmity rather than by severity of the seizure disorder.

- Dependent (98 residents)—all meals and hot drinks provided and help given with personal care.
- Intermediate (94 residents)—all meals provided, some residents make their own hot drinks and there is a variable level of independence with personal care.
- Independent (111 residents)—independent with regard to hot drinks, personal care, and some meals.

The number of residents in each category were calculated at the end of the year and remained relatively constant during the 1 year period. Seizures occurring in all patients were recorded prospectively in seizure diaries.

The number of seizures in the same 1 year period (June 1995 - June 1996) were calculated from the case records of the 303 residents. The number of seizures in the 28 bed assessment unit could not be calculated for the year, as there was a high patient

turnover with a median stay of 32 days. The number of seizures from the residential houses were therefore extrapolated to allow for the number of patients in the assessment unit. No burns occurred in the assessment unit, which was classified as dependent with regard to the level of care made available to the inpatients.

A χ^2 test was applied to the number of burns occurring in houses of differing levels of dependence to examine the significance of observed variation. This was then repeated correcting for the median number of seizures per person, in the three groups.

The results are shown in the table. Whereas the frequency of burns is regarded as accurate, the number of seizures may be underreported as nocturnal or minor seizures may not have been seen and recorded. Information as to whether an injury was seizure related or not was unavailable in three, and the cause of the burn was unavailable in two cases. Of the severe burns, one required skin grafting with healing occurring over 3 months and one patient attended an accident and emergency department, but was not admitted, and the burn took 4 months to heal. Two of the severe burns were complicated by methicillin resistant *Staphylococcus aureus* (MRSA).

Infection, but healed uneventfully. Two other patients had burns that were also complicated by an infection but without growth of a particular organism. One received antibiotics and recovered quickly, the other was initially treated only with topical therapy and took longer to recover.

Most burns were related to seizures (19 of 34). This was particularly true of the more severe burns (five of six). A total of 18 631 seizures were recorded during the year meaning that about one seizure in every 980 resulted in a burn and one in every 3105 seizures resulted in a severe burn.

Most burns were caused by hot water injuries (25 of 34). More severe burns were mostly caused by larger quantities of water. Cookers, hot pipes, and heaters were rare causes of burns in this study.

There were significantly more burns in the more independent houses ($p < 0.05$). The median number of seizures per person in the more independent houses was also greater as residents were placed in houses according to their physical disability rather than on the severity of their seizure disorder. Correcting for seizure frequency abolished the significance of the effect of level of dependence on the number of burns ($p > 0.1$).

Burns are a serious but underrecognised complication of epilepsy. Previous studies have used two different approaches. The first has been to give patients attending an epilepsy clinic a questionnaire about any burns ever sustained.^{3,4} This method relies on the memory of the patients involved and therefore biases recording towards serious injuries. These studies also gave little idea of frequency, as there was no time limit imposed. The second approach has been to consider patients with epilepsy admitted to burns units.¹ This again restricts the survey to severe burns. Neither method gives any idea of frequency as there is no set population with no calculation of the number of seizures over a period of time, hence no idea of level of risk.

The residents at the Chalfont Centre for Epilepsy live as normal a life as their level of disability allows. This means that some residents are not exposed to the same risk of burns as those living in the community. A

Factors influencing burns in patients with epilepsy

	Severity of burns (time to heal (days))				Total
	0-7	7-14	14-21	>21	
Burns (n)	17	8	3	6	34
Relation to seizure:					
Seizure	8	3	3	5	19
No seizure	8	4	0	0	12
Not known	1	1	0	1	3
Cause of burn:					
Kettle	2	1	1	2	6
Teapot	1	1	0	2	4
Cup	10	3	1	1	15
Cooker	2	0	0	0	2
Chemical	0	1	0	0	1
Cigarettes	1	1	0	0	2
Hot pipe	0	0	1	0	1
Heater	1	0	0	0	1
Unknown	0	1	0	1	1
Relation to independence and seizure frequency:					
Burns/100 people (n):					
Independent	9	1.8	1.8	3.6	16.2
Intermediate	5.3	4.3	0	2.1	11.7
Dependent	2	2	1	0	5
Burns/seizure ($\times 10^{-4}$) (n):					
Independent	21	4.2	4.2	8.4	37.8
Intermediate	18	15	0	7.5	40.5
Dependent	14	14	7	0	35
Seizures/person/year (median):					
Independent	43				
Intermediate	28				
Dependent	14				