

Olmsted Syndrome Associated with Somatic Type of Delusion

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Abstract

The inheritance of Olmsted syndrome that is a very rare congenital with transgredient palmoplantar keratoderma is distinguished by the presence of massive hyperkeratosis with fissured skin and periorificial chaps. It usually appears during the early life and mostly in male patients. Herein we report a case of Olmsted syndrome which is associated with ichthyosis and somatic type of delusion during adulthood

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Introduction

The first case of Olmsted syndrome (OS) was reported in 1927 as a combination finding of congenital palmoplantar keratoderma with periorificial hyperkeratosis.¹ In subsequent years 20 more cases were reported.²⁻⁵ Olmsted syndrome is an extremely rare disorder, and often is distinguished by severe bilateral palmoplantar keratoderma and periorificial keratotic lesions.^{3,4} It seemed that these two main symptoms constitute characteristic composition of findings to the exclusion of other syndromes, but next reported cases undermined it. Hence two main symptoms constitute characteristic composition of findings, to the exclusion of other syndromes.²

Although, the thickness of the skin lesions differ during the development of the disease, most lesions tend to become thicker and harder with time.² Orofacial keratosis is one of the consistent findings of OS.¹ The condition more often is confused with acrodermatitis entropathica, hidrotic ectodermal dysplasia, mal de Meleda.¹ Herein, we present a case of OS in association with ichthyosis and delusion disorder in an adult patient.

Case Report

A 29-yr-old man with massive palmoplantar and periorificial hyperkeratosis associated with generalized ichthyosis has been referred to the Dermatology Clinic of Psychiatric Field of Shahid Beheshti Hospital of Kerman University of Medical sciences (Fig 1). According to his parents, progressive palmoplantar hyperkeratosis had appeared of six months age, which gradually led to massive hyperkeratosis and painful deep fissures on the palms and soles. In familial history his parents had been unaffected and had no family genetic relations. He had four brothers one of which had the same disease and died at the age of five-months-old. He also had three sisters who were healthy and had no signs of the disease.

Although his general condition was good his psychiatric abnormality, mono symptomatic hypochondriac psychosis,

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Fig1: Palmar hyperkeratosis and nail deformity

was compatible with somatic type of delusion. Flexion deformities of his fingers began in his early childhood and his delusion status has been focused on the right side organ dysfunction. Thickening and cumulating of keratin materials as well as composing giant horn-like palmar masses made him not being able to straighten his fingers, hold or catch objects and as a result of deep plantar fissuring he was not able to walk normally (Fig 2).

Ichthyoses and generalized skin chaps in association with severe verrucous hyperkeratotic plaques were seen on the extensor surfaces of sacral area and also on both knees and elbows. Scalp hair was brittle and turbid especially on the parietal area. The nails were dystrophic hyper-curvature and brownish to black color. The mucosal surfaces and the teeth development were normal. Topical therapy such as soaking in potassium permanganate, wet dressing, using salicylic or boric acid, topical corticosteroids etc. were not effective. Systemic retinoid was not used by him because of financial problem and delusional disorder.

Pathological examination of the lesions on the palms and knees showed severe hyperkeratosis, focal parakeratosis, acanthosis, with hyper vascular and mild mononuclear cell infiltration in papillary dermis, and of a biopsy of ichthyotic skin showed mild orthokeratosis diminishing of granular layer marked follicular keratin plugging was compatible with *ichthyosis vulgaris*.



Fig2: Plantar hyperkeratosis

Discussion

Some authors believe that hyperkeratotic lesions may appear in response to trauma.⁶ The illness usually begins in early childhood, at the time when the child starts walking and holding things.⁶ Gioigii and coworkers reported a case when the skin lesions appeared at the time the patient was 30-yrs-old (late onset).⁷ Whereas, the first presentation sign of this syndrome in our case was palmoplantar keratoderma, and according to his parents it appeared when he was six-months-old.

Our case had Olmsted syndrome, in association with somatic type of delusion as mentioned by Kaplan.⁸ Ichthyosis and false belief that was based on the correct inference about external reality, which was not consistent with his intelligence and cultural background; so he could not describe corrections by reasoning.⁸

Somatic type delusional disorder is also known as a mono symptomatic hypochondriac psychosis and usually is a false belief function of the body.⁹ His psychological disorder was somatic type delusion, with unilateral abdominal dysfunction. This association seemed to have sporadic occurrence.¹⁰ Although, the causes of delusional disorders are not known, certain risk factors may be relevant to the etiology and may deserve further explanations.

The risk factors may be social isolated personality, with positive family history of sensory

impairment and social isolation and immigration during older ages. Social isolation was a risk factor in this patient; however, it is not easy to say that there was any a strong relationship between the two disorders. Therefore, more investigations are needed in this field as stated in a twin child by Cambiahi et al.¹⁰ A familial case with an autosomal or X-linked dominant mode of inheritance is also reported about this syndrome.⁴ Hereditary process in our case guided us as an X-linked or rarely as an autosomal dominant type with variable penetration, but the severity of disease in this case and similar finding in his brother that has been died at last, lead us to assume that autosomal recessive inheritance could be the cause of this syndrome in this family.^{4,10}

In summary, Olmsted syndrome is a congenital palmoplantar and periorificial keratoderma and is a rare syndrome that usually appears in the first few years of life. Olmsted syndrome reported in this study was also associated with ichthyosises and somatic delusion, a false belief involving function of the right side of the body.

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