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PAPILLON - LEFEVRE SYNDROME

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A case of Papillon-Lefevre syndrome is reported in a six years old boy. He had early loss of deciduous teeth with periodontosis, palmoplantar keratoderma extending on to the pressure points of the body, hepatosplenomegaly and congenital hydrocele.

Key words : Papillon - Lefevre syndrome

Introduction

Papillon-Lefevre syndrome is a disorder, described by Papillon and Lefevre in 1924. They noticed severe destructive periodontitis affecting both deciduous and permanent dentitions associated with palmo-plantar hyperkeratosis. These manifestations usually appear in childhood between 1-4 years of age and decidous teeth exfoliate within or at the age of six. Permanent teeth erupt normally but soon get affected by periodontal disease. Individual becomes edentulous within teenage. Hyperkeratotic lesions extend to knees and elbows.[1] Some cases have shown inconsistent manifestations like calcification of falx cerebri and choroid plexus, calcification of the dura, attachment of the tentorium, thumb nail dystrophy and hyperhidrosis.[2,3]

Case Report

A 6-year-old boy born to a consanguinous married couple presented with hyperkeratotic lesions of soles and palms, extending on to the dorsae of feet and hands, elbows and knees. He also had hepatosplenomegaly and congenital hydrocele. Extra oral examination showed painless bilateral submandibular lymphadenopathy. Intra oral examination revealed complete edentulous ridges except for the presence of maxillary second deciduous molar on both the sides. The gingiva in relation to these teeth was red, soft and oedematous. Teeth showed grade III mobility with periodontal pocket, gingival recession and mucosa over edentulous area was normal.

Orthopantomograph showed bone loss around maxillary deciduous second molar. Permanent teeth were within the bony crypts. Haematological investigations like bleeding time, clotting time, total count and differential counts were within normal limits. Skin biopsy showed irregular hyperkeratosis, marked acanthosis and a conspicuous perivascular infiltrate of lymphocytes and histiocytes. With the available other investigations, paediatricians could not find the cause for hepatosplenomegaly.

Comments

Papillon-Lefevre syndrome is probably inherited as an autosomal recessive disease with an incidence of 1-4 per million people. Our patient had palmo-plantar keratoderma, extension of skin lesions beyond the palms and soles and associated manifestations like hepatosplenomegaly and congenital hydrocele. It is quite interesting to know that these inconsistent findings have not been recorded in the earlier literature.[4]

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