Progressive Muscular Dystrophy
A New Clinical Sign
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Abstract
Meryon gave the first description of progressive muscular paralysis in 1852. Later, Duchenne gave complete description of progressive muscular dystrophy (PMD) in 1868, now disorder has been given his name. Gowers provided the first comprehensive details in English in 1879. Both Duchenne and Gowers emphasized the pseudohypertrophic enlargement of certain muscles.

P.M.D. is a primary progressive degeneration of the musculature of the body of unknown etiology. P.M.D. is the commonest muscular dystrophy having x-linked recessive inheritance and usually affects males. In the early stage or in doubtful cases, accurate diagnosis should be made after proper clinical examination and investigations like estimation of creatine kinase (CPK), serum aldolase, electromyography (EMG) and histopathological examination of muscle.

Incidence
The incidence of Duchenne muscular dystrophy ranges from 13 to 33 per 100,000 live born males and its prevalence in population varies from 19 to 39 per 100,000.

Symptoms
Usually parents describe clumsiness in walking and frequent falls in the affected child. In many cases, walking is delayed. Whereas in other cases, child is apparently normal until third year and parents do not report any abnormality. Later child shows laziness or pesplanus and inability to run, followed by increasing difficulty in climbing stairs and also in raising from floor by climbing upto legs, to attain standing position (Gowers sign). Later child walks with waddling gait.

A new clinical sign has been observed in these cases due to atrophy of Brachioradialis muscle.

The wasting is seen in musculature of proximal part of upper and lower limbs alongwith muscular hypertrophy is present in calves. Weakness of hip and knee extensors is observed followed by weakness around shoulder girdle. Contracture develops usually in tendoachillus & flexors of hip and knee.

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Technique
The upper limb is kept on the side of the chest and elbow at 90°, along with forearm in mid-prone and then the child is asked to flex the elbow against the resistance on the forearm. Normally there is bulge or prominence of belly of a brachioradialis.

A positive sign means that there is no bulge or prominence of belly of brachioradialis.
A negative sign means that there is bulge or prominence of belly of brachioradialis (photograph 2).

The weakly positive sign means that there is mild bulging of belly of brachioradialis.

Discussion

This new clinical sign (Agarwal’s sign) has been observed practically in all cases of already proved PMD who have attended O.P.D. of Department of Physical Medicine and Rehabilitation, RALC, K.G. Medical College, Lucknow from August 1997 till April 2002. During this period a total of 36 cases of PMD have been documented wherein this new clinical sign was observed.

This new clinical sign (Agarwal’s sign) is simple, easily detectable and requires no tools or special training. Further elicitation of this new sign requires less exposure of the upper limb and hence suitable for young girls as well. It has been observed in cases of Myopathy where there is minimum hypertrophy of calves, the new clinical sign was positive.

The positive sign denotes complete atrophy of brachioradialis whereas negative sign denotes normal bulge of brachioradialis.

References

1. Duchenne G.B. (1868), Recherché sur la paralysio musculaire pseudo-hypertrophique au paralysie myoscl erosique, Archives Generatis de Medicine, II, 5,178,305,421,552