CASE REPORT

MENINGOENCEPHALOMYELITIS AND NEURITIS CLINICALLY DIAGNOSED AS MULTIPLE SCLEROSIS

Samruay Shuangshoti

Department of Pathology, Faculty of Medicine, Chulalongkorn University, Bangkok 10330, Thailand

Meningoencephalomyelitis may give clinical manifestations imicking multiple sclerosis. It. hence, should be clearly distinguished from the latter before rendering the diagnosis of multiple sclerosis, especially in Thailand where it is generally known that multiple sclerosis is exceedingly rare (Shuangshoti and Tangchai, 1983). To be presented herein is an example of meningioencephalomyeloneuritis which was erroneously diagnosed as multiple sclerosis.

An 18-year-old single female student, the native of Pattani Province, complained of quadriparesis for 3 days before entering Chulalongkorn Hospital.

One year prior to occurrence of tetraparesis, she experienced bilateral photophobia wihch required wearing black spectacles. She, developed opsillopsia. Two weeks later, she was found by a private physician to have Romberg's sign. These symptoms and sign disappeared by medical treatment within 1 month. The details of treatment, however, were not available.

Eleven months before admission to Chulalongkorn Hospital, she experienced discomfort at the nape of the neck and numbness in both hands. She, however, could move her neck and limbs as usual. There was no weakness of any extremity.

Ten months prior to hospitalization, she experienced dull deep pain in the neck when she hit her hand on a pillow in practising Judo. This episode prevented her from further preactice.

Nine months prior to admission to hospital, the patient's left eye became progressively blurred and was then blind within 1 month. An ophthalmologist gave her a drug which brought back her normal vision within 1 month.

Twelve days prior to admission to hospital, the deep dull nuchal pain recurred, especially when she extended backward her head and neck. Five days before admission, she developed weakness of the left upper limb. It was hard for her to hold any thing with the left hand. The right upper limb and lower extremities were intact.

Three days prior to admission to Chulalongkorn Hospital, quadriparesis ensued, with particular involvement of the left upper and lower limbs. She, moreover, had numbness of the skin from the chest downward to the lower limbs, dysuria, and dyspnea. The patient then was admitted to a provincial hospital where a myelogram disclosed an enlarged spinal cord from C_1 to C_7. The cerebropinal fluid (CSF) revealed 68 mg/100 ml of protein and 42 mg/100 ml of sugar. A laminectomy from C_1 to C_7 confirmed an enlarged cervical segment of the spinal cord. There was no improvement of the patient's neurologic condition although she received intravenous steroid. The dyspnea persisted. She then was transfered to Chulalongkorn Hospital, 3 days after surgical intervention.

Upon hospitalization, body temperature was 37°C, pulse rate 78 beats/ minute, respiratory rate 28/minute, and blood pressure 120/80 mmHg. The patient was tachypnic, fully conscious, and cooperative. Each pupil, 3 mm in diameter, was reactive to light. There was a left Marcus Gunn phenomenon. Visual acuity was 20/20 bilaterally. Visual fields were normal. Quadriparesis was detected. There was impaired pinprick sensation up to T_4 on the right side and T_6 on the left. Impaired propioception of the upper limbs and Romberg's sign were observed. Babinski's test showed bilateral dorsiflexion of toes.

Laboratory data revealed 13 g /100 ml of hemoglobin, 11,700 leukocytes/mm^3 with 77% neutrophils, and 23% lymphocytes. There were 248,000 thrombocytes/mm^3. Plasma glucose was 130 mg/100 ml, BUN 16 mg/100 ml, creatinine 0.6 mg/100 ml, total bilirubin 0.5 mg/100 ml, direct bilirubin 0.3 mg/100 ml, alkaline phosphatase 127 IU/l, SGOT 12 IU/l, albumin 3.7 g/100 ml, globulin 3.2 g/100 ml,

calcium 9.5 mg/100 ml, phosphate 4.5 mg/100 ml, sodium 144 mEq/l, potassium 3.9 mEq/l, chloride 109 mEq/l, and carbondioxide 22 mEq/l. Examination of the clear CSF demonstrated 1 lym-phocyte/mm^3, 15 mg of protein/mm^3, 68 mg of glucose/mm^3 and negative VDRL test. The urine contained numerous leukocytes and erythrocytes/mm^3 (on Foley's catheter). Culture of the surgical wound yielded slight growth of *Pseudomonas aeruginosa.*

The clinical impression was multiple sclerosis. Steroid was administered. The patient developed intermittent fever (37.2°-39°C). Antibiotics (cefotaxine, cloxacillin and ceftazidine), thus, were added. Terminally, the patient developed severe dyspnea which required artificial oxygenation. She died with generalized convulsions, 30 days in Chulalongkorn Hospital, or 33 days after cervical laminectomy. The total course of ailment was about 1 year.

The general autopsy findings consisted of acute and chronic hemorrhagic cystitis, and hyaline membrane disease of the lungs (780 g).

The main findings were in the central nervous system (CNS). A 1,440-g brain showed severe congestion, edema, and prominent herniation of the cerebellar tonsils. The optic nerves, chiasm, tracts, lateral geniculate bodies, and oculomotor nerves were grossly unremarkable. Various blood vessels were thin-walled. Coronal sections of brain disclosed numerous petechiae on the congested and edematous cut surfaces (Fig 1, 2). The entire spinal cord was also congested and edematous, especially the cervical segment. Petechiae were disseminated throughout substance of the spinal cord (Fig 2).

Microscopically, the optic nerves and chiasm were infiltrated by a moderate number of lymphocytes and microglias. These cells were as well present in the optic nerve sheaths (Fig 3). There was no demyelination in luxol-fast blue stain of these nerves.

The left oculomotor nerve was also infiltrated by similar reactive cells as in the optic nerves. A few foci of demyelination were observed in some bundles (Fig 4).

The pons showed infiltration of lymphocytes and microglias of moderate number in its substance and in adjacent leptomenignes. The neurons comprising pontine nuclei were frequently shrunken; the cytoplasm was cloudy, and nuclei were pyknotic. The nucleoli were obscure (Fig 5). There was status spongiosus of the neuropil.

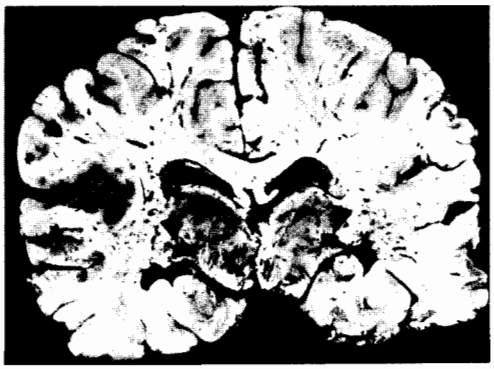

Fig 1–Coronal section of brain through intact lateral geniculate bodies (arrowheads) showing severe congestion and scattered petechiae of the cut surface. Flattening of gyri and narrowing of sulci indicate edema of brain. Scale is in centimeters.

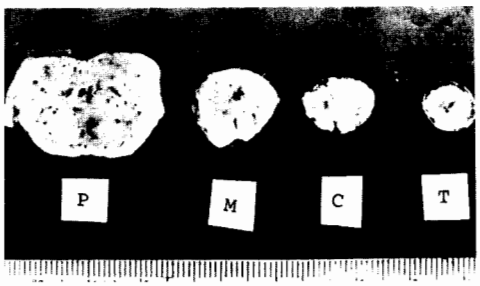

Fig 2–Cross sections of Pons (P), medulla oblongata (M), cervical segment of spinal cord (C) and thoracic portion of spinal cord (T) to show congestion, petechiae, and edema. Scale is in centimeters.

The medulla oblongata, especially its caudal part, showed infiltration of its central portion by many lymphocytes, microglias, and astrocytes to represent glial nodules. (Fig 6). This was also true for the posterior white columns (fasiculi gracillis and cuneatus) of the spinal cord which, in addition, had prominent status spongiosus (Fig 7). The gray matter was infiltrated by many lymphocytes and microglias. The neurons in the anterior gray horn exhibited degenerative changes characterized by cloudy perikaryon with central chromatolysis, pyknotic nuclei which were often eccentric, and indistinct nucleoli. The fish-eye ap-pearance of many nuclei, then, was indistinct. Some anteiror horn cells were surrounded by microglias (Fig 8).

The spinal nerves in the subarachnoid space were

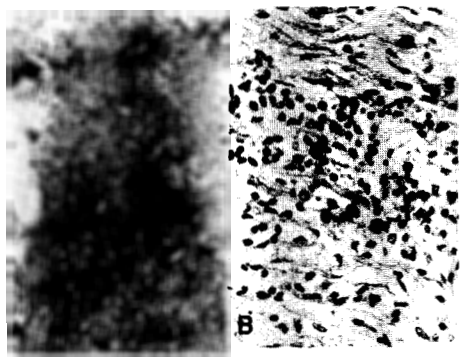

Fig 3–Microscopic features of optics nerve.

(A) Optic nerve and sheath are infiltrated by many reactive cells representing neuritis. Hematoxylin and eosin, × 100.

(B) At high magnification, lymphocytes, histiocytes, and rod-shaped microglias are recognized. Hematoxylin and eosin, × 400.

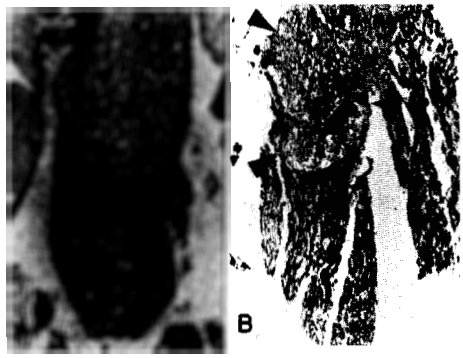

Fig 4–Oculomotor nerve, left.

(A) Infiltration of reactive cells, chiefly lymphocytes, is depicted to represent neuritis. Hematoxylin and eosin, × 100.

(B) The pale area of demyelination (among arrowheads) is shown. Luxol-fast blue stain, × 100.

often edematous and occasionally infiltrated by lymphocytes and microglias. Some showed demyelination of axons (Fig 9) as those perceived in the left oculomotor nerve (Fig 4).

The pathologic diagnoses were nonspecific and nonpurulent meningoencephalomyelitis and neuritis of optic nerves, left oculomotor nerve, and some spinal nerves; and demyelination of left oculomotor nerve and some spinal nersves.

Clinically, the characteristic features in multiple

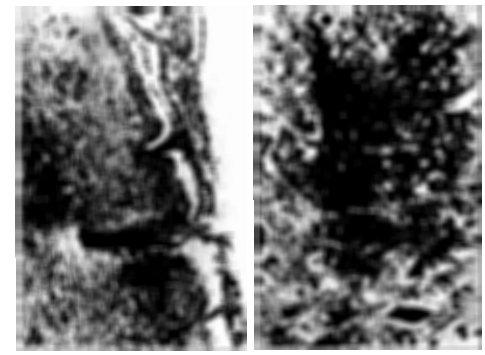

Fig 5–Medulla oblongata.

(A) Infiltration of many reactive cells mainly lymphocytes is observed in the leptomeninges and peripheral portion of medulla. Also note spongy state of the neuropil. Hematoxylin and eosin, × 50.

(B) Many lymphocytes and microglias are scattered individually as well as in clusters. A degenerated neuron lies below; note absence of fish-eye appearance of nucleus and cloudy perikaryon. Hematoxylin and eosin, × 100.

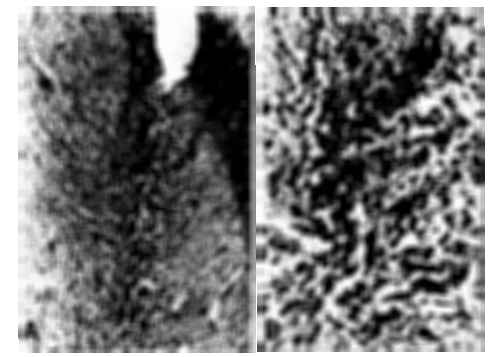

Fig 6–Medulla oblongata.

(A) A large inflammatory focus is below the fourth ventricle (between arrowheads) which is lined by ependyma. Hematoxylin and eosin, × 50.

(B) Microglias and astrocytes form a large glial nodule. Hematoxylin and eosin, × 100.

sclerosis are symptoms and signs disseminated in time. The malady may start with paresthesia, diplopia, decreased vision, weakness, and virtigo. The initial manifestations characteristically disappear spontaneously, and to be replaced by other symptom. The relapse and remission course usually occurs and is suggestive but not diagnostic of multiple sclerosis. In some cases, moreover, the course is steadily progressive with frequent addition of novel findings. Charcot's triad (nystagmus, intention tremor, and scanning speech) is not necessory for diagnosis, but

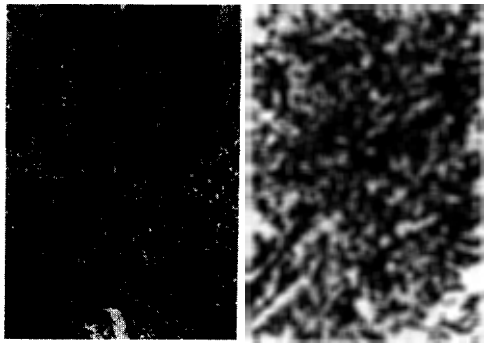

Fig 7–Cervical segment of spinal cord.

(A) The posterior white columns with posterior median sulcus between them (arrowheads) show status spongiosus and reactive cells. Hematoxylin and eosin, × 50.

(B) Lymphocytes, microglias, and oligodendroglias are mingled in white matter of posterior column. Hematoxylin and eosin, × 100.

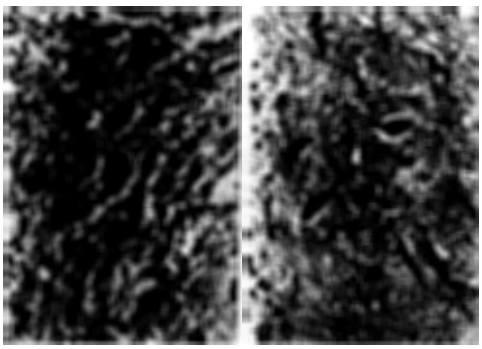

Fig 8–Cervical segment of spinal cord.

(A) and (B) show degenerated anterior horn cells intermingled with many microglias and lymphocytes in gray matter. Hematoxylin and eosin, × 400 each.

often occurs in cases of longstanding multiple sclerosis. The patient is usually young adult, but is rare before10 years of age.

The clinical manifestations in the current young woman thus fit well to multiple sclerosis because the symptoms and signs of the eyes as well as disturbance in sensation and motor weakness in relation to lesions of the spinal cord or brain are all compatible with the aforementioned descriptions. The clinical diagnosis of multiple sclerosis in this case thus was reasonable (Matthews, 1985).

The external pathologic changes of the CNS in

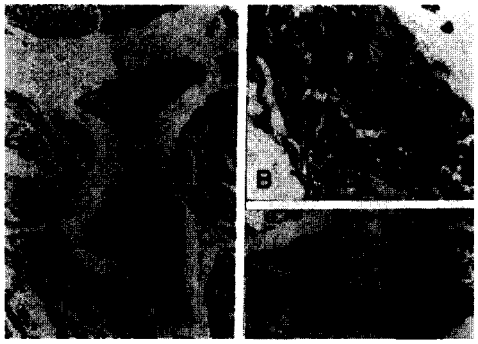

Fig 9–Spinal nerves.

(A) Several spinal nerves lie in subarachnoid space. A nerve (arrowhead) is infiltrated by reactive cells. Others are edematous and pale. Hematoxylin and eosin, × 100.

(B) The same nerve idicated by the arrowhead in (A) demonstrates infiltration of lymphocytes. Hematoxylin and eosin, × 400.

(C) Demyelination is shown in a nerve. Luxol-fast blue stain, × 100.

multiple sclerosis are not clear. There may only be mild gyral atrophy, leptomeningeal opacity, and thin optic nerves. Other cranial and spinal nerves remain unchanged. When the brain is cut plaques of sclerosis are often seen, predominantly in the white matter and paraventricular zone. These plaques are often irregular but well demarcated, and are visible with the naked eye (Weller, 1985).

The pathologic appearance of the brain in the current case, differed from the aforesaid features. The brain and spinal cord were congested, focally hemorrhagic, and edematous. The most important finding that was strongly against the diagnosis of multiple sclerosis in this patient was the involvement of the oculomotor and some spinal nerves. Typically, the cranial and peripheral nerves are not affected in multiple sclerosis, except the optic nerves (Weller, 1985).

The current malady was not subacute necrotizing encephalomyelopathy (Leigh's disease) (Kalimo *et al,* 1979; Hegedüs and Nemeth, 1981; Shuangshoti *et al,* 1994). The individual lesions did not tend to be symmetrical, and proliferation of small blood vessels and endothelium was not observed. Neurons within the lesions, moreover, often showed degenerative changes instead of being relatively intact as in Leigh's disease.

The presence of lesions in the posterior columns of the spinal cord and Romberg's sign suggested the possibility of tabes dorsalis in this young woman. However, this probability was excluded on the basis of a negative VDRL test.

Bechet's disease (or syndrome) must be excluded from the current case of meningoencephalomyeloneuritis. Bechet's disease is a chronic inflammatory disorder characterized by recurring apthous ulceration affecting the oral and pharyngeal mucosae, genital and dermal lesions (such as furunculosis, and erythema nodosum), ocular affections (such as uveitis, iridocyclitis with hypopyon, keratoconjunctivitis, and retinitis or retinal detachment), optic neuritis, arthritis, serositis, and ulcer of the alimentary tract. Involvement of the nervous system which includes meningoencephalitis or myelitis, and neuropathy of the cranial and peripheral nerves has been reported and associated with poor prognosis. The malady is more common in males than in females with a ratio of 3:1. The etiology is unknown but viral, allergic or collagen vascular diseases with vasculitides have been suggested (Wolf *et al,* 1965; Chajek and Fainaru, 1975). Although the present patient had an ocular problem Bechet's disease was excluded. Mucosal, genital, and dermal lesions as well as vasculitides were not observed. Uveitis cannot be excluded because the eyeball was not permitted to be examined postmortem. The ocular symptoms and signs, nevertheless, could be associated with inflammation affecting the optic and oculomotor nerves.

The uveomeningoencephalitic (Vogt-Koyanagi-Harada) syndrome must also be ruled out. This syndrome is characterized by exudative iridocyclitis and choroiditis associated with patchy depigmentation of the skin and hair including lashes and eyebrows. There may be also retinal detachment and deafness (Pattison, 1965; Riehl and Andrews, 1966). Some authors considered this syndrome to be a variant of Bechet's syndrome (Rubinstein and Urich, 1963). The current patient did not have depigmentation of the skin and hair. It is, hence, less likely that she had uveomeningoencephalitic syndrome.

The question might be raised regarding the correlation between the age of the CNS lesions and the approximate 1-year clinical course of the patient. The author is well aware that individual CNS lesions of this patient could have different ages. Some may be old and others may be young. Nevertheless, the occurrence of status spongiosus, glial nodules, and gliosis in the posterior column of the spinal cord and medulla oblongata suggests lesions in these two regions to be old enough and in accordance with 1-year clinical course of the patient's ailment.

The cause of the present malady of the CNS is considered as obscure. Based on the nonspecific and nonpurulent nature as well as occurrence of glial nodules, however, viral etiology is suggested. The author is well aware that this suggestion is not in accordance with the CSF findings in which pleocytosis was absent. However, there were only two CSF studies. The findings, thus, may be less reliable. No further discussion will be made on the viral nature because of lacking of data on the virologic study.

ACKNOWLEDGEMENTS

This study is supported by the Chulalongkorn Faculty of Medicine - China Medical Board Scholar Development Fund, 1994-1995.

REFFERENCES

Chajek T, Fainaru M. Behcet's disease : report of 41 cases and a review of the literature. *Medicine* 1975; 54 : 179-96.

Hegedüs K, Nemeth G. The adult form of subacute necrotizing encephalopathy. *Surg Neurol* 1981; 21 : 572-6.

Kalimo H, Lundbery PO, Oloson Y. Familial subacute necrotizing encephalomyelopathy of the adult form (adult Leigh syndrome). *Ann Neurol* 1979; 6 : 200-6.

Matthews WB. Symptoms and signs. In: Matthews WB, ed. McAlpine's Multiple Sclerosis. Edinburgh : Churchill Livingstone, 1985 : 96-119.

Pattison E. Uveomeningoencephalitic syndrome (Vogt-Koyanagi-Harada). *Arch Neurol* 1965; 12 : 197-205.

Riehl J-L, Andrews JM. The uveomeningoencephalitic syndrome. *Neurology* 1966; 16 : 603-9.

Rubinstein LJ, Urich H. Meningo-encephalitis of Behcet's disease: case report with pathological findings. *Brain* 1963; 86 : 151-60.

Shuangshoti S, Kasantikul V, Lerdlum S. Subacute necrotizing encephalomyelopathy (Leigh's disease) in adult clinically diagnosed as multiple sclerosis. *Chula Med J* 1994; 38 : 145-63.

Shuangshoti S, Tangchai P. Neuropathology. 2nd ed. Bangkok : Thai Watana Panich Press, 1983 : 239-43.

Weller RO. Pathology of multiple sclerosis. In : Matthews WR, ed. McAlpine's Multiple Sclerosis. Edinburgh : Churchill Livingstone, 1985 : 301-45.

Wolf SM, Schotland DL, Phillips LL. Involvement of nervous system in Behcet's syndrome. *Arch Neurol* 1965; 12 : 315-25.

CASE REPORT

CAPILLARIASIS WITH CHRONIC INTESTINAL PSEUDO-OBSTRUCTION

Polrat Wilairatana

Department of Clinical Tropical Medicine and Hospital for Tropical Diseases, Faculty of Tropical Medicine, Mahidol University, Bangkok 10400, Thailand

The common symptoms of capillariasis are diarrhea, recurrent vague abdominal pain, weight loss, malaise, and anorexia. However, there has been no report of capillariasis presenting with chronic intestinal pseudo-obstruction.

A 35-year-old Thai man came to the hospital with history of chronic voluminous, watery diarrhea with 8 to 10 stools daily for 4 months, loss of appetite, and loss of 6 kg body weight. The patient was a farmer working in Prachinburi Province in the northeast of Thailand. The patient suffered from recurrent vague abdominal discomfort with progession of the symptoms prior to admission. Physical examination revealed body weight of 43 kg, muscle wasting, pale conjunctiva, and abdominal distention witth prominent borborygmi. Minimal ascites was detected. Laboratory investigation showed Hb 8 g/dl, WBC 6,500/mm^3, neutrophils 41%, lymphocytes 48%, eosinophils 7%, monocytes 3%, basophils 1%, and adequate platelet count. Blood chemistry showed Na 130 mEq/l, K 2.5 mEq/l, total Ca 7.7 mg/dl, fasting plasma glucose 70 mg/dl, creatinine 0.95 mg/dl. Liver function test showed total bilirubin 0.74 mg/dl, SGOT 21 U/l, SGPT 22 U/l, albumin 1.8 mg/dl, globulin 2.3 mg/dl, prothromin time 15.4 seconds (control 12.4 seconds). Anti-HIV was negative. Ascites fluid showed transudative profile. Stool examination revealed *Capillaria philippinensis* ova. Plain abdomen film demonstrated multiple air-fluid levels in small

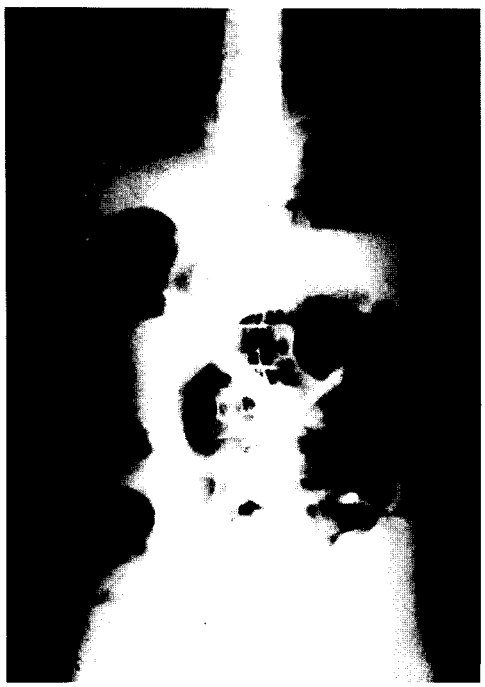

Fig 1–An upright abdominal film on admission showed air-fluid levels in small intestine.

Fig 2–Barium study on prone position showed malabsorption pattern of small intestine.