POROKERATOSIS OF MIBELLI IN A FAMILY

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The occurrence of porokeratosis of Mibelli in all members of a family when only one parent (mother) had the disease is reported. The 17 members of the third generation are still under 17 years of age and have not revealed any lesions of porokeratosis.

Key Words : Familial Porokeratosis of Mibelli, Autosomal dominant

Introduction

Porokeratosis of Mibelli is a rare genodermatosis with an autosomal dominant mode of inheritance.[1] Males are more often affiliated than females and the onset of disease is usually during childhood.[2] Cases have been documented singularly or in multiple members of family.[3] In the absence of many reports of familial involvement of porokeratosis from our country, we are tempted to report porokeratosis of Mibelli occurring in two generations in which all members of 2nd generation were affected.

Case Report

A 35-years-old male reported to skin out-patient department with multiple asymptomatic irregularly shaped skin lesions of 13 years duration. The examination revealed numerous annular lesions which were widely spread over body and ranged from pea size to about 7 cm in diameter. There was a typical hyperkeratotic, raised border with deep furrow. The central area of the lesions showed atrophy.The histopathological examination from the margin of a representative lesion confirmed the diagnosis of porokeratosis of Mibelli.

The family history revealed the presence of similar lesions in the mother and all his brothers and sisters (2 brothers and 3 sisters). Three members of above family could be examined in the department who had similar lesions with atrophic centre and raised border with furrow. The other members who are living in remote areas could not be called for examination. The 17 members of third generation are under 17 years of age and no one is still affected by the disease (Fig.1).

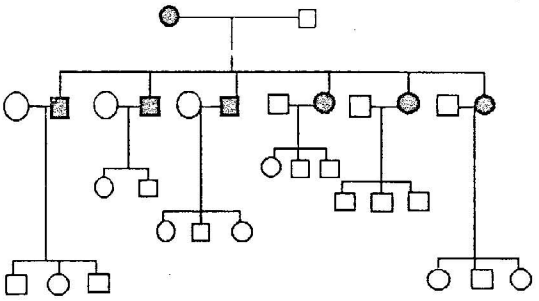

Fig. 1. Showing Pedigree of family.

Comments

The familial occurrence of porokeratosis of Mibelli has been reported only once by Prasad et al,[3] however there are occasional reports available in the literature. Prasad et al reported familial occurrence of porokeratosis in 2 separate families involving 18 and 4 cases respectively. They showed involvement of all the members in the first family when both parents were involved & hypothesized it on the basis of double dosing. In other family both children expressed the disease when only

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father had the disease. The family reported by us manifested disease in all offsprings when only mother expressed porokeratosis. This is in accordance with observations of Prasad et al[3] in second family of 4 cases and to best of our knowledge, only other instance where 100% expression of porokeratosis is reported and is difficult to explain by simple rule of autosomal dominant inheritance.[4]

The 17 members of third generation (the eldest one 17 year old and the youngest one 10 years old) have yet not expressed porokeratosis which is again unusual since porokeratosis usually manifests in early childhood. This could probably be explained on the accepted fact that additional trigering factors besides the inherited abnormal clones are necessary for clinical manifestation of porokeratosis.[5]

References

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