Papillon-Lefevre Syndrome: A Report Of Two Cases

INTRODUCTION

Papillon-Lefevre syndrome (PLS) is a rare (1-4 cases per million) autosomal recessive inherited disorder of keratinisation characterised by diffuse palmoplantar hyperkeratosis and aggressive periodontitis affecting both primary and permanent dentition.

CASE REPORTS

CASE 1

A 13 year old boy presented with persistent thickening and scaling of skin of palms and soles since the age of 2 years, with h/o winter aggravation. There was history of recurrent cutaneous infections and swelling of gums. There was no history of excessive sweating. Orthopantomograph showed moderate alveolar bone loss.

CASE 2

An 8 year old girl came with complaints of thickening of skin of palms and soles along with itching, erosions & reddish thickened skin over knees and elbows for last 6 years. She also had h/o repeated gum swelling. Intraoral examination showed loss of most teeth.

DISCUSSION

Papillon-Lefevre syndrome usually appears in childhood. Males and females are equally affected. Patients are normal at birth. Transgradient palmo-plantar keratoderma develops concurrently with oral lesions. Other symptoms include hyperhidrosis, arachnodactyly, intracranial calcification, increased susceptibility to infections and mental retardation. Dermatological manifestations of PLS are usually treated with emollients, salicylic acid and topical steroids. Oral retinoids such as acitretin, etretinate and isotretinoin are beneficial in treating both dental and cutaneous lesions of PLS. PLS-associated periodontitis is treated with appropriate periodontal/antimicrobial treatment to prevent or delay tooth loss and enhance early replacement of missing teeth for preservation of function and aesthetics.

CONCLUSION

Complete history, clinical examination and relevant investigations can enable a prompt and timely diagnosis of a rare genetic disorder like Papillon-Lefevre syndrome. A multidisciplinary approach involving a team of dermatologist, pediatrician, and dental surgeon team (periodontist, pedodontist, and prosthodontist) is important for overall care of the patient with PLS.