PSEUDO-AINHUM ASSOCIATED WITH SPINA-BIFIDA OCCULTA AND HEMIVERTEBRA

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A 7-year-old girl had pseudo-ainhum, spina-bifida occulta, a hemivertebra and focal neurological deficit. The disease process was progressive for the last three years leading to amputation of the third digit and formation of constricting bands around the fourth and the fifth digits of the left foot.

Key words : Pseudo-ainhum, Spina-bifida occulta, Hemivertebra.

Constricting bands on the digits can be classified into true-ainhum and pseudo-ainhum.[1] True-ainhum is characterized by the development of a constricting band around a digit, which may be followed by spontaneous amputation, whereas pseudo-ainhum is a sequence of similar events occurring in various other disorders or as an isolated developmental anomaly.

Spina-bifida results from incomplete closure of the spinal canal, the commonest site being the lumbo-sacral region. In spina-bifida occulta, there is radiological evidence of failure of fusion of the posterior arches of the lumbar vertebrae. If it is associated with a cutaneous marker such as a lumbo-sacral lipoma, dermal sinus, hairy patch or an angiomatous malformation, the lower end of the spinal cord is at risk from traction injuries resulting from tethering due to congenital anomalies in the spinal canal. The traction injury to the cord can result in motor, sensory or sphincter disturbances.[2]

A typical vertebra develops from three primary ossification centres; one in each half of the vertebral arch at the roots of the transverse processes and the third in the body (centrum). The body (centrum) is ocassionally ossified from bilateral centres which may fail to unite. If one of the centres fails to develop, a hemivertebra results which causes lateral curvature of the spine (scoliosis).[3]

Pseudo-ainhum has been reported in association with certain disorders, such as, pathologies of the nervous system. The present case had association of focal neurological deficit without evidence of syringomyelia. There are very few reports available in the Indian literature describing pseudo-ainhum.[4]

Case Report

A 7-year-old girl developed annular constricting bands around the third, fourth and fifth digits of the left foot 3 years back. This was followed by auto-amputation of the third digit about 2½ years back. She was born as a full-term baby of non-consanguinous marriage. The developmental milestones were within normal limits but she had nocturnal enuresis. There was no family history of congenital anomalies, syphilis, diabetes mellitus, palmar and plantar hyperkeratosis. She was moderately built and well-nourished with a tuft of hairs on the lumbar region and a mild scoliosis with concavity to the left in the same region. There was absence of the distal two-thirds of the third digit and constricting annular bands were present near the bases of the fourth and fifth digits of the left foot. There was no associated club foot, cleft-lip, cleft-palate, syndactyly or microdactyly. The neurological deficit was confined to the left lower limb i.e. the site of pseudo-ainhum. She

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also had wasting of the left thigh and calf-muscles. The tone was normal, power was grade 5/5 and there were no fasciculations. Fine touch was impaired in L_5 and S_1 dermatomes. Sensations of both spinothalamic and posterior column tracts were normal. The knee-jerk was diminished, the ankle-jerk was absent and the plantars were poorly elicitable but were flexor on the left side. Other limbs revealed no neurological abnormality. Other systems were normal.

Hemoglobin, total and differential leucocyte counts, sedimentation rate, urinalysis and stools examination were normal. Investigations to detect any possible primary cause, such as, diabetes mellitus, syphilis, leprosy, polyneuropathy etc, were non-contributory. Roentgenograms of the left foot and vertebral column revealed complete loss of the distal and middle phalanges, and partial absorption of the proximal phalanx of the third digit, complete loss of the distal phalanx and absorption of the middle phalanx of the fourth digit, incomplete spina-bifida at D_9 level, hemivertebra at L_2 level and complete spina-bifida caudal to L_3 involving even the lowest sacral vertebra.

Comments

True-ainhum (spotaneous dactylolysis) as first described by Messum in 1821 and Clarke in 1860,[5] is a pathological phenomenon in a class of its own, having no aetiological or morphological identity with congenital or acquired conditions commonly referred to as pseudo-ainhum or atypical-ainhum.[6] It eventuates in auto-amputation of the digit as the constricting band deepens and bony absorption occurs.[7] It occurs more frequently in tropics, amongst adult Negroes and involves invariably the fifth toe. It is more common in the males especially in bare-foot walkers. The exact aetiology is yet unknown though a role of infection is postulated.[6,8] Pseudo-ainhum, a term coined possibly by Neumann,[9] now represents a condition in which constricting bands occur for reasons other than ainhum. Its causes can be divided into, (a) congenital bands, (b) traumatic causes, and (c) associated with or secondary to specific diseases, such as leprosy, syphilis, diabetes mellitus, ankylostomiasis, scleroderma, syringomyelia, spinal cord tumors, pachyonychia congenita, hyperkeratosis palmaris et plantaris etc.[1]

Izumi and Arnold[10] reported pseudo-ainhum with club foot and syndactyly, and hypothesized that an underlying mesenchymal developmental defect was responsible for such associations. Similarly, spina-bifida occulta and hemivertebra with pseudo-ainhum in our case can be explained on the basis of this hypothesis as all these defects involve mesenchymal tissues. Alternatively, spina-bifida occulta and hemivertebra causing a neurological deficit, leading to pseudo-ainhum could possibly explain this association in our case.

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