TECHNICAL REPORT
11-73
業 績 報 告 書

CHRONIC MENINGOENCEPHALOMYELITIS WITH SPASTIC SPINAL PARALYSIS

けいれん性脊髄麻痺を呈した慢性髄膜脳脊髄炎

CASE REPORT
症例報告

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ABCC JNIH

ATOMIC BOMB CASUALTY COMMISSION

国立予防衛生研究所－原爆傷害調査委員会

JAPANESE NATIONAL INSTITUTE OF HEALTH OF THE MINISTRY OF HEALTH AND WELFARE

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本症例についてご指導を仰ぎ，かつ有益な討議によって本報告をご援助いただいた次の各病理専門医に謝意を表したい．Washington D. C. の米軍病理学研究所 Dr. J. L. Hansen，Dr. M. N. Hart，および Dr. J. M. Henry，国立九州メディカル・センター武谷止孝博士，ならびに東京都鶴風会脳性麻痺研究所松山春郎博士．

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SUMMARY

A case of chronic meningoencephalomyelitis in a 48-year-old housewife is presented. The onset was characterized by spastic paralysis of the lower extremities. The course was progressive with repeated remissions and exacerbations, and the patient died approximately 7 years after the onset of disease. Laboratory tests showed slightly increased cell count in the spinal fluid, accelerated sedimentation rate, positive CRP and RA, and increased ASLO and gamma globulin levels. Neuropathologic examination revealed such changes as perivascular cellular infiltration, glial nodules, poorly demarcated demyelination, and recent necrosis in the spinal cord and basal ganglia. Only mild inflammatory findings were noted in the telencephalon and brain stem. The clinicopathologic findings in this case supported a diagnosis of chronic meningoencephalomyelitis which could not be classified as any known type of encephalomyelitis.

INTRODUCTION

In recent years there has been much progress in elucidating the etiology of certain previously obscure types of encephalitis. There is increasing evidence to establish measles virus as one of the causes of a sporadic form of this disease, and a slow acting

要 約

48歳の主婦に認められた慢性髄膜脳脊髄炎の1症例について報告する．発病時の特徴としては，下肢のけいれん性麻痺が認められた．寛解と悪化とを繰り返しながら進行性に経過し，発病後約7年で死亡した．臨床検査では髄液細胞の軽度の増加，赤沈値の促進，C反応性蛋白およびリウマチ因子検査の陽性，および抗ストレプトリジンO値およびガンマーグロブリン値の亢進を認めた．神経病理学的検査においては，脊髄・基底核に血管周囲の細胞浸潤，グリア結節，境界不鮮明な脱髄，新鮮壊死などの病変を認めた．終脳および脳幹においてはごく軽度の炎症性所見しか認められなかった．本症例の臨床・病理学的所見は既知の脳炎に属せしめ難い慢性髄膜脳脊髄炎の診断を支持した．

はじめに

近年，これまで原因不明の型であった脳炎の病因究明に非常な進歩がみられる．この疾患のうち，散発型のものの1因として麻疹ウイルスが確立されつつあり，非定型

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virus has been suspected to be the cause of atypical encephalitis.[1-7] However, except for general paresis, there has been little progress in establishing the etiology of chronic encephalomyelitis.

Recently we studied a case of chronic encephalomyelitis in which, at autopsy, the pathologic lesions were predominantly in the spinal cord and basal ganglia. The clinical course was one of spastic spinal paralysis. The etiology was not determined. This case is felt by the authors to be quite different from previously described cases of chronic encephalomyelitis and therefore worthy of report.

CLINICAL HISTORY

The patient, a 48-year-old housewife at the time of death on 22 March 1971, first developed neurological symptoms sometime in 1964. She denied a family history of neurologic disease and had not been exposed to the atomic bomb.

Present History. The patient first noted pain in the soles of both feet, pain and cramping in the calves, and a stiffness in her legs while walking. These symptoms gradually progressed accompanied by increased difficulty in walking. Sometime during 1964 she suffered an illness diagnosed as pyelonephritis. About 1966, the pain in the legs decreased and she was able to walk unassisted. In 1967, marked weakness of the legs recurred but without pain. There was severe gait disturbance.

Neurologic examination during initial hospitalization in July 1969 showed no cranial nerve involvement. The gait was spastic and slow. There was no muscular atrophy or fibrillary tremor. Grip strength was 14kg on the left and 17 kg on the right. There were no cutaneous sensory disturbances. The deep tendon reflexes of all extremities were bilaterally symmetrical, but markedly accentuated especially in the upper extremities. Babinski's, Chaddock's, Schaeffer's, and Mendel-Bechtrew's reflexes were positive bilaterally, but Oppenheim's and Rossolimo's reactions were negative. Ankle clonus was positive bilaterally. No cerebellar signs or abnormalities of the extrapyramidal system were noted.

Laboratory Findings. On lumbar puncture, the pressure was normal with normal Queckenstedt's test; cell count 21 mm3; protein 30 mg/100 ml; sugar 74 mg/100 ml; Nonne-Apelt negative; Pandy negative. Serological tests of the blood and spinal fluid for syphilis were all negative. X-ray examination of the cervical, thoracic and lumbar vertebrae was unremarkable. Funduscopy showed mild arteriosclerosis. Serum studies: CRP weakly positive;

脳炎の病因として遅活性ウイルスが推定されている.[1-7] しかし，慢性脳脊髄炎の病因については，進行麻痺を除けば，ほとんど確立されていない.

最近，著者らは，剖検で病理学的病変が主として脊髄および基底核に認められた慢性脳脊髄炎の1症例を経験した．臨床経過はけいれん性脊髄麻痺に一致するものであった．病因は決定するに至らなかった．この症例は，従来報告されている慢性脳脊髄炎例とはかなり異なっているように思われるのでここに報告する.

臨床経過

死亡時の1971年3月22日に48歳であった主婦．最初に神経症状が発現したのは1964年であるが詳細は不明．神経系疾患の家族歴はないという．原爆に被爆していない.

現病歴： 最初，両足底部の疼痛を訴えるようになり，歩行時，腓腹筋に疼痛やけいれん並びに足が棒のようになることに気づいた．これらの症状は次第に進行し，それに伴って歩行障害も強くなった．1964年ころに腎盂腎炎の診断を受けた．1966年ころ，下肢の疼痛は軽減し，介助がなくて歩行できるようになった．1967年には下肢の脱力が再発したが，疼痛はなかった．歩行障害は悪化していた.

1969年7月の初回入院時における神経学的検査では，脳神経障害は認められなかった．歩行はけいれん性で緩徐．筋萎縮または筋線維性攣搦はみられない．握力は左14kg，右17kg．皮膚の知覚障害はなかった．四肢深部腱反射は両側対称性であるが，高度に亢進していた．これは上肢よりも下肢に著しかった．Babinski, Chaddock, Schaeffer, および Mendel-Bechtrew の病的反射が，両側ともに認められたが，Oppenheim および Rossolimo の反射は陰性．足間代は両側とも陽性．小脳症状や錐体外路症状は認められない.

臨床検査所見： 腰椎穿刺では髄液圧は正常で，Queckenstedt 試験の結果も正常．細胞数21 mm3；蛋白30 mg／100 ml；糖74 mg／100 ml；Nonne-Apelt 陰性；Pandy 陰性．血液および髄液の梅毒反応はすべて陰性．頚椎，胸椎，および腰椎のX線検査でも著変なし．眼底検査では軽度の動脈硬化を認めた．血清生化学的検査:

antistreptolysin O titer 333 Todd units; RA strongly positive; LE cell and factor negative; NPN 29.4 mg/100 ml; alkaline phosphatase 4.7 k.k. units; SGOT 17 units; SGPT 20 units; Fe 54.8 μg/ml: TTT 19.4 units; ZTT 24.0 units; total protein 7.4 g/100 ml; A/G 0.94. Serum protein fractions: albumin 48.0%; alpha-1 globulin 2.5%; alpha-2 globulin 10.0%; beta globulin 7.5%; gamma globulin 32.0%; IgG 2102 mg/100 ml; IgA 668 mg/100 ml; IgM 475 mg/100 ml; total Ig 3245mg/100ml. Sedimentation rate 104mm (60 min); 136 mm (120 min).

She was rehospitalized from 27 September to 6 November 1969. At this time EMG showed an upper motor neuron lesion of the muscles of both lower extremities. Myelography was unremarkable. During this hospitalization she developed dysuria. During her third hospitalization from 26 December 1969 to 1 April 1970, ECG revealed coronary insufficiency. She developed pyelonephritis but her neurological symptoms remained unchanged, and no cutaneous sensory disturbances were noted. Recorded blood pressure varied from 160/104 to 186/90 mmHg. Subsequently she had loss of appetite, became markedly emaciated and died on 22 March 1971. There was no record of any definite cerebral symptoms during the entire course of her illness (approximately 7 years).

Treatment. The following medications were begun at the time of initial hospitalization and were continued throughout her course: 1-α-methyl-dopa (Aldomet), Diazepam (Cercine), 2-6-Bis (Diethanollamine)-4-8-dipiperidin-pyrimid-pyrimin (Persantin), Serpacil, Apresolin, Esidrex, Prednisolon (Predonin), and vitamins.

Autopsy Findings. The brain weighed 1275 g. The dura and leptomeninges were normal. There were no external central nervous system abnormalities. The vessels at the base of the brain showed moderate arteriosclerosis, but were not occluded. There was no evidence of increased intracranial pressure. Multiple cut surfaces revealed no focal lesions.

Examination of histologic sections of the cerebrum showed mild focal cellular infiltration, mostly by lymphocytes, in the pia mater of the left occipital and right frontal lobes (Figure 1). The nerve cells of the cortex were well preserved. Mild perivenous cellular infiltration and occasional mild glial accumulations were seen in the subcortical white substance of the parietal, occipital, and frontal lobes. There was no definite demyelination or tissue necrosis.

In the basal ganglia small foci of fresh necrosis were seen in the left caudate nucleus and globus pallidus

CRP は弱陽性．ASLO 値は，333 Todd 単位，RA 強陽性，LE細胞およびLE因子は陰性； 残余窒素29.4 mg ／100 ml； アルカリ・フォスファターゼ 4.7 kk 単位； SGOT 17単位； SGPT 20単位； 鉄54.8 μg／ml； TTT 19.4単位； ZTT 24.0； 総蛋白量 7.4 g／100 ml； A／G 0.94. 血清蛋白分画：アルブミン48.0%； α－1 グロブリン 2.5%； α－2 グロブリン10.0%； β－グロブリン 7.5%； γ－グロブリン32.0%； IgG 2102 mg／100 ml； IgA 668 mg／100 ml； IgM 475 mg／100 ml； 総 Ig 3245 mg／100 ml ．赤沈率 104 mm（60分）～ 136 mm（120分）．

第2回目の入院は1969年 9 月27日から同年11月 6 日まで．EMGでは，両下肢筋に上位運動ノイロンの病変が認められた．脊髄造影法X線検査では著変なし．この入院中排尿障害が発現した．第 3 回目の入院は1969年12月26日から1970年 4 月 1 日まで．心電図検査では冠動脈不全が認められた．腎盂腎炎を併発したが，神経学的症状はいぜんとして変わらず，皮膚知覚障害は認められなかった．記載された血圧は，160／104－186／90 mmHg であった．その後，しだいに食欲が減退し，るいそう著しく，1971年 3 月22日に死亡．全経過の約 7 年間に明確な脳症状の記載はみられない．

治療： 初回入院時に次の投薬が開始され，入院期間中継続された： 1－α－methyl－dopa (Aldomet)，Diazepam (Cercine)， 2－6－Bis (Diethanollamine)－4－8－dipiperidin - pyrimid - pyrimin (Persantin)，Serpacil，Apresolin，Esidrex，Prednisolon (Predonin)，およびビタミン類．

剖検所見： 脳重1275 g．硬膜および軟膜は正常．中枢神経系に肉眼的異常はない．脳底血管には中等度の動脈硬化像が認められたが，閉塞の所見はない．脳圧亢進の所見もない．多くの割面でも限局巣は認められない．

大脳の組織学的検査では，左後頭葉および右前頭葉の軟膜に主としてリンパ球による軽度の細胞浸潤が認められた（図 1）．皮質の神経細胞はよく保たれている．頭頂後頭葉および前頭葉の皮質下白質にごく軽度の血管周囲性細胞浸潤および軽度のグリア集簇が散在している．明確な脱髄や組織壊死はなかった．

基底核では，左尾状核および淡蒼球に新鮮な小壊死巣が

with cellular infiltration around the vessels in the neighboring areas. Small foci of fresh malacia containing gitter cells and hypertrophied glia were present in the right caudate nucleus and globus pallidus, and in these areas advanced arteriosclerosis (Figure 2), perivascular cellular infiltration and glial nodules were seen. Similar lesions were also seen in the medial thalamic nucleus bilaterally (Figure 3). Occasional mild glial accumulations and glial nodules were seen in the right lateral thalamic nucleus and hypothalamus (Figure 4). These lesions were definitely more marked than those in the telencephalon. Several calcified nerve cells were seen near foci of necrosis.

Mild perivascular cellular infiltration and mild glial accumulations were seen in the left substantia nigra and periaqueductal area in the pons as well as in the medulla oblongata. There was no demyelination of the pyramid. The lesions of the brain stem were definitely less marked than those of the basal ganglia. Mild, focal perivascular cellular infiltration of the same degree as in the telencephalon was noted in the white substance of the cerebellum but without glial nodules. The optic nerves showed no demyelination or cellular infiltration.

Lesions of the white substance were most striking in the spinal cord and included demyelination of the pyramidal tract, glial nodules and perivascular cellular infiltration. Glial nodules were also seen in the grey substance of the cervical spinal cord (Figure 5). Anterior horn cells were decreased throughout, especially in the thoracic and lumbar spinal cord. The remaining cells showed chromatolysis and atrophy. The lateral and anterior funiculi of the entire pyramidal tract were spongy, and myelin stain showed poorly demarcated areas of demyelination. Trichrome stain showed proliferation of connective tissue in areas corresponding to the areas of demyelination. Here, there was loss and destruction of both the myelin sheath and axons, with occasional gitter cells and astroglia but axons were better preserved than myelin sheath (Figure 6). In addition, many corpora amylacea were seen in the white substance throughout the spinal cord, but the relation of these bodies to the lesions described above is not known. Histologic sections from both psoas, gastrocnemius and biceps muscles showed diffuse neurogenic atrophy (Figure 7). No inclusion bodies were seen in any of the sections examined. The lesions were most marked in the thoracic spinal cord, followed by those in the cervical and lumbar spinal cord (Figure 8). The vessels in the spinal cord were completely normal.

The principal autopsy diagnosis was chronic meningoencephalomyelitis, etiology undetermined.

あり，それに近い血管周囲に細胞浸潤が認められた．右尾状核および淡蒼球に，脂肪顆粒細胞および肥大グリアなどを含む新鮮な小軟化巣がみられ，これらの領域に進行した動脈硬化（図２），血管周囲性細胞浸潤およびグリア小結節が存在する．左右視床内側核にも同様な病変がある（図３）．さらに右視床外側核および視床下部には，軽度のグリア集簇およびグリア結節が散在する（図４）．これらの病変は，終脳のそれよりも明らかに著明である．壊死巣の近くには石灰化した神経細胞がいくつか認められる．

左黒質および橋の中脳水道周囲ならびに延髄に軽度の血管周囲性細胞浸潤および軽度のグリア集簇がみられる．錐体の脱髄はない．脳幹の病変は基底核のそれよりも明らかに軽い．小脳の白質には，終脳と同程度の血管周囲性細胞浸潤が認められるが，グリア結節はない．視神経には，脱髄も細胞浸潤もない．

脊髄では，白質の病変が最も著しくて錐体路の脱髄，グリア結節および血管周囲性細胞浸潤が認められる．グリア結節は頚髄の灰白質にも認められる（図５）．前角細胞は全域にわたって減少しており，胸髄および腰髄においてそれが強い．残りの細胞も染色質融解および萎縮の像を呈する．錐体路の全域にわたって側索および前索が海綿状を呈し，髄鞘の染色では境界不鮮明な脱髄が認められる．Trichrome 染色を行ったところ，この脱髄部に一致して結合織の増生がみられる．この部では，髄鞘・軸索ともに消失・崩壊しており，脂肪顆粒細胞および星型グリアが散見されるが軸索は髄鞘よりもよく保存されている（図６）．その他，脊髄の全域にわたって白質にアミロイド小体が多数認められるが，これと上記の病変との関係は明らかでない．腰筋，腓腹筋および二頭筋の組織切片を調べたが，これらはすべて瀰漫性神経原性萎縮を示した（図８）．検索した限りでは，封入体はみられなかった．この病変は胸髄において最も強く，次いで頚髄，腰髄の順である（図８）．脊髄内の血管は全く正常である．

主要剖検診断は病因不明の慢性髄膜脳脊髄炎であった．

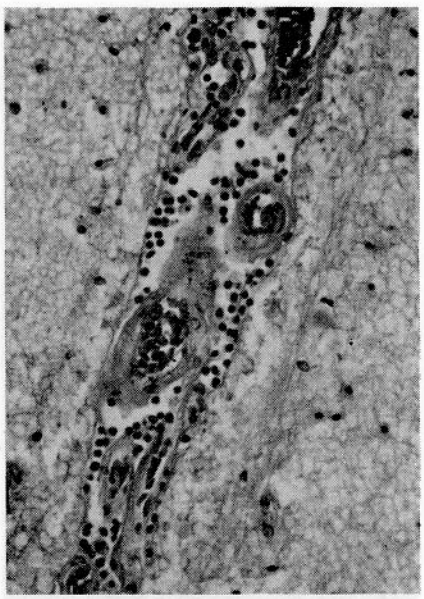

Figure 1. Left frontal lobe, mild meningeal infiltration with lymphocytes (hematoxylin-eosin ×260).

図1. 左前葉の髄膜における軽度のリンパ球浸潤(H－E染色，×260)

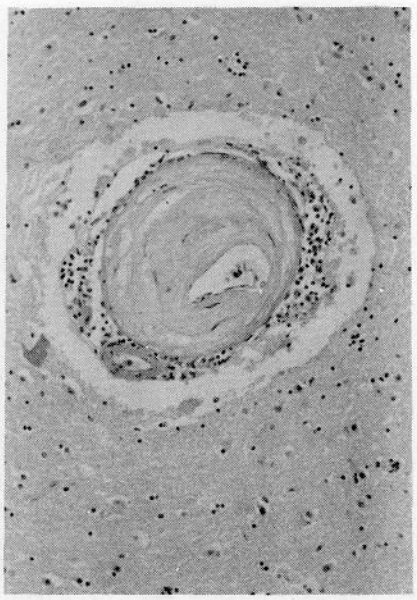

Figure 2. Right putamen, advanced arteriosclerosis (hematoxylin-eosin ×130).

図2. 右被殻における進行した動脈硬化(H－E染色，×130

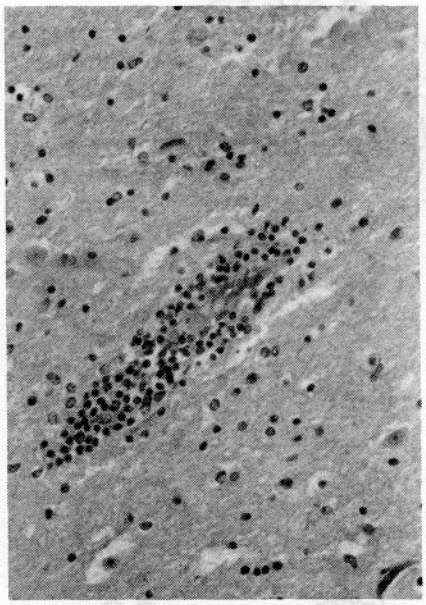

Figure 3. Left thalamus, perivascular infiltration (hematoxylin-eosin ×260).

図3. 左視床における血管周囲性細胞浸潤(H－E染色，×260)

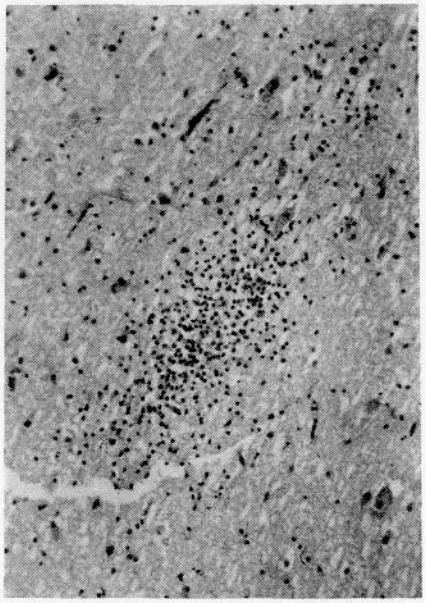

Figure 4. Right hypothalamus, glial nodule (hematoxylin-eosin ×130).

図4. 右視床下部におけるグリア結節(H－E染色，×130)

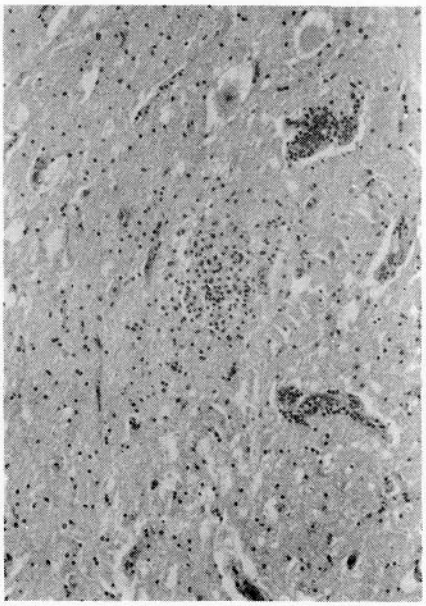

Figure 5. Anterior horn in the cervical spinal cord, glial nodule and perivascular infiltration (hematoxylin-eosin ×130).

図5. 頸髄の前角におけるグリア結節と血管周囲性細胞浸潤
（H－E染色，× 130）

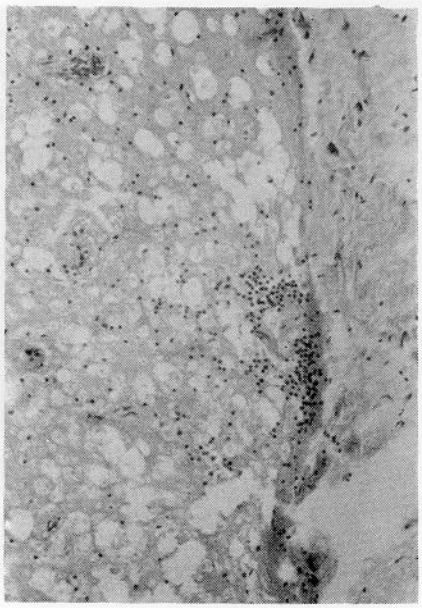

Figure 6. Lateral tract in the thoracic spinal cord, demyelination and glial nodule (Luxol fast blue, ×130).

図6. 胸髄の側路における脱髄とグリア結節
（Luxol fast blue ×130）

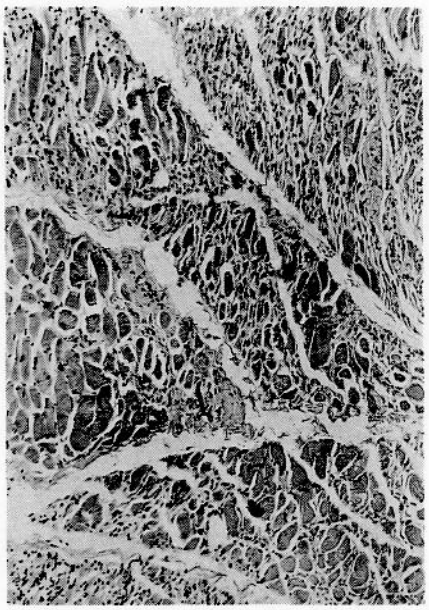

Figure 7. Right gastrocnemius, neurogenic atrophy (hematoxylin-eosin ×67).

図7. 右腓腹筋における神経原性筋萎縮
（H－E染色，× 67）

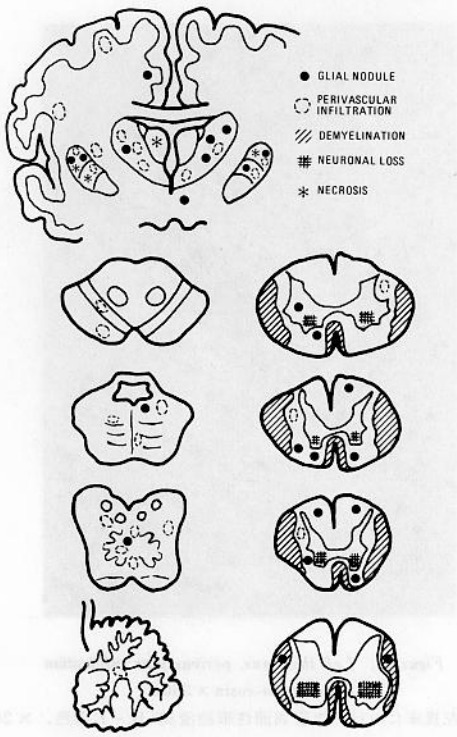

Figure 8. Distribution of lesions.

図8. 病変の分布

The following additional autopsy diagnoses were made: (1) Coronary arteriosclerosis, mild; (2) Arterio- and arteriolosclerosis, severe, kidneys and brain; (3) Acute pyelonephritis with abscess, left; (4) Chronic pyelonephritis, bilateral; (5) Hydronephrosis, bilateral, moderate; (6) Hydroureter, bilateral, moderate; (7) Acute and chronic cystitis, severe; (8) Pulmonary edema, moderate; (9) Pulmonary emphysema, mild; (10) Submucosal abscess, esophagus; (11) Diffuse muscular atrophy, mild to moderate; and (12) Decubitus ulcers, sacrum.

DISCUSSION

The clinical findings in this case included: (1) illness in a 48-year-old female; (2) gradual onset; (3) spastic symmetrical paralysis spreading from lower extremities to upper extremities; (4) presence of diffuse muscular atrophy; (5) progressive course with incomplete remission; and (6) increased cell count in spinal fluid (mild), elevated sedimentation rate, positive CRP and RA, moderately increased ASLO level, and increased serum gamma globulin. Neuropathologic findings included: (1) both perivascular infiltration mainly by lymphocytes and glial nodules and small accumulations of glial cells most striking in the spinal cord and basal ganglia, but also present in the white substance of telencephalon, medulla oblongata, pons, and mesencephalon; (2) irregular demyelination of the spinal pyramidal tract; (3) decrease and degeneration of anterior horn cells; (4) small, localized, fresh areas of necrosis in basal ganglia; and (5) advanced arteriosclerosis localized in basal ganglia.

Although the clinical course might suggest a degenerative or demyelinating disease such as amyotrophic lateral sclerosis and multiple sclerosis, the absence of a bulbar syndrome, the distribution of the muscle atrophy and the occurrence of remissions are different from the clinical course of ALS and the absence of multiple leucoencephalopathy lesions is against the diagnosis of multiple sclerosis. Further, the histologic changes do not support these diagnoses. The mild increase of cells in the spinal fluid and the results of blood chemistry tests cause one to suspect infection. The presence of perivascular cellular infiltration, glial nodules, mild glial accumulations and tissue necrosis as well as the irregularity and non-systemic nature of the demyelination of the spinal cord support the suggestion that this is probably an infectious and possibly a viral disease. The generally mild nature of the inflammatory changes presumably is related to the prolonged use of adrenal cortical agents.

The lesions in the basal ganglia are of at least two types. As part of the generalized arteriosclerosis

その他の剖検診断は次のとおりである：(1) 軽度の冠動脈硬化症；(2) 腎および脳の強度動脈硬化症および細動脈硬化症；(3) 左腎の膿瘍を伴う急性腎盂腎炎；(4) 両側慢性腎盂腎炎；(5) 両側の中等度水腎症；(6) 両側の中等度尿管水瘤；(7) 強度の急性および慢性膀胱炎；(8) 中等度の肺浮腫；(9) 軽度の肺気腫；(10) 食道の粘膜下膿瘍；(11) 軽度から中等度の瀰漫性筋萎縮；(12) 仙骨の褥瘡性潰瘍.

考　察

本症例における臨床所見は次のとおりである：(1) 48歳の女性患者；(2) 緩徐な発病；(3) 下肢から上肢にわたる対称性けいれん性麻痺；(4) 瀰漫性筋萎縮の存在；(5) 不完全寛解を伴う進行性経過；(6) 髄液の細胞増多（軽度），赤沈率の亢進，CRPおよびRA反応陽性，ASLO値の中等度亢進；ならびに血清ガンマグロブリン値の増加．神経病理学的所見は次のとおりである：(1) 主としてリンパ球およびグリア結節から成る血管周囲性浸潤およびグリア細胞の小集簇は，脊髄および基底核において最も顕著であるが，終脳，延髄，橋および中脳の白質にもみられる；(2) 脊髄錐体路の不規則な脱髄；(3) 前角神経細胞の減少および変性；(4) 基底核に限局性の新鮮な小壊死；(5) 基底核に限局性の進行した動脈硬化．

臨床経過から筋萎縮性側索硬化症，多発性硬化症のような変性または脱髄性の疾患も考えられるであろうが延髄症候群の欠如，筋萎縮の分布，寛解の存在等は，筋萎縮性側索硬化症の臨床経過と異なる．一方，多発性白質脳症の病変の欠如は，多発性硬化症の診断を支持しない．また，組織学的変化もこの診断を支持するものではない．髄液細胞の軽度の増加という臨床所見や血液化学検査の結果は感染症を疑わせる．血管周囲性細胞浸潤，グリア小結節，軽度のグリア集簇，組織壊死の存在，ならびに脊髄の不規則性および非系統性脱髄は，本症例がおそらく感染性疾患で，さらにはウイルス性感染であることも考えさせられる．炎症性変化が全般的に軽度であるのは，副腎皮質剤の長期投与の影響によるものと考える．

基底核の病変には少なくとも2種類ある．体の他の部位

present elsewhere in the body, there are severe arteriosclerotic vascular changes and in addition, there are areas of recent necrosis and glial accumulations which presumably are related to reduced local resistance to the underlying encephalitic process.

The most appropriate neuropathologic diagnosis seems to be meningoencephalitis universalis proliferativa et necrotica. Since the lesions of the telencephalon are mild and scattered, the use of the term universalis may be questioned. However, the similarity of the lesions in the telencephalon to those elsewhere indicates that they are all part of the same process, and the term seems justified.

The possibility that this was a case of Japanese summer encephalitis was considered. Initially, the patient had neurologic symptoms of the lower extremities which developed so gradually that the seasonal time of onset is vague. The usual acute symptoms of Japanese encephalitis (fever, headache, vomiting, disturbances of consciousness, etc.) were never noted. Serologic tests of the spinal fluid for Japanese encephalitis were not made, but, except for slight increase of cells, examination of the spinal fluid showed normal pressure and protein content and no abnormality. A clinical diagnosis of Japanese encephalitis seems to be unreasonable in this case. From the neuropathologic standpoint, the main site of lesions in Japanese encephalitis are in the diencephalon, mesencephalon and telencephalon, and there are, almost without exception, lesions in the thalamus even in protracted cases.[8-12] One of the authors,[13] reported that many patients with subacute or chronic Japanese and other encephalitis have marked lesions of the mesencephalon and brain stem and that this region seems to be one of the areas susceptible to encephalitis. However, lesions of the telencephalon in this case consisted only of occasional, mild perivascular cellular infiltrations and small glial accumulations in the subcortical white substance, with no evidence of polioencephalitis. Further, the dominant lesion in the spinal cord militates against the possibility that this patient had Japanese encephalitis.

Of the known types of chronic encephalitis, general paresis was ruled out in this case by the negative blood and spinal fluid serological tests and by the absence of marked lesions in the frontal lobe and of Hortega's glia.

Conditions previously called inclusion body encephalitis and subacute sclerosing leukoencephalitis (SSLE) are now considered to belong to a single category described as subacute sclerosing panencephalitis with lesions which are mainly in the white

にある全身性動脈硬化症としての強度の動脈硬化性血管変化がある．それと新鮮な壊死やグリア集簇であるこれらは恐らく根底にある脳炎過程による局所の抵抗減弱と関連があると考えられる．

神経病理学的診断としては，増殖性および壊死性びまん性髄膜脳炎とするのが最も妥当であると思われる．終脳の病変は軽度でかつ散在性であるため，びまん性という用語の使用には疑問があるかもしれない．しかし，終脳の病変とその他の部位の病変とが近似していることは，それらがすべて同じ疾患過程の部分であることを示しておりその用語も妥当であろう．

本症例が，日本夏期脳炎でありうるとも考えられる．下肢の神経症状が初発症状できわめて徐々に発病したので，発病季節もあいまいであった．日本脳炎に見られる急性症状(発熱頭痛，嘔吐，意識障害など)は全く認められていない．日本脳炎を調べる髄液の血清学的検査は行なわれていないが，軽度の細胞増加を除けば，髄液検査では髄圧および蛋白量は正常で，異常は認められなかった．本症例を臨床的に日本脳炎と診断することは妥当ではないと思われる．神経病理学的に考えると，日本脳炎の病変の主座は間脳，中脳および終脳にあり，視床には遷延例でさえもほとんど例外なく病変が存在する．[8-12] 著者の１人[13]は，亜急性または慢性に経過した日本脳炎およびその他の脳炎では，間脳および脳幹に病変の著しい例が多く，この部位は脳炎に浸されやすい領域の一つであると思われる，と報告している．しかし，本症例における終脳の病変は，皮質か白質にごく軽度の血管周囲性細胞浸潤とグリア小集簇を散見した程度であり，灰白質脳炎の像はみられない．さらに，主として脊髄に病変があることから考えれば，本例では日本脳炎の可能性が除外される．

慢性脳炎として知られているもののうち，本例では進行麻痺は，血液・髄液の両検査ではともに梅毒反応は陰性で，前頭葉の著明な病変も Hortega のグリアの反応も認められなかったので否定される．

従来の封入体脳炎と亜急性硬化性白質脳炎(SSLE)は，今日では亜急性硬化性全脳炎と呼ばれ，同一範疇に属するものと考えられている．この場合，病変は主として終

substance of the telencephalon. In contrast, in our case the lesions of the white substance were mild compared with those of spinal cord and basal ganglia, and no evidence of a sclerosing process was found.

脳の白質に存在する．それに反して，本症例の白質の病変は，脊髄や基底核のものに比べてきわめて軽く，硬化性の徴候は認められなかった．

This case has a number of clinical and pathologic similarities to the brain stem encephalitis described by Iizuka.[14] Clinically these included: (1) onset in adulthood; (2) chronic course; (3) repeated exacerbations and remissions; and (4) increased cell count in the spinal fluid. Pathologically there were: (1) lesions involving the diencephalon and spinal cord; (2) periarteriolar and perivenous cellular infiltration; (3) localized and diffuse glial proliferation and glial growths; (4) absence of inclusion bodies; and (5) chronic degeneration with poorly demarcated demyelination and glial proliferation in the lateral or posterior funiculi of the spinal cord. However, the lesions in our case were concentrated in the spinal cord and basal ganglia and in this respect were quite different from the condition reported by Iizuka.

この例には，飯塚[14]の提唱した脳幹脳炎と臨床的・病理学的にいくつかの類似性がある．すなわち，臨床的には：(1) 成人における発病；(2) 慢性経過；(3) 好転と悪化の反復；(4) 髄液中の細胞数の増加．また，病理学的には：(1) 間脳および脊髄にわたる病変；(2) 細小動静脈周囲性細胞浸潤；(3) 限局性および瀰漫性のグリア細胞増殖ならびにグリア集簇；(4) 封入体の欠如；(5) 脊髄の側索または後索における境界不鮮明な脱髄とグリア細胞の増殖をともなう慢性変性像．しかし，本症例における病変は脊髄および基底核に集中しており，この点においては病変は飯塚が報告した状態とはかなり異なっていた．

The pathologic findings in this case differ from the chronic disseminated encephalomyelitis, brain stem type, described by Shiraki,[15] in which the main site of lesions is the basal portion extending from the medulla oblongata to the mesencephalon, with lesions also present in the optic nerve. The demyelination of the spinal cord in our case was poorly demarcated, non-systematic, and not a primary demyelinating process or demyelinating encephalomyelitis.

白木[15]が記載している脳幹型の慢性播種性脳脊髄炎は，病変の主座が延髄から中脳に及ぶ脳底部にあって視神経にも病変が存在するとされており，本症例における病理学的所見とも異なっている．本症例では，脊髄の脱髄は境界不鮮明で非系統性であって一次性脱髄過程とも脱髄性脳脊髄炎とも考えられない．

Abe[16] reported a case which clinically presented as spastic spinal paralysis and histologically was found to be encephalitis. He noted inflammatory findings in the mesencephalon and medulla oblongata, but no evidence of myelitis even though there was systematic demyelination of the spinal cord. He interpreted his case as being one of encephalitis complicated by demyelinating disease. However, the lesions of the spinal cord he described were quite different from those in our case.

安陪[16]は，臨床的にはけいれん性脊髄麻痺を呈し，組織学的には脳炎であると認められた症例について報告している．彼は，間脳および延髄に炎症所見を認めたが，脊髄に系統的な脱髄はみられたものの，脊髄炎の徴候はなかったと述べている．そして，その症例を脳炎と脱髄疾患との合併例として報告している．しかし，安陪が述べている脊髄の病変は，本症例とはかなり異なったものであった．

West Indian neuropathy[17] is reported as presenting with myelitis and clinical symptoms of spastic paraplegia and is suspected to be due to a viral infection or intoxication. It is described as showing retrobulbar neuropathy, nerve deafness, selective lower motor neuron lesions and posterior column degeneration in addition to spastic paraplegia. Although there is little similarity to our case, it is mentioned in view of the possibility of some common etiology.

脊髄炎が存在しけいれん性対麻痺の臨床的症状を呈するものとしては，西インド諸島の原住民における神経病[17]が報告されており，その病因はウイルスまたは中毒によるものではないかと考えられている．それは，けいれん性対麻痺のほか，球後神経病，神経性難聴，選択的下位運動ノイロンおよび後柱変性を示すものとして記載されている．本症例との類似性はわずかであるが，病因になにか共通点のあることを考えさせる．

As described above, the major known types of chronic encephalitis have been reviewed and our case did not

以上，既知の慢性脳炎について考察したが，そのいずれ

seem to correspond to any of them. The spinal cord lesions were the prominent and characteristic neuropathologic finding in our case, and this was consistent with the clinical course. The necrosis seen in the basal ganglia is considered to be relatively recent in view of the predominance of gitter cells and hypertrophied glia. It is for this reason that psychoneurologic symptoms due to thalamic disturbances were not noted in the initial period.

にも本症例は相当しないように思われる．本症例においては，脊髄病変が顕著であって特徴的神経病理学的所見であったが，これは臨床的経過とも一致するものであった．基底核にみられる壊死は，脂肪顆粒細胞および肥大グリアが優勢であることからみても，比較的新鮮なものと考えられる．そのため，発病初期には，視床障害による精神神経学的症状が認められなかったのであろう．

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