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Resolution of Pseudoainhum with Acitretin in Lamellar Ichthyosis

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Abstract

Lamellar ichthyosis is an autosomal recessive type of ichthyosis characterized by abnormal skin scaling, ectropion and ear abnormalities. Pseudoainhum is the appearance of constriction bands around digits which can lead to autoamputation of digits. Here we report a case of Lamellar ichthyosis with Pseudoainhum, which showed reversal of Pseudoainhum with 6 months of acitretin therapy.

Key-words: Pseudoainhum, congenital ichthyosis, acitretin

Introduction

Lamellar ichthyosis is an autosomal recessive type of ichthyosis which often presents as a collodion baby at birth and later manifests as ichthyosis with large polyclonal, dark adherent scaling. Other clinical features include ectropion, onychodysplasia, palmoplantar keratoderma, cicatricial alopecia and ear abnormalities.[1] Pseudoainhum is the term used for appearance of annular constriction bands around digits.[2] Pseudoainhum occurs in various conditions like palmoplantar keratoderma, trauma, systemic sclerosis, chronic plaque psoriasis, pachyonychia congenita, erythropoietic protoporphyria, and Olmsted's syndrome.[3] Autoamputation of digits is a dreaded complication of pseudoainhum. Pseudoainhum is generally managed surgically; here we have a case of lamellar ichthyosis with pseudoainhum which resolved with acitretin therapy.

Case Report

A 29 - year- old female came with complaints of scaling all over the body with extensor predilection since birth which was exacerbated during winter. She was a single full-term baby, born of a consanguineous marriage (second degree), by normal vaginal delivery. History suggestive of collodion membrane was present at birth. There was a history of developing large brown scaling all over the body predominantly on the extensor aspect of limbs since childhood along with watering and difficulty in closure of eyes. There was no history suggestive of erythroderma, blisters, hearing/visual abnormality or any other systemic complaints.

On examination, she had generalized ichthyosis, with large dark brown scales seen all over the face, trunk and extensor aspect of extremities. Ectropion and scaling were seen in face with no gross abnormality of the ears (Figure-1 A, B, C).

Distal onycholysis and longitudinal ridging were seen in toe nails. Constriction band (Pseudoainhum) was seen in left little finger (Figure- 1D). Ophthalmology and ENT evaluation were normal. All routine investigations like complete blood count, renal profile, liver profile, lipid profile were within normal limits. Skin biopsy showed hyperkeratosis, parakeratosis and focal spongiosis (Figure 2).

Based on history, examination and investigation, she was diagnosed as a case of congential autosomal recessive lamellar ichthyosis and was treated with keratolytics, moisturizers and antihistamines. She was started on acitretin 25 mg OD after routine investigation and ensuring negative pregnancy test. As she was not willing for surgical management of pseudoainhum, she was followed up for 6 months. After 6 months, she showed near complete resolution of pseudoainhum while she was on acitretin. (Figure-3)

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Figure 1A: Clinical image showing scaling of face | Figure 1B: Clinical image showing generalized ichthyosis, with large dark brown scales seen all over the extensor aspect of upper extremities | Figure 1C: Clinical image showing generalized ichthyosis, with large dark brown scales seen all over the extensor aspect of lower extremities | Figure 1D: Constriction band (Pseudoainhum) was seen in left little finger

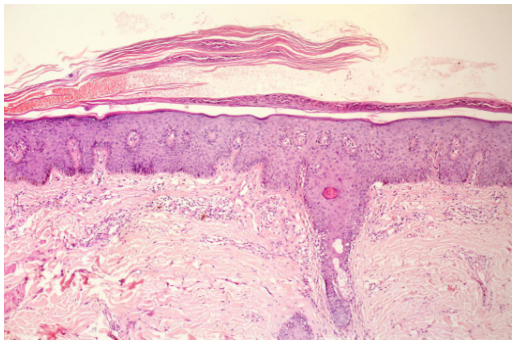

Figure 2: Histopathology- hematoxylin and Eosin staining at 10x showing hyperkeratosis, parakeratosis and focal spongiosis

Figure 3: Pseudoainhum post acitretin treatment

Discussion

Lamellar ichthyosis is an autosomal recessive ichthyosis due to multiple gene defects such as transglutaminase 1 expression (chromosomal 14q11), NIPAL4 gene (protein ichthyin), CYP4F2 (cytochrome 450 polypeptide), CERS3 gene (ceramide synthase3), LIPN (acid lipase), PNPLA1 (patalin like phospholipase). It is characterized by ichthyosis, ectropion, alopecia, palmoplantar keratoderma. Lamellar ichthyosis is rarely associated with pseudoainhum, only few cases are reported previously.[4] Here we report a case of pseudoainhum associated with lamellar ichthyosis who was started on acitretin. She showed reversal of pseudoainhum and significant resolution of ichthyosis at follow up after six months of acitretin. There are only 2 previous reports of reversal of pseudoainhum with acitretin published in literature. First report was by Kura et al in a case of Camisa syndrome observed complete reversal of pseudoainhum with 6 months of acitretin.[5] Second was a case of palmoplantar keratoderma with alopecia and pseudoainhum improving with 2 months of acitretin reported by Richey et al.[6] Therefore, this case highlights the potential role of acitretin as a non-surgical option in managing pseudoainhum associated with ichthyosis.

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