

not been reported before.

5. *Normal MRI of the spine:* MRI abnormalities are almost universal. In an Indian study, focal cord atrophy was seen in 100% of cases at the level of cervical 4 to 7 vertebral bodies in MRI done in neutral neck position(4).

Secondly, I disagree with the inclusion of Madras motor neuron disease (MMND) as a differential diagnosis in this case. MMND typically presents with the involvement of lower cranial nerves (seventh and ninth to twelfth) and sensorineural deafness along with features of chronic anterior horn cell disease(5,6).

The diagnosis in this child could still be a genetically determined distal spinal muscular atrophy. Genetic and molecular studies to identify survival motor neuron (SMN) 1 and 2 gene mutations would have been useful towards making a definite diagnosis(7).

Finally, the association of elevated serum lead level in this child appears to be coincidental. However, chelating therapy is required for the same.

In conclusion, the diagnosis of Hirayama disease cannot be made confidently in this case. Genetic studies would have been useful in making a definite diagnosis of spinal muscular atrophy.

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Reply

Our comments are as under:

- None of the previous authors have used the criteria of 'four to five years of symptoms' before making a diagnosis of Hirayama disease(1). Even in the study quoted by Dr. S. Kumar(2) the duration of illness at presentation ranged from 3- 64 months with 25% patients having duration of illness less than 12 months.
Given the reported arrest of disease progression with the use of a cervical collar(2), it is imperative that the diagnosis is made as early as possible rather than waiting for a period of "at least 4-5yrs. of symptoms".
- The patient reported had asymmetric

bilateral involvement with more severe wasting on the right side. The 'oblique atrophy' referred to is due to the wasting of the forearm and hand muscles with sparing of the brachioradialis.

3. We agree that the disease is uncommon in females. However, female sex alone does not exclude the diagnosis.
4. The disease usually presents in teens or early twenties and the earliest reported age at onset is 10 years, as mentioned in the text. The purpose of the case report itself was to comment on the early age of presentation and to speculate on the possible role of lead in accelerating the rate of disease progression (and thereby an early onset).
5. Hirayama, *et al.* studied 73 consecutive patients with the disease by myelography, CT-myelography and MRI(3). In neutral neck position, MRI could detect mild to moderate atrophy of lower cervical cord in only 49% of the patients (23 of 47).

Genetic analysis of patients with distal upper limb spinal muscular atrophy has been previously carried out(4). In four patients of this disease which were studied, no homozygous deletions of exon 7 and 8 of the SMNtel gene were found, and no deletions in exon 5 of the NAIP gene were detected. The diagnosis is essentially clinical supplemented

by results of electrophysiological investigations and muscle biopsy. Even though the parents were unwilling to go for further tests, we think that genetic studies would not have been of any help in the diagnosis.

We have discussed the possibility of elevated lead being a coincidental finding. Chelation therapy was offered but refused by the parents.

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Resuscitation of Asphyxiated Newborns(1)

We read with interest your article(1). We feel the conclusion should be interpreted with caution due to the following reasons:

Methodology

1. Use of date-based method of randomization is not a strict randomization procedure. This issue should not be brushed aside by authors as 'lacunae for purist'. If a proper randomized control trial would have been done, the results could have been different. Similarity of results of