Brain (1988), **111**, 1025–1037

FAMILIAL MYOPATHY WITH CHANGES RESEMBLING INCLUSION BODY MYOSITIS AND PERIVENTRICULAR LEUCOENCEPHALOPATHY

A NEW SYNDROME

by ANDREW J. COLE,[1] RUBEN KUZNIECKY, GEORGE KARPATI, STIRLING CARPENTER, EVA ANDERMANN *and* FREDERICK ANDERMANN

(*From the Montreal Neurological Hospital and Institute, Montreal, Canada*)

SUMMARY

Five of 6 male siblings were affected by a progressive myopathy beginning in early childhood. Muscle biopsies in all patients showed the characteristic changes of inclusion body myositis. Computerized tomography and magnetic resonance imaging revealed a markedly abnormal appearance of cerebral white matter in the 4 affected patients tested, but clinical and other laboratory examinations failed to demonstrate evidence of central white matter dysfunction. Muscle biopsies and brain imaging were normal in all clinically unaffected family members. On the basis of the genetics, muscle biopsy findings and cerebral white matter changes, we conclude that this constellation represents a hitherto undescribed syndrome.

INTRODUCTION

Inclusion body myositis (IBM) is a well recognized variety of idiopathic inflammatory myopathy characterized by slowly progressive weakness, usually starting in the sixth or seventh decade, and predominantly affecting men. The diagnosis can be suspected on clinical grounds, but can be confirmed only by the finding of typical inclusions in muscle fibres consisting of 15 to 18 nm filaments (Carpenter *et al.*, 1978). A number of reports have described the pathological findings of IBM in younger patients (Sato *et al.*, 1969; Yunis and Samaha, 1971; Tomé *et al.*, 1981; Eisen *et al.*, 1983) and several examples of apparent familial IBM have been reported (Fukuhara *et al.*, 1982; Matsubara and Tanabe, 1982; Eisen *et al.*, 1983).

We investigated a family of 6 male siblings, 5 of whom were affected by a progressive myopathy. In each patient, muscle biopsy demonstrated the character-

Correspondence to: Dr G. Karpati, Montreal Neurological Hospital and Institute, 3801 University Street, Montreal, Quebec H3A 2B4, Canada.

[1] Present address: Johns Hopkins University School of Medicine, Department of Neuroscience, 725 North Wolfe Street, Baltimore, Maryland, 21205, USA.

istic findings of IBM by light and electron microscopy. Each of the affected patients also showed a striking but asymptomatic abnormality of cerebral white matter on magnetic resonance imaging (MRI).

CASE REPORTS

The familial incidence of this disease is illustrated in fig. 1.

Case II-2

A 35-yr-old man, the first of twins, was born 1 wk before term. He walked at 18 months and was never able to run. Gowers' sign was present from the time he began to stand. Muscle weakness, mainly proximal, progressed slowly during adolescence. Following a motor vehicle accident in which he sustained a femoral fracture at the age of 26 yrs, he required canes to walk.

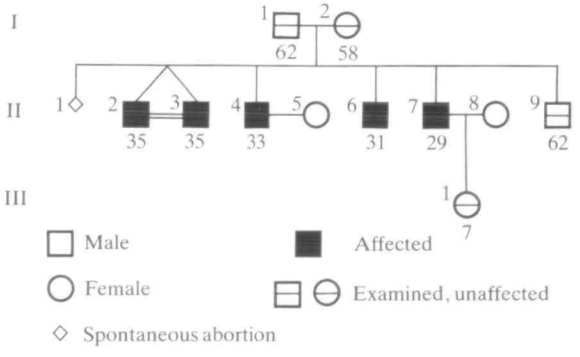

FIG. 1. Pedigree of Family D. Proband is II-6.

At 35 yrs he showed an exaggerated lumbar lordosis and bilateral genu recurvatum. He had moderate proximal weakness in all 4 limbs with minimal wasting. His calves were slightly enlarged but had normal texture. Distal muscle strength was normal. Tendon jerks were absent in the arms, reduced at the knees and preserved at the ankles. The plantar responses were flexor. He required crutches to walk, and had a waddling gait. Mentation, cranial nerves, sensation and coordination were intact.

Case II-3

This man was the second-born identical twin of Case II-2. His motor milestones were delayed in spite of normal intellectual development. Muscle weakness, apparent from early childhood, was slowly progressive, and he currently requires one cane to walk.

At 35 yrs he showed mild proximal muscle weakness in the upper limbs, with moderate proximal weakness in the legs. Distal muscles were of normal strength. Calf bulk was slightly increased. Tendon jerks were absent except at the ankles. He had a waddling gait with increased lumbar lordosis. Sensation and coordination were normal.

Case II-4

A 33-yr-old man had been delivered at term following a normal gestation. Motor milestones were delayed: he walked unassisted at 20 months. Intellectual function was normal. He was never able to run. Muscle weakness progressed slowly throughout his life so that by the age of 30 yrs he had difficulty ascending stairs.

At 33 yrs he showed moderate weakness of proximal and distal muscles of the lower limbs but relative sparing of the intrinsic muscles of the feet. Mild shoulder girdle and proximal arm weakness was present with normal hand strength. Craniobulbar musculature was normal. Tendon jerks were absent except at the ankles. Sensation and mentation were normal. Gait was moderately impaired.

Case 11-6

The proband, a 31-yr-old man, first walked unassisted at 18 months of age. Intellectual development was normal. He had difficulty in school athletics, and muscle weakness continued to progress until the present time. In addition he has suffered from complex partial seizures and occasional generalized convulsions since the age of 7 yrs.

Examination at 31 yrs of age showed moderate kyphoscoliosis. Mentation was normal. A right exotropia was present as were bilateral cataracts and retinal scars considered by a neuro-ophthalmologist to be traumatic in origin. Muscle bulk was diminished in association with diffuse weakness which was worse in proximal muscles of all 4 limbs. Ankle jerks were preserved. Gowers' sign was present and his gait was mildly impaired. Sensation was normal.

Case 11-7

A 29-yr-old man was the product of a normal gestation and delivery. Intellectual development was normal, but motor milestones were markedly delayed. He was able to sit unassisted at the age of 13 months, and to walk at the age of 24 months. He was unable to participate in school athletics because of muscle weakness. There was steady progression of muscle weakness so that by the age of 28 yrs he had great difficulty going up and down stairs. There were no cramps, muscle pains, or fasciculations.

At 29 yrs of age he showed mild kyphoscoliosis and exaggerated lumbar lordosis (fig. 2). The gait was waddling. Gowers' sign was present. Muscle bulk was diminished throughout, especially in the upper limbs. There was moderate diffuse weakness which was most apparent in the proximal muscles of the upper and lower limbs. Neck flexors were also weak, and there was winging of the scapulae; intrinsic

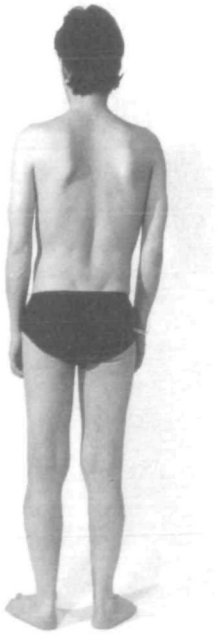

FIG. 2. *Case 11-7.* Note patchy wasting of arm and leg musculature, including muscles of the shoulder girdle.

hand and foot muscles were well preserved. Tendon reflexes were absent except ankle jerks. The plantar responses were flexor. The mental state, cranial nerves, cerebellar and sensory examinations were normal.

RESULTS

Results of laboratory investigations of patients are presented in Table 1. All tests in unaffected family members were normal or negative.

TABLE 1. SUMMARY OF LABORATORY EXAMINATIONS IN FAMILY D*

	Case				
Test	*II-2*	*II-3*	*II-4*	*II-6*	*II-7*
CK(IU/dl) (normal range 5-140)	404	391	668	848	737
EMG	Small polyphasic motor units, fibrillation, positive sharp waves, bizarre high frequency discharges, hyperrecruitment				
NCV	Minimal slowing of motor nerve conduction velocities with slight prolongation of distal F wave latencies				
CSF					
Protein (g/l)	0·61	0·68	NA	0·56	NA
Oligoclonal IgG bands	Neg	Neg	NA	Neg	NA
VLCFA	N	N	N	NA	NA
VEP	N	N	N	N	N
BAEP	N	N	N	N	N

* All tests in unaffected family members were normal. CK = serum creatine kinase. EMG = electromyography. NCV = nerve conduction velocity. NA = not available. Neg = negative. N = normal. VEP = visual evoked potential. BAEP = brainstem auditory evoked potential. VLCFA = serum very long chain fatty acids.

Muscle biopsy

Biopsies of biceps brachii muscle from all family members except Case III-1 were processed for histochemical and electron microscopic examination in the routine manner (Carpenter and Karpati, 1984). Each of the affected patients demonstrated the characteristic findings of IBM on histochemical, phase and electron microscopic examination of muscle biopsies (Carpenter *et al.*, 1978) (Table 2). Histochemical study of cryostat sections revealed many abnormally small calibre fibres mixed with some abnormally large fibres. Centrally situated myonuclei were increased in number. There was a moderate increase in endomysial connective tissue. Rimmed vacuoles were present in many muscle fibres throughout the specimens (fig. 3). Some rimmed vacuoles contained impressive eosinophylic inclusions (fig. 4A). In a few

TABLE 2. SUMMARY OF PATHOLOGICAL FINDINGS IN MUSCLE IN FAMILY D

	Case				
	II-2	*II-3*	*II-4*	*II-6*	*II-7*
Age (yrs)	34	34	32	29	28
Muscle fibre loss	+ + +	+ + +	+ + +	+	+
Necrosis	+ +	+	+	0	+
Rimmed vacuoles	+ + +	+ +	+	+ + +	+ + +
Variability of fibre calibre	+ + +	+ + +	+ + +	+ + +	+ + +
Abnormal 15-18 nm cytoplasmic filamentous inclusions observed	Y	Y	Y	Y	N

0 = absent. + = mild. + + = moderate. + + + = severe. Y = yes. N = no.

fibres, the chromatin of some nuclei was displaced to the periphery of the nucleus by masses of inclusion material (fig. 4B). In the most severely affected patients (II-2, II-3, II-4), massive replacement of muscle fibres with adipose tissue was seen (fig. 5). Phase microscopy on resin sections showed dark granules in several muscle fibres in all biopsies (fig. 6), except that of Case II-7, where there were very few fibres. Nuclear abnormalities were not observed. Electron microscopy showed membranous whorls and accumulations of characteristic 15 to 18 nm filaments in the cytoplasm but not in nuclei of muscle fibres (fig. 7). The mother (I-2) and the unaffected brother (II-9) had

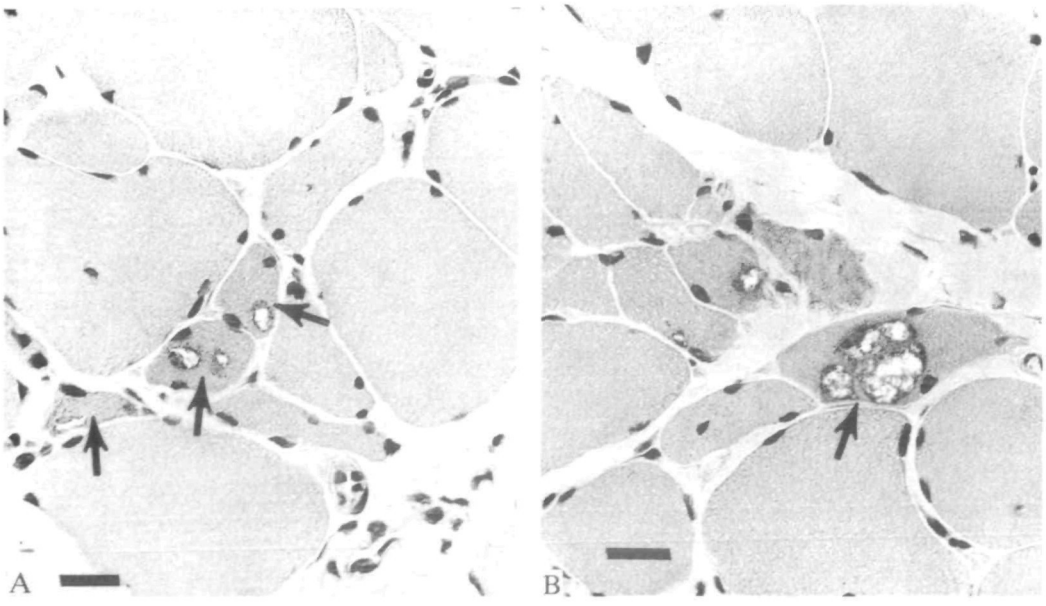

FIG. 3. Biopsy of *Case II-7*. A, arrows mark 3 fibres with prominent rimmed vacuoles. B, a fibre in the centre (arrow) is overwhelmed by rimmed vacuoles. Haematoxylin and eosin. Bar = 25 μm.

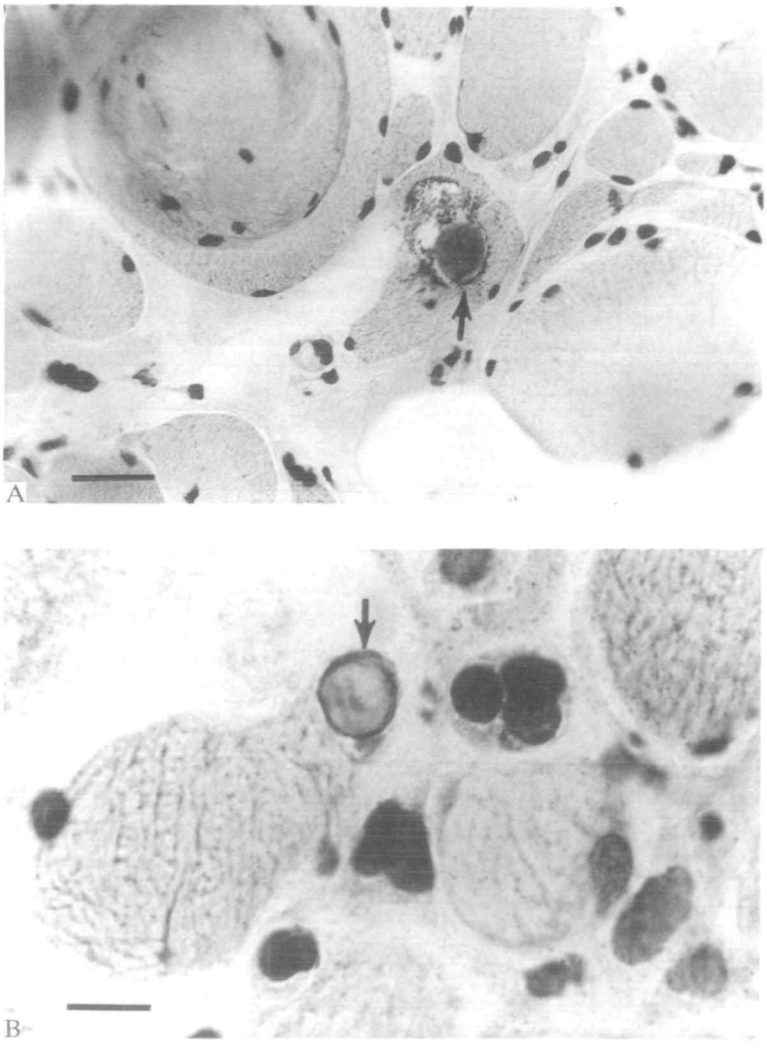

FIG. 4. *Case II-7.* A, muscle biopsy. A massive eosinophilic inclusion body is present in a large rimmed vacuole (arrow). In the left upper corner, a massively hypertrophied muscle fibre shows spatial disorientation of myofibrils. Haematoxylin and eosin. Bar = 25 μm. B, muscle biopsy. In an otherwise normal muscle fibre, a nucleus (arrow) is greatly enlarged by material that displaces the nuclear chromatin to the periphery. Haematoxylin and eosin. Bar = 10 μm.

normal biopsies. The father (I-1), who was entirely asymptomatic, had a number of tubular aggregates in his biceps biopsy. Intramuscular nerves in each case were normal.

Brain imaging

Cranial CT scanning was performed in 4 of the affected patients (II-2, II-3, II-6, II-7) and showed hypodensity of cerebral white matter (fig. 8). MRI of the brain in all

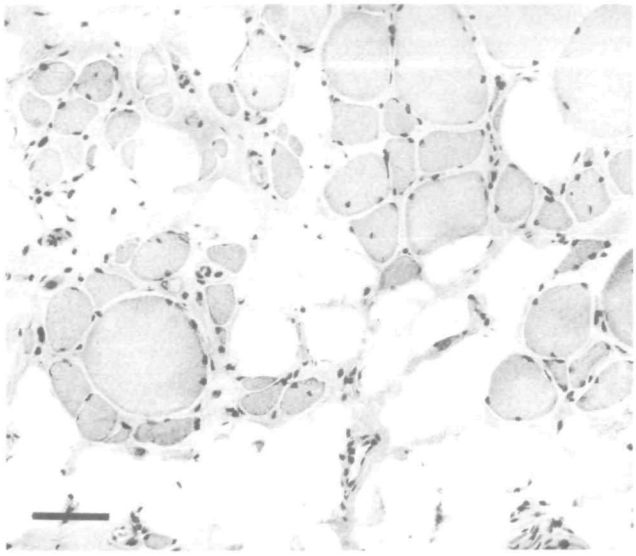

FIG. 5. Biopsy of *Case II-2*. There is prominent loss of muscle fibres with replacement by adipocytes. Most of the remaining fibres are abnormally small or large in calibre. Haematoxylin and eosin. Bar = 100 μm.

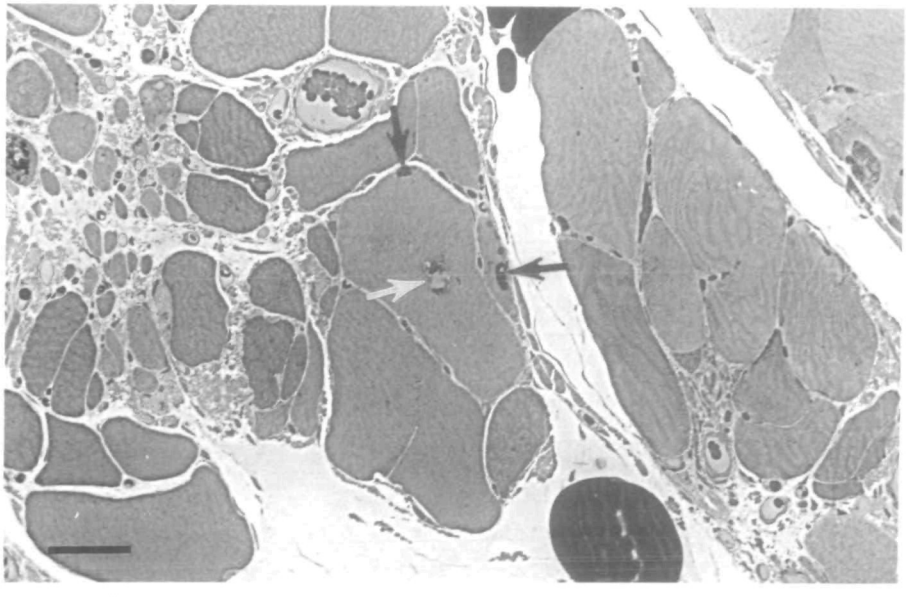

FIG. 6. Biopsy of *Case II-6*. Dark arrows point to membranous whorls, white arrow to a cytoplasmic inclusion bordered by membranous whorls. Note the marked variation in fibre size and collagen deposition between fibres. Resin section, paraphenylene diamine. Bar = 50 μm.

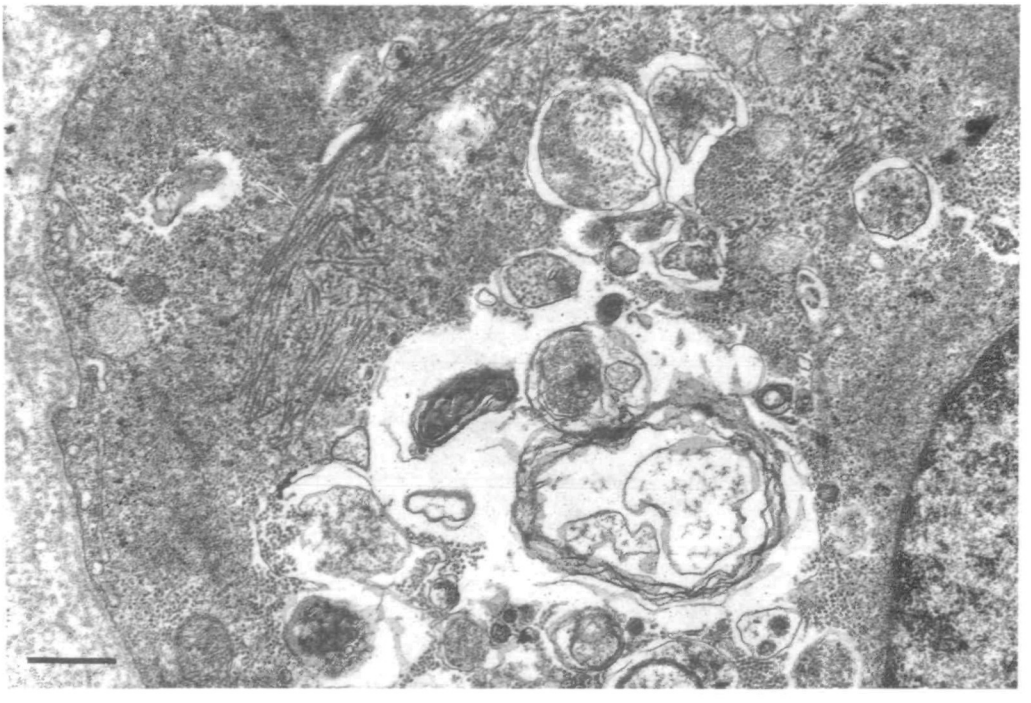

FIG. 7. Biopsy of *Case II-6*. This electron micrograph shows a collection of abnormal filaments (arrow) in a muscle fibre near some membranous whorls. Bar = 0.5 μm.

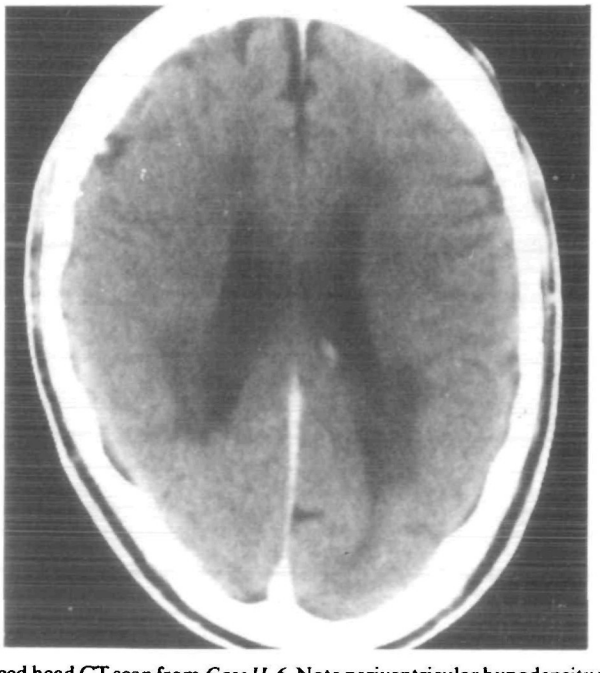

FIG. 8. Contrast-enhanced head CT scan from *Case II-6*. Note periventricular hypodensity with accentuation in the region of the frontal and occipital horns.

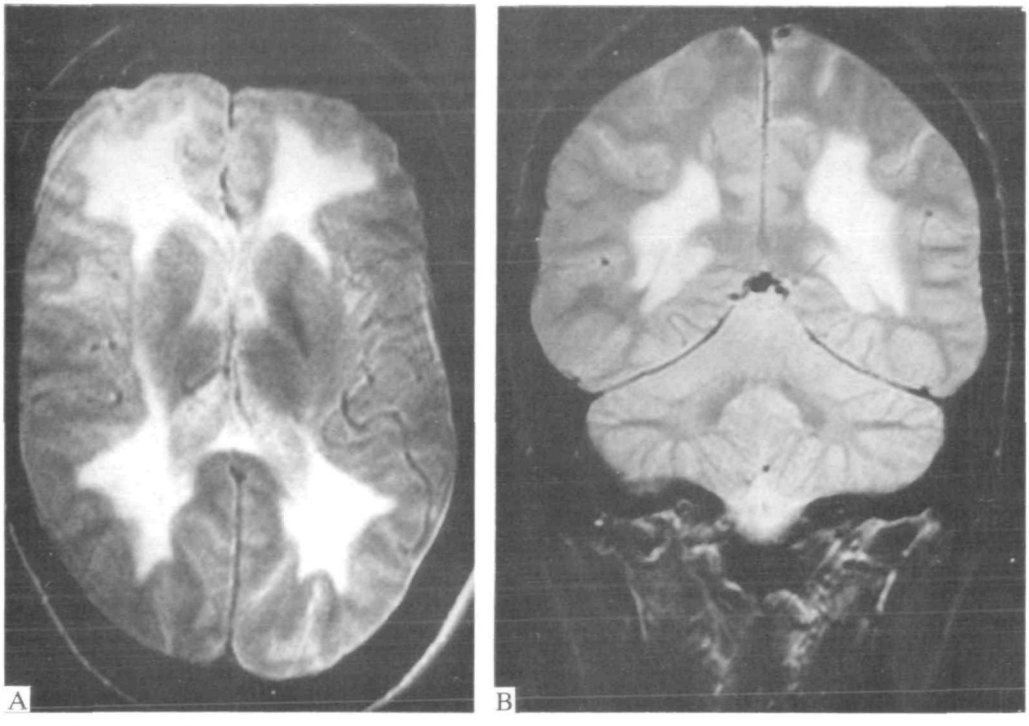

FIG. 9. A, magnetic resonance imaging of brain from *Case II-2*, axial slice. TR = 2100 ms, TE = 60 ms. The cerebral white matter is conspicuous by abnormally high intensity signal. B, magnetic resonance imaging of brain, from *Case II-2*, coronal cut. TR = 2100 ms, TE = 60 ms. The abnormally high intensity signal of the cerebral white matter appears to spare the subcortical U fibres.

affected brothers showed strikingly increased signal intensity from cerebral white matter, including periventricular white matter and the corona radiata, but sparing the subcortical U-fibre system (fig. 9). In 2 cases (II-6, II-7), the increased signal intensity took on a nodular appearance. The severity of white matter change roughly correlated with the severity of myopathy in affected patients. Imaging studies in all unaffected family members were normal.

Additional studies

A 68-yr-old man with pathologically documented typical sporadic IBM was examined with MRI: his cerebral white matter was entirely normal. An unrelated 71-yr-old woman with well documented IBM died of respiratory failure and had a complete postmortem examination. No abnormality of cerebral white matter was detected on gross or microscopic examination. An Iranian-Jewish woman with a progressive myopathy beginning in the third decade, who had a similarly affected brother, demonstrated the typical pathological changes of IBM on muscle biopsy. A brain MRI scan in that patient was normal.

DISCUSSION

Our patients presented with a progressive myopathy associated with evidence of abnormal cerebral white matter on brain imaging. Since the pathological features of the myopathy were indistinguishable from those of inclusion body myositis (IBM), but the clinical picture was atypical for that disease, we shall compare our cases to those in the literature with typical and with atypical forms of IBM. Furthermore, we shall discuss the relationship of the cerebral white matter changes to the syndrome.

The pathological changes found in muscle biopsies in our patients are entirely consistent with the diagnosis of IBM. Variations in fibre calibre, rimmed vacuoles, proliferation of endomysial connective tissue, membranous whorls and inclusions composed of 15 to 18 nm filaments are all characteristic, and in combination have been considered pathognomonic of that disorder. While inflammatory infiltrates may be present in typical IBM, they are not required for diagnosis (Carpenter and Karpati, 1981, 1984) and were absent in our patients. The typical filamentous nuclear or cytoplasmic inclusions present in these patients' biopsies are almost exclusively seen in classical IBM. In a few recent reports, these filaments have been described in association with neuromuscular diseases variously labelled as oculopharyngeal muscular dystrophy and distal myopathy (Fukuhara *et al.*, 1982; Matsubara and Tanabe, 1982; Smith and Chad, 1984). Whether these myopathies are distinct from IBM, or whether they represent variants of IBM cannot presently be determined. Thus the disease specificity of the 15 to 18 nm filaments remains uncertain.

The clinical features of the myopathy observed in these patients differ in a number of significant respects from the usual presentation of IBM. IBM most often begins in the sixth or seventh decade of life. While cases have been described beginning as early as the second decade (Sato *et al.*, 1969; Yunis and Samaha, 1971; Jerusalem *et al.*, 1972; Tomé *et al.*, 1981), onset in early childhood would be extraordinary and has been reported only in 1 case by Eisen *et al.* (1983). Weakness in classical IBM may involve all muscle groups, but is often predominant distally (Carpenter *et al.*, 1978), in contrast to the present cases where weakness was mainly proximal.

A number of reports have described familial myopathies that fulfil the pathological criteria required for a diagnosis of IBM. Eisen *et al.* (1983) described a 23-yr-old man with a slowly progressive distal myopathy beginning in early childhood whose muscle biopsy revealed the typical findings of IBM. That patient had a brother affected with a similar illness who was not studied. Matsubara and Tanabe (1982) reported a brother and sister affected by a progressive distal myopathy during the third decade who also met the biopsy criteria for a diagnosis of IBM. They characterized this disease as an 'autosomal recessive distal myopathy'. Fukuhara *et al.* (1982) described a 39-yr-old man with the clinical picture of 'oculopharyngeal muscular dystrophy' (OMD) that began at the age of 23 yrs with ptosis and proximal muscle wasting. That patient's son was noted to be clumsy at 2 yrs of age, and developed leg weakness and wasting at the age of 6 yrs. Facial

weakness with pronounced distal weakness led to muscle biopsy at 9 yrs. Muscle biopsies in each of these patients showed rimmed vacuoles. Electron microscopy was reported in the second patient only, and demonstrated cytoplasmic inclusions associated with 13 to 19 nm filaments. These authors speculated on the relationship between OMD and distal myopathy and suggested that there might be an aetiological link between the two disorders. They concluded that 'distal myopathy', OMD and IBM were variant forms of the same disease. Argov and Yarom (1984) reported 9 examples of a generalized myopathy sparing the quadriceps in Iranian Jews beginning in the third or fourth decade. Biopsies were characterized by the presence of rimmed vacuoles in affected muscles. Abnormal filaments were not mentioned in the description of electron microscopic findings. Inheritance was felt to be autosomal recessive. Our finding of typical 15 to 18 nm filamentous inclusions in the muscle biopsy from an Iranian-Jewish woman with a familial myopathy beginning during the third decade suggests that this population may indeed suffer from a form of familial IBM. Detailed neuroimaging studies were not described in any of these reports of familial IBM. The Iranian-Jewish woman whom we studied had a normal MRI brain scan, confirming that changes in cerebral white matter are not characteristic of all familial examples of IBM.

The family that is the subject of the present report differs significantly from each of these reported examples of 'familial IBM'. Our patients were all affected early in life, and all demonstrated mainly proximal weakness. Because of the absence of female siblings, as well as the occurrence of this syndrome in only a single sibship, the mode of inheritance cannot be determined with certainty. The absence of female siblings makes it impossible to differentiate between autosomal recessive or X-linked recessive inheritance. The absence of muscle biopsy findings of IBM or of white matter abnormality in either of the parents argues strongly against the possibility of dominant transmission, although incomplete penetrance of the trait or a mutation in either of the parental germ-cell lines cannot be totally eliminated. Maternal inheritance, described in certain mitochondrial cytopathies (Giles *et al.*, 1980; Egger and Wilson, 1983; Rosing *et al.*, 1985) cannot be eliminated in view of the marked phenotypic variability from one generation to the next in those disorders (DiMauro *et al.*, 1985). There are numerous examples of neurological diseases that apparently occur in both familial and sporadic forms, such as Creutzfeld-Jakob disease, Alzheimer's disease, and amyotrophic lateral sclerosis. In each of these diseases hypotheses of vertical transmission of an infectious agent, genetic susceptibility to an environmental agent, and similarity of phenotypic expression of different disease processes have been suggested. Each of these hypotheses could explain the relationship between familial and sporadic IBM. Evidence that IBM is caused by a persistent mumps virus infection (Chou, 1986) has not yet been confirmed.

The white matter hypodensity on CT scans, and the high signal intensity from white matter seen on MRI in these patients appeared as severe in degree as that seen in cases of leucodystrophy, yet the patients had no symptoms of white matter dysfunction. A number of myopathies have been shown to be associated with

abnormal CNS function and/or appearance. It has long been recognized that Duchenne muscular dystrophy and myotonic dystrophy may be associated with mild or moderate mental retardation, and with abnormalities of cerebral cortical architecture (Rosman and Kakulas, 1966). Several reports have emphasized the association of certain congenital muscular dystrophies with CNS changes (Bernier *et al.*, 1979; Nogen, 1980; Fukuyama *et al.*, 1981; Egger *et al.*, 1983; Echenne *et al.*, 1986) which may range from apparently asymptomatic hypodensity of cerebral white matter on CT scanning to widespread abnormality of cerebral architecture involving grey and/or white matter. A number of putative mitochondrial cytopathies have also been associated with CNS white matter abnormalities, for example the Kearns-Sayre-Shy syndrome of progressive external ophthalmoplegia. The clinical picture in our family, however, is inconsistent with any of these diagnoses. Nevertheless, the correlation between severity of white matter change and both clinical and pathological severity of myopathy in our patients suggests that the white matter change and the muscle disease may be related to the same pathogenetic factor. We cannot, however, rule out the possibility that these changes are only related by genetic linkage; completely coincidental occurrence seems unlikely.

The disease described in this report appears to be unique. The familial occurrence of a slowly progressive myopathy beginning in early childhood meeting the pathological criteria for a diagnosis of IBM, associated with a distinctive abnormality of cerebral white matter, suggests that this is a newly recognized syndrome. This may indicate either that the clinical spectrum of IBM is wider than originally thought, or that the pathological changes of IBM are nonspecific and may be found in other disorders. To put it differently, not all progressive muscle diseases with 15 to 18 nm filaments necessarily represent IBM. Only the identification of an underlying biochemical, molecular or immunological defect would allow clarification of the relationship between the disease occurring in our family and cases of typical IBM.

ACKNOWLEDGEMENTS

We wish to thank Dr D. Melanson for interpretation of neuroradiological investigations, Dr L. S. Wolfe for biochemical examinations, Dr D. Gendron for performance of EMG and NCV studies, Dr L. F. Quesney for interpretation of VER and BAEP studies, Dr P. Ashby for performing an MRI scan on one of the additional patients and Drs J. Mathieu and A. Bellevance for referring the patients for investigation.

REFERENCES

ARGOV Z, YAROM R (1984) 'Rimmed vacuole myopathy' sparing the quadriceps: a unique disorder in Iranian Jews. *Journal of the Neurological Sciences*, **64**, 33-43.

BERNIER J-P, BROOKE MH, NAIDICH TP, CARROLL JE (1979) Myoencephalopathy: cerebral hypomyelination revealed by CT scan of the head in a muscle disease. *Transactions of the American Neurological Association*, **104**, 244-246.

CARPENTER S, KARPATI G (1981) The major inflammatory myopathies of unknown cause. *Pathology Annual*, **16**, Part 2, 205-237.

CARPENTER S, KARPATI G (1984) Methods of tissue removal and preparation. In: *Pathology of Skeletal Muscle*. New York and Edinburgh: Churchill Livingstone, pp. 39-61.

CARPENTER S, KARPATI G, HELLER I, EISEN A (1978) Inclusion body myositis: a distinct variety of idiopathic inflammatory myopathy. *Neurology, New York*, **28,** 8-17.

CHOU SM (1986) Inclusion body myositis: a chronic persistent mumps myositis? *Human Pathology*, **17,** 765-777.

DIMAURO S, BONILLA E, ZEVIANI M, NAKAGAWA M, DEVIVO DC (1985) Mitochondrial myopathies. *Annals of Neurology*, **17,** 521-538.

ECHENNE B, ARTHUIS M, BILLARD C, CAMPOS-CASTELLO J, CASTEL Y, DULAC O, FONTAN D, GAUTHIER A, KULAKOWSKI S, MEURON G DE, MOORE JR, NIETO-BARRERA M, PAGES M, PARAIN D, PAVONE L, PONSET G (1986) Congenital muscular dystrophy and cerebral CT scan anomalies: results of a collaborative study of the Société de Neurologie Infantile. *Journal of the Neurological Sciences*, **75,** 7-22.

EGGER J, WILSON J (1983) Mitochondrial inheritance in a mitochondrially mediated disease. *New England Journal of Medicine*, **309,** 142-146.

EGGER J, KENDALL BE, ERDOHAZI M, LAKE BD, WILSON J, BRETT EM (1983) Involvement of the central nervous system in congenital muscular dystrophies. *Developmental Medicine and Child Neurology*, **25,** 32-42.

EISEN A, BERRY K, GIBSON G (1983) Inclusion body myositis (IBM): myopathy or neuropathy? *Neurology, Cleveland*, **33,** 1109-1114.

FUKUHARA N, KUMAMOTO T, TSUBAKI T, MAYUZUMI T, NITTA H (1982) Oculopharyngeal muscular dystrophy and distal myopathy. *Acta Neurologica Scandinavica*, **65,** 458-467.

FUKUYAMA Y, OSAWA M, SUZUKI H (1981) Congenital progressive dystrophy of the Fukuyama type: clinical, genetic and pathological considerations. *Brain and Development*, **3,** 1-29.

GILES RE, BLANC H, CANN HM, WALLACE DC (1980) Maternal inheritance of human mitochondrial DNA. *Proceedings of the National Academy of Sciences of the USA*, **77,** 6715-6719.

JERUSALEM F, BAUMGARTNER G, WYLER R (1972) Virus-ähnliche Einschlüsse bei chronischen neuromuskulären Prozessen: elektronenmikroskopische Biopsiebefunde von 2 Fällen. *Archiv für Psychiatrie und Nervenkrankheiten*, **215,** 148-166.

MATSUBARA S, TANABE H (1982) Hereditary distal myopathy with filamentous inclusions. *Acta Neurologica Scandinavica*, **65,** 363-368.

NOGEN AG (1980) Congenital muscle disease and abnormal findings on computerized tomography. *Developmental Medicine and Child Neurology*, **22,** 658-663.

ROSING HS, HOPKINS LC, WALLACE DC, EPSTEIN CM, WEIDENHEIM K (1985) Maternally inherited mitochondrial myopathy and myoclonic epilepsy. *Annals of Neurology*, **17,** 228-237.

ROSMAN NP, KAKULAS BA (1966) Mental deficiency associated with muscular dystrophy: a neuropathological study. *Brain*, **89,** 769-788.

SATO T, WALKER DL, PETERS HA, REESE HH, CHOU SM (1969) Myxovirus-like inclusion bodies in chronic polymyositis: electron microscopic and viral studies. *Transactions of the American Neurological Association*, **94,** 339-341.

SMITH TW, CHAD D (1984) Intranuclear inclusions in oculopharyngeal dystrophy. *Muscle and Nerve*, **7,** 339-340.

TOMÉ FMS, FARDEAU M, LEBON P, CHEVALLAY M (1981) Inclusion body myositis. *Acta Neuropathologica, Berlin*, Supplement 7, 287-291.

YUNIS EJ, SAMAHA FJ (1971) Inclusion body myositis. *Laboratory Investigation*, **25,** 240-248.

(Received August 13, 1987. Revised November 3, 1987. Accepted November 16, 1987)