CLINICAL REVIEW

Diagnostic dilemma revealing a rare association of pseudoainhum with underlying vascular anomaly

Banashree Majumdar, Atul Jain, Aggarwal Ishad, Anindya Bandyopadhyay[1], Arghyaprasun Ghosh
Departments of Dermatology, Venerology and Leprosy, and [1]Radiology, IPGMER and SSKM Hospital, Kolkata, West Bengal, India

ABSTRACT

We report a case of pseudoainhum with underlying vascular malformation in a 4½-year-old male patient over distal part of left lower extremity with a constriction involving the fifth toe.

Key words: Constriction, pseudoainhum, vascular malformation

INTRODUCTION

Pseudoainhum is a rare acquired or congenital disorder characterized by progressive development of a fibrotic band on a finger or toe until spontaneous auto amputation occurs. Ainhum is an idiopathic disease involving the fifth toes of black people mainly in tropical zones.[1] Although there are various congenital and acquired causes, pseudoainhum is rarely reported to be associated with underlying vascular anomaly. Hence we are reporting the same.

CASE REPORT

This was a case report of a 4½-year-old Muslim boy arising out of a non-consanguineous marriage presented to us with mildly pigmented thickened skin over distal part of left lower extremity with a constriction involving the fifth toe [Figure 1]. There was no history of any pain or discomfort in the affected limb and the child carried out his normal daily activities. On examination, there was a sharply demarcated scaly thickened moderately pigmented, warm, plaque extending from lower third of left lower limb on to the toes. A constricting band was seen over the base of the fifth toe. Further thick circumscribed scales were seen enclosing the affected 5th toe. Just proximal to the lesion, at the junction of middle and lower third of the affected limb, an increased localized bounding pulsation was felt. There were no sensory alterations in any limbs nor was there any abnormality in sweating. Temperature in other limbs was normal. There was no palmoplantar keratoderma. There was no family history of icthyosis or keratoderma. Routine haematological tests were within normal limits. Histopathological examination of a punch biopsy specimen obtained from the dorsum of affected foot showed compact hyperkeratosis and papillomatosis in a church spire manner with preserved granular cell layer. Along with an irregular acanthosis and broad and pointed rete ridges, the basal cell layer showed exaggerated pigment [Figure 2]. Color Doppler study of the left lower limb showed the normal triphasic pattern of flow being replaced by a monophasic pattern in the anterior and posterior tibial and dorsalis pedis arteries [Figure 3]. Screening for human immunodeficiency virus was non-reactive: X-ray chest and the affected foot were within normal limits. Mantoux test was negative.

DISCUSSION

Pseudoainhum cases which we generally encounter can either be congenital or acquired. In congenital cases the cause is usually unknown but few worth mentioning causes are amniotic bands, adhesions, after amniocentesis and in Ehler-Danlos syndrome. Among the causes

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ADDRESS FOR CORRESPONDENCE
Dr. Banashree Majumdar,
Department of Dermatology, Venerology and Leprosy, IPGMER and SSKM Hospital, Kolkata - 700 020, West Bengal, India
E-mail: banashreemajumdar1@gmail.com

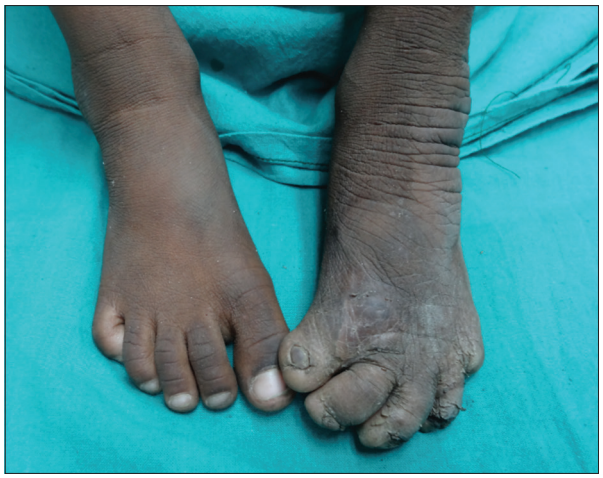

Figure 1: Pseudoainhum affecting left little toe

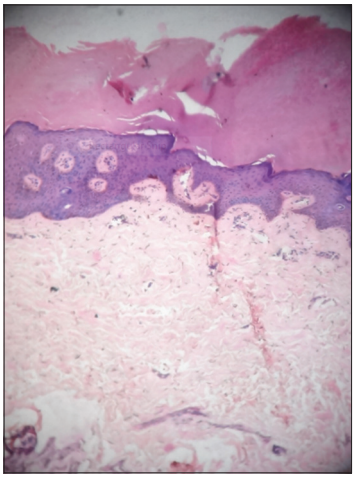

Figure 2: Photomicrograph showing compact hyperkeratosis and papillomatosis in a church spire manner with preserved granular cell layer (H and E, ×100)

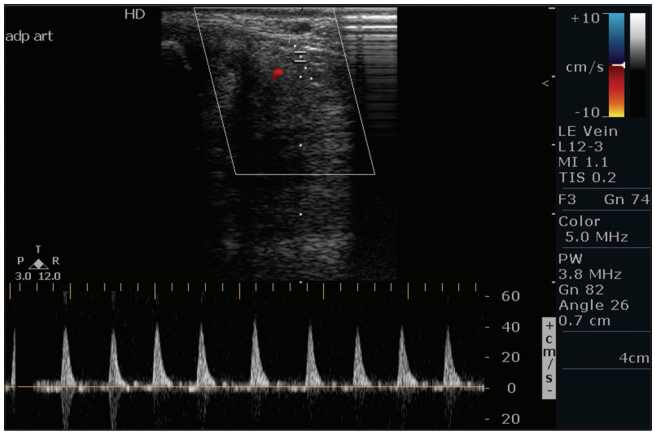

Figure 3: Doppler study showing replacement of the normal triphasic flow by a monophasic pattern

responsible for acquired pseudoainhum are palmoplantar keratoderma, Olmsted syndrome, pachyonychia congenita, erythropoeitic protoporphyria, infections like leprosy, trauma, cold injury, neuropathy and systemic sclerosis.[2] Congenital cases of pseudoainhum must be distinguished from aplasia, hypoplasia and acromelia. Papillon-Lefevre syndrome is also associated with pseudoainhum but can be ruled out by the presence of periodontitis and diffuse palmoplantar keratoderma.[3] One of the close differential diagnosis is keratoderma hereditarium mutilans or Vohwinkel's syndrome, which is inherited as an autosomal dominant disease, but a recessive type has occasionally been described.[4]

A peep into pathology tells us that congenital annular bands are developmental anomalies in which hyperplasic collagen nearly replaces the subcutaneous tissue.

In summary we present a rare case of pseudoainhum in combination with dynamic peripheral arterial disturbance with no Doppler ultrasonographic evidence of limb ischemia In our case pseudoainhum is associated with unilateral clubfoot, also according to one study in more than half of reported cases, they are accompanied by other mesenchymal anomalies, particularly syndactyly and clubfoot and thus common embryologic derivation of these anomalies suggests a basic mesenchymal developmental defect.[5] Reperfusion of the affected limb has been suggested.[6] Depending on the limb anomaly, surgical management with staged Z-plasty may be performed.[7] We referred the child to cardio thoracic and vascular surgeons for further management.

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