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THEVENARD SYNDROME

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Six cases of Thevenard syndrome presented as trophic ulcers on the toes with shortening and loss of pain and temperature sensations. Four cases had spina-bifida out of which two cases had associated faun-tail and one case had hypertrophy of the lower extremity of one side.

Key words : Thevenard syndrome, Trophic ulcers, Spina-bifida, Faun-tail.

Thevenard syndrome is an unusual familial disorder transmitted as an autosomal dominant trait. Trophic ulcers on the toes and heels are its major clinical manifestations. These are largely located on the plantar surface of the great toe, and are associated with the loss of pain and temperature sensations. Osteolysis, osteoporosis and spontaneous amputation of digits are characteristically seen later in the course of the disease.[1] Deafness, spina-bifida, nystagmus, muscular atrophy and shooting pains may also be its neurological accompaniments.[2-5] Occurrence of the disease is infrequent in children, for only a few reports are thus far available.[6,7] We report six cases with interesting clinical manifestations.

Case Reports

Case 1

A 5-year-old male had non-healing, painless ulcers over the plantar aspect of the great and little toes of the right foot noticed when the child was 6-month-old. The ulcer over the great toe was 2 cm × 1.5 cm, with a well-defined margin, dirty looking base and surrounded by hyperkeratotic collar, while that of little toe was crusted, progressed slowly to ultimately result in shortening of all the toes. There was no history of deafness, muscular weakness, shooting pains or eye complaints. There was no history of a similar condition in the family.(Fig.1)

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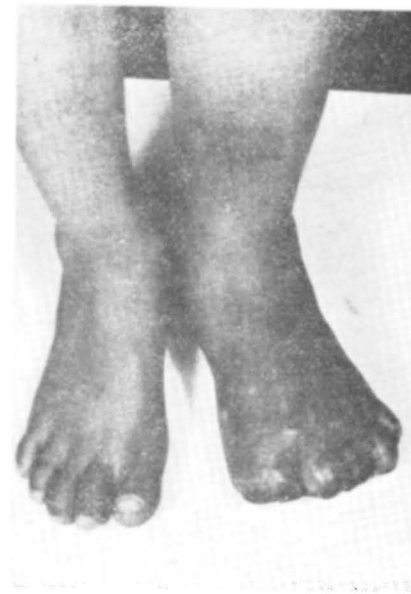

Fig. 1. Shortening of all the toes of right foot with ulcers over the plantar aspects of the great and the little toes.

The sensations of pain and temperature were absent in the right foot upto its middle with a convex border towards the ankle joint. There was a tuft of hair (Faun-tail) in the lumbo-sacral region. A similar tuft of hair was also seen over the antero-medial aspect of the right leg. Muscles of the right leg were hypertrophic, the deep tendon reflexes however, were normal. Radiological examination of the lumbo-sacral spine revealed spina-bifida. Osteoporosis and partial destruction of phalanges of all the toes was seen in the right foot. Hypertrophic changes of the right tibia and fibula were also observed.

The other 5 cases ranging in age from 3 to 13 years also had non-healing, painless ulcer's over the plantar aspects of the right great toe (cases 2 and 3), 4th and 5th toes of left foot (case 4), right 1st toe (case 5), and 2nd and 3rd toes of left foot (case 6) for 6 months to 2 years. Corresponding shortening of the toes was also noted in these cases. Loss of pain and temperature sensations was observed around the ulcer in all the cases. However, deep reflexes were normal. Only 1 case (case 4) showed faun-tail on the lumbo-sacral area. The family history was not positive in any case. X-ray lumbo-sacral spine revealed spina-bifida in cases 3, 4 and 5. X-ray of the foot showed osteoporosis and partial destruction of the distal phalanges of the corresponding shortened toes.

Comments

Thevenard syndrome envisages occurrence of an ulcerative multilatory acropathy characterised by severe and relapsing ulcers of the skin of the feet and hands. Dissociated sensory deficit and osteolysis of bones are its cardinal associations. Nelaton[8] described a family with such features in 1852 and ever since, the disease has been variously described by many workers. Hicks[2] described the disease in ten members in four generations of a family. Denny-Brown[9] subsequently observed that the primary cause of ulceration of the feet was due to degeneration of the cells of dorsal root ganglia, dorsal roots and peripheral nerve trunks. He coined the term Hereditary Sensory Radicular Neuropathy and documented three sporadic cases, and emphasized that such a situation is frequent than those of their occurrence in the family. However, the symptomatology was not as typical or striking.

Thevenard[10] and many other workers[2,3,5] observed that the sensory loss is of dissociated type, affecting pain and temperature sensations only, while in some cases, area of this disturbed sensations being limited to a circular area around the ulcer with no suggestion of segmental distribution. Interestingly, we had similar findings in our cases, reiterating their findings. Spina-bifida was observed in four cases out of which two had associated Faun-tail in our series. Similar findings have been reported by earlier workers.[2,3,5] Other associations of the disease like loss of tendon reflexes, shooting pains, muscular atrophy, deafness, nystagmus and choreiform movements as observed by various workers[2-5] were not noticed in our cases. However, case 1 presented with hypertrophy of the muscles, subcutaneous tissues and bones of the right lower extremity.

Thevenard syndrome is inherited by a dominant gene with reduced penetrance and variable expressivity. Though the disease is relatively unusual in children, Heller and Robb[6] reported two cases 9 and 12 years of age, who suffered from the disease but without any lightening pains or deafness.

Walker[7] described both a sporadic type and a familial type of the disease in two children with both of them having dissociated sensory deficit, lightening pains and deafness. However, only the child with the sporadic type exhibited the characteristic ulcero-mutilatory acropathy, as seen in our cases.

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