LETTERS TO THE EDITOR

THEVENARD SYNDROME

This has reference to the case report entitled "Thevenard syndrome" published in Ind J Dermatol Venereol Leprol, 1987; 53 : 305-307. I have the following comments to offer :

Thevenard syndrome is a rare autosomal dominant disorder due to hereditary degeneration of cranio-spinal ganglia. The sensory neurons that connect centrally with the spinothalamic tract are particularly affected. These are often associated with nerve deafness.[1]

The authors in their case report have reported six cases of this rare syndrome. None of these cases had family history or nerve deafness. Four of these cases had spina bifida. I feel these four cases are simple spina bifida with trophic changes. The incidence of lumbosacral spina bifida occulta has been reported to be as high as 17% of all the spines X-rayed.[2] In lumbo-sacral spina bifida, the spinal cord often retains its foetal length and extends down to the sacrum. The variety of lesions including diastematomyelia, intramedullary dermoid, hydromyelia, cauda equina compression due to the fibrous band or adhesions, ectopic dorsal nerve roots and lipoma have been reported in association of spina bifida occulta.[3] Muscle weakness and wasting below the knees, impaired or loss of ankle jerk and trophic ulceration[4] may be the presenting feature in some cases.

In view of the above, I do not agree with the authors for clubbing all these cases to a group of rare syndromes, specially in these four cases with spina bifida, a compression on or tractional damage of nerve roots associated with this defect led to trophic ulceration and trophic bone changes (Sudeck atrophy).

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REPLY

We appreciate the critical reading of our article by Dr Talwar. However, in this connection, we state that our cases were of sporadic types which are quite frequent than those of familial types.[1] As regards nerve deafness, it develops in later life and our cases ranged from 3 to 13 years. Dissociated sensory deficit and osteolysis of bones are the cardinal association of Thevenard syndrome. Those patients with suspected spina bifida were investigated by the neurologist and pathology like dermoid, diastometomylia, tethered cord etc within spinal canal was ruled out. Presence of ulcero-multilatory acropathy in our cases without any intraspinal disease prompted us to label these cases as of Thevenard syndrome.

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References

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