Clinical Observation on Our Patients and Epidemiology in Osaka, Nara and Wakayama Prefecture

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Recently several patients were reported in Japan, who had been noticed diarrhea or other gastrointestinal symptoms in their prodromal stage and then followed by the signs resembling myelopathy or encephalomyelopathy. It remains still unknown whether they might be any of acute myelitis, neuromyelitis optica, multiple sclerosis, funicular myelosis and myeloradiculoneuritis in old criteria, or not.

Since 1959, 22 cases of such disorder have been experienced also in our clinic. Clinicals and epidemiology of these instances will be presented here, including the same disorder observed in Osaka, Nara and Wakayama Prefecture known by enquête.

I. Observations on nonspecific encephalomyelopathy in our clinic

1) Method and clinical classification (Table 1)

There were 3 patients of nonspecific encephalomyelopathy among 2,686 in-patients during 1949 to 1958, while 26 among 2,760 during 1959 to 1963. Of these 26 cases, 15 were noticed diarrhea, including 3 of mucobloody stools, and 7 complaining of abdominal pain, fullness in abdomen, nausea, meteorism and constipation in the prodromal stage. These 26 cases could be classified into following groups: i.e. 17 of transverse myelitis, 2 of disseminated encephalomyelitis, 5 of myelitis with optic neuritis and others. Transverse myelitis was most frequent regardless of the diarrhea and abdominal symptoms, and myelitis with optic neuritis was more common in the group with diarrhea.

2) Clinicals of 22 patients with diarrhea or other gastrointestinal symptoms.

There were found each one case in 1959 and 1960, 3 in 1961, 7 in 1962 and 10 in 1963, or they were increasing year by year. There were 10 males and 12 females. Eight cases were of sixth decade, 5 of third, 4 of fourth, 3 of fifth and 2 of seventh. Six were found in spring, 7 in summer, 4 in autumn and 5 in winter. Signs of myelopathy appeared acutely or subacutely after several days (4 cases), 3 to 7 weeks (8), 2 to 4 months (8), or 5 to 6 months (2) from the onset of abdominal symptoms. Fever elevated over 38.0°C in 2 cases. Muscle pain in both legs was seen in 10 cases.
Table 1. Classification of the Nonspecific Encephalomyelitis

<table>
<thead>
<tr>
<th>Classification</th>
<th>Number of cases in our clinic</th>
<th>Number of cases by enquête</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>with diarrhea</td>
<td>with abdominal symptoms except diarrhea</td>
</tr>
<tr>
<td>Transverse myelitis</td>
<td>9</td>
<td>4</td>
</tr>
<tr>
<td>Disseminated encephalomyelitis</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Myelitis with optic neuritis</td>
<td>4</td>
<td>1</td>
</tr>
<tr>
<td>Multiple sclerosis syndrome</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Encephalomyelitis</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Meningomyelitis</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Landry type</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Miscellaneous</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Total</td>
<td>15</td>
<td>7</td>
</tr>
</tbody>
</table>

Remarks: ( ) shows the number of cases during 1949~1958
Sensational disturbance below the navel or the legs was found in all cases, but it was often imperfect and more commonly superficial than deep. Vesico-rectal disturbance was seen in 7 cases, perfect paralysis of the lower extremities in 5, of whom 1 was followed by imperfect paralysis of the upper extremities, and imperfect paralysis of the lower extremities in 17. Knee jerks were weak or absent in some cases at the first stage, but became elevated later in 20 cases, and Achilles tendon reflexes were elevated in 15. Babinski reflex was positive in 16 cases and patellar or ankle clonus was positive in 5. Deep reflexes in the upper extremities were elevated in 9 cases. Positive Hoffmann or Trömner reflex was seen in 10 cases.

All patients were treated with steroids, antibiotics and vitamins, of which large amount of thiamine was most commonly used, but they were of little effect. γ-Globulin was administered noncontributorily in only one case.

As to the outcome, death occurred in 2 cases, including one of coexisting cirrhosis of the liver, blindness or high grade visual disturbance with disturbed gait in 3, disability of standing in 2, moderately disturbed gait in 3, slightly disturbed gait in 7, and nearly perfect recovery in 5.

According to our experiences, they can be separated into 2 groups: the one is accompanied by retrobulbar neuritis and the other without it. Most of the former were more serious than the latter. Six of our 22 patients seemed to belong to the former and 16 to the latter.

Typical clinical course of each one case will be presented here.

![Fig. 1.](image-url)
1) Report of a case with retrobulbar neuritis (Fig. 1)

A 24 year-old clerk, having often noticed diarrhea for one year, had complained of diarrhea 3-4 times per day with occasional abdominal pain for previous several weeks. Visiting our clinic on July 23, 1961, he was treated as enteritis with gradual improvement. Then, numbness in the both legs was suddenly noticed on Aug. 6, and increased in degree until the end of Aug. He was admitted on Sept. 24, complaining of disturbed gait with spastic paralysis of the lower extremities. On admission, knee and Achilles tendon reflexes were elevated, with positive Babinski reflex, but normal were the reflexes of the upper extremities and abdomen. Hypoesthesia was found below the navel and deep sensation was slightly disturbed. Steroids, vitamins and ATP were prescribed with little effect. Numbness was so stubborn that sleep was often disturbed. At the end of September, he noticed visual disturbance, which was getting worse gradually. Ophthalmologic examination revealed retrobulbar neuritis optica. At the beginning of October, temporary recovery of gait with negative Babinski was seen, but at the end of that month gait grew again worse, thereas numbness of the hands, elevated reflexes of the upper extremities and positive Trömmer and Hoffmann reflex appeared. Finally, numbness of the extremities was gradually improved with a little variety, and he became to be able to walk on crutches, though visual power got worse progressively until he lost it perfectly.

2) Report of a case without optic neuritis (Fig. 2)

A 44 year-old maid noticed, on May 27, 1963, calls of nature over 20 times per
day, with lower abdominal pain and absent or scanty passage of stools. In spite of our treatment as acute enteritis, abdominal discomfort, nausea mushy stools had continued for 2 weeks. She was admitted on June 14, complaining of general weariness, powerlessness and numbness in the legs, pricking pain in the both thighs and disturbed gait. On admission she was constipated, restless, irritable and sleepless owing to her complaints. Though abdominal reflexes were absent, reflexes of the both upper extremities were elevated. Hoffmann and Trömner reflexes were positive in left side. Somewhat weak knee jerks, nearly absent Achilles tendon reflexes and positive Babinski reflex were found in both sides. Hypesthesia was observed below the navel, but almost normal was the deep sensation. Gait was somewhat spastic. Test of the cerebrospinal fluid was normal. She was treated with steroids and vitamins. Numbness of the legs diminished after 2 weeks, though knee jerks remained markedly elevated. At the end of July gait was improved with normal knee jerks and negative Babinski, and it became nearly normal at the beginning of August. She was discharged with but a little hypesthesia below the navel on Sept. 10, 1963.

II. Observations on nonspecific encephalomyelopathy in Osaka, Nara and Wakayama Prefecture

1) Method and clinical classification of the cases by enquête (Table 1)
   Primary enquête was sent to total 75 clinics, 48 in Osaka, 6 in Nara and 21 in Wakayama Prefecture. Answers were obtained from 46 clinics. By secondary more detailed enquête to 29 clinics, which had had the experiences of such disorder, 60 cases of encephalomyelopathy including 33 with diarrhea and 4 with abdominal symptoms except diarrhea and 23 without them were collected. They could be classified into following groups, i.e. 39 cases of transverse myelitis, 3 of disseminated encephalomyelitis, 11 of myelitis with optic neuritis, 2 of multiple sclerosis syndrome and one of another figure. There was little relationship between the types of the clinical figure of encephalomyelopathy and diarrhea or abdominal symptoms, although transverse myelitis was most commonly experienced, regardless of the presentation of diarrhea and abdominal symptoms.

2) Epidemiology
   Epidemiology was studied on total 89 cases, of which 29 were ours and 60 were obtained by enquête.
   Sex and age: Patients were ranged in third to sixth decades. Males were dominant in the group without diarrhea and abdominal symptoms, but as frequent as females in the group with them.
   Season: Among 48 cases with diarrhea, 16 were discovered in May to June, 15 in July to September, or they occurred mostly in early summer to early autumn. Patients with abdominal symptoms except diarrhea were scattered equally in every month, and patients without diarrhea and abdominal symptoms had little relation to the season.
Yearly frequency: Though enquête was done as to the patients experienced during January, 1960 to June, 1963, 2 cases with diarrhea in 1959 and 2 without diarrhea and abdominal symptoms in 1956 were included here. Three cases of ours seen before 1958 without diarrhea and abdominal symptoms were also included. Cases with diarrhea seemed to be increasing year by year, while cases without it had no definite tendency.

Distribution: Of 59 cases with diarrhea and abdominal symptoms, 31 were found in Osaka Prefecture, 20 in Wakayama and 1 in Nara.

Summary

For recent several years, marked increase of nonspecific encephalomyelopathy has been experienced in our clinic, and most of them are accompanied by prodromal diarrhea or other abdominal symptoms. Their clinical manifestations, however, have little difference from the nonspecific encephalomyelopathy in old criteria. Since death, blindness and disability of standing follow them not uncommonly, their prognosis can not be always benign. It seems to be, at present, still too early to say that nonspecific encephalomyelopathy with diarrhea or other abdominal symptoms might be a definite clinical entity.