

## Short report

# Cerebellar ataxia in enteric fever

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**SUMMARY** In a study of enteric fever, cerebellar ataxia was found to be the commonest neurological manifestation, second only to toxic delirium. Excluding toxic delirium (found in 25-30% of cases) neurologic deficit was noted in 5.0% of a series of 718 consecutive cases; 2.3% showed cerebellar ataxia, either as an isolated feature or in association with other lesions. The ataxia usually appeared in the second week, and lasted a mean of 9.4 days. In 90% of cases it had cleared completely within a month.

Neurological manifestations occur in enteric fever. Besides toxic delirium which may be seen in 22-57% of cases<sup>1-4</sup> the two most frequently mentioned are encephalitis or encephalopathy<sup>3-7</sup> and post-typhoid psychosis.<sup>6</sup> The encephalopathy may take various forms from deep unconsciousness and convulsions to focal deficits such as aphasia and hemiplegia.<sup>1,2,4,6</sup> Perhaps the largest collection of cases is that of Osuntokun *et al*<sup>2</sup> who reported on 959 patients and found toxic delirium in 57%, varying depth of coma in 3.5%, bilateral pyramidal signs in 3.1%, transient extrapyramidal signs in 1% and rare cases of peripheral neuropathy, mononeuritis multiplex and the late development of schizophreniform psychosis. The authors did not mention any case with cerebellar ataxia. We have noted a cerebellar syndrome in a considerable number of cases of enteric fever. In fact, cerebellar ataxia was the commonest manifestation (if delirium is excluded).

### Materials and methods

The paper deals with 28 cases who developed cerebellar ataxia during the course of enteric fever, derived from two sources. The first was a consecutive series of 718 cases from Sassoon Hospital, seen between March 1978 and February 1981. Of these, 36 (5%) showed neurological manifestations (excluding toxic delirium) and 16 (2.3%) showed cerebellar ataxia. To these are added 12 further cases, seen either subsequent to February 1981 or at an

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Received 7 June 1983 and in final revised form 23 November 1984.  
Accepted 15 December 1984

attached institution by one of the authors (RSW).

The criteria for the diagnosis of enteric fever, in these 28 cases, were (a) Culture and isolation of the organism from blood, stool or marrow in 13 cases. (b) A Widal titre rising to above 1 in 250 in the remainder. In nine cases there was a fourfold rise in titre. In the rest the initial titre itself was very high (for example 1 in 300) and a fourfold rise was not obtained (for example 1 in 300 rising to 1 in 900). According to standard texts an initial Widal titre of the 0 antigen of 1 in 100 with a subsequent rise is proof of enteric fever.<sup>8</sup> Others feel that a diagnosis based on the Widal test alone is less reliable.<sup>9</sup> For this reason we chose deliberately stringent criteria for the Widal titre. The 13 culture-positive cases and fifteen culture-negative cases were similar in all respects, including clinical manifestations, course and response to treatment. They will therefore be discussed together.

### Clinical findings

The cerebellar ataxia was seen either in isolation or in combination with a variety of other signs which have been described before (table). We have restricted the use of the term encephalopathy to patients showing unconsciousness lasting over 48 hours (excluding patients who only showed the usual toxic delirium). In this period there were eight cases of encephalopathy, of whom two developed cerebellar ataxia after recovery from unconsciousness. A protracted confusional state was one which persisted long after fever subsided. It was noted to last up to 90 days (mean 23.2 days). The table lists the clinical manifestations of the cerebellar syndrome. All cases showed marked ataxia of gait. If a patient showed only mild ataxia, during tandem gait testing, this was ignored as possibly being due to prolonged illness. Twenty two of the 28 cases were totally unable to walk unaided, and seven were unable to stand unaided because of ataxia. The commonest finding was a combination of gait ataxia and limb ataxia generally more

Table Cerebellar ataxia in enteric fever. (28 cases)

Types of syndrome	No of cases
Isolated cerebellar ataxia	10
Cerebellar ataxia + protracted confusional state	8
Cerebellar ataxia + Bilateral pyramidal signs	3
Cerebellar ataxia + deafness	2
Initial encephalopathy followed by cerebellar ataxia	2
Cerebellar ataxia + Extrapyramidal signs	1
Cerebellar ataxia + confusion + Bilateral pyramidal signs	1
Ataxia + confusion + pyramidal signs + extrapyramidal signs	1
<i>Neurological signs</i>	
Gait ataxia	28
Bilateral finger-nose ataxia	24
Unable to walk unaided	22
Bilateral heel-knee ataxia	22
Dysidiadochokinesia	14
Hypotonia	12
Rebound phenomenon	11
Unable to stand alone	7
Pendular knee jerk	6
Cerebellar speech	5
Nystagmus	4

marked in the lower limbs. As many cases had gait ataxia, and as limb ataxia affected upper and lower limbs the figures in the table exceed 28. Nystagmus and dysarthric speech was noted in only a few cases.

Most cases of enteric fever who were to develop neurologic signs had a toxic delirium in the earlier stages (87%). However, three of the 28 cases came to hospital for the neurologic deficit alone. Two of these had isolated cerebellar ataxia. In these cases the fever had been treated at home and had not apparently been severe or associated with delirium. The cerebellar ataxia appeared in the first week of fever in seven (25% of the ataxia cases), in the second week in 17 (61%) and in the third week in four (14%). Twelve cases were actually apyrexial when ataxia was first noted. The mean time of onset was 10.4 days, and the mean duration of significant ataxia 9.4 days. At the end of one month only three patients still showed ataxia, and none showed ataxia by the end of 6 weeks.

During the course of our study only three cases of enteric fever with neurologic manifestations died. One had cerebellar ataxia. Two were studied post mortem. The only abnormality noted was mild oedema and congestion of the brain. There was no evidence of perivascular cuffing and no perivenous demyelination.

## Discussion

Cerebellar ataxia as a complication of enteric fever is not mentioned in standard texts.<sup>6,10</sup> Osuntokun *et al*<sup>2</sup> in a review of 959 cases do not mention cerebellar ataxia, yet it was the commonest manifestation we encountered.

Probably the first description of cerebellar ataxia in enteric fever was by Westphal in 1872.<sup>11</sup> Joshi<sup>7</sup> reviewing 135 cases of typhoid fever described 15 cases of encephalitis and listed four as having ataxia.

Since then a few isolated case reports have appeared in the Indian literature.<sup>12,13</sup> Ukadgaonkar *et al*<sup>12</sup> described three cases. All had "enteric toxæmia" to start with and the ataxia lasted 10–14 days. A number of cases have also been described in the French literature, chiefly from Africa.<sup>14,15</sup> Perhaps the largest series in the English literature to date is that of Scragg *et al*<sup>4</sup> who reviewed the complications in 340 cases of enteric fever and found eight with ataxia giving a prevalence of 2.5%. In Joshi's series<sup>7</sup> the prevalence of cerebellar ataxia was 3.5%. Joshi does not describe the ataxia further. Scragg *et al*<sup>4</sup> only noted that in all but one of the cases the ataxia was "cerebellar in type".

The explanation of the neurologic manifestations is not clear. Osuntokun *et al*<sup>2</sup> found no abnormality in the cases studied at necropsy. In our two cases examined at necropsy, only cerebral oedema and congestion were seen. Neither of these two cases had cerebellar ataxia. One case of Scragg *et al*<sup>4</sup> showed a large cerebellar haemorrhage but we doubt if this is the cause of the transient cerebellar ataxia described here. Ramchandran *et al*<sup>16</sup> have described perivascular cuffing and perivenous demyelination in a single case. The case did not have cerebellar ataxia and the changes were noted only in the midbrain and pons. Other authors also have seen nonspecific oedema and vascular changes as their only findings in fatal cases.<sup>17</sup>

In view of the paucity of pathological data implicating the cerebellum and the frequency with which the ataxia followed toxic delirium, could it be that the signs described were manifestations of frontal ataxia? The low prevalence of dysarthria and nystagmus lend some support to such a view. Though it is accepted that at times it is not possible to differentiate frontal from cerebellar ataxia we feel the syndrome we describe to be of cerebellar origin for the following reasons: (a) Several cases showed isolated ataxia without confusion or any change in mood, memory or attention when the ataxia was evident and gross, (b) There was nearly always marked finger-nose ataxia (83%) and heel-knee ataxia (77%) or both (71%). Intention tremor was frequent. These signs are unusual in frontal lobe ataxia. (c) The gait was thought to be typically cerebellar with swaying from side to side and irregularity and variability of steps quite unlike the slow, short, shuffling gait typical of frontal lobe ataxia,<sup>10</sup> (d) In all cases with isolated ataxia, hypotonia was present. This was of course not seen when there were associated pyramidal or extrapyramidal signs. It was less noticeable in those with confusion. Frontal lobe ataxia is usually associated with increased resistance to movement.<sup>18</sup>

The cerebellar ataxia seen in alcoholic cerebellar

degeneration<sup>19</sup> is clinically very similar in that it involves stance and gait chiefly and affects lower limbs more than upper. In that syndrome too, nystagmus and dysarthria are quite uncommon. In these alcoholic cases Victor *et al*<sup>19</sup> showed the lesions to involve the cerebellar cortex and to be most marked in the anterior and superior region of the vermis and cerebellar hemispheres.

Thus it is our belief that the isolated ataxia seen in some of our cases was of cerebellar origin. In other cases (those with associated signs) other factors may have played a part but the ataxia was in most respects similar to the isolated ataxia group. Final verification of the pathogenesis of this ataxia is not possible in this reversible disorder. All our survivors recovered completely, and no residual ataxia was seen after 6 weeks. Nevertheless the point is worth making that in Poona, India (and probably elsewhere on this subcontinent) the commonest cause of acute or subacute cerebellar ataxia occurring after fever, implies that the fever was enteric fever. We have in fact since the start of this study, diagnosed a fever as being enteric because of the appearance during its course of a bilateral cerebellar ataxia.

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