Coexistence of disseminated superficial and giant porokeratosis of Mibelli with squamous cell carcinoma

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ABSTRACT

Porokeratosis of Mibelli is a genetic disease transmitted as an autosomal dominant trait. The giant type of porokeratosis is a relatively rare entity and is associated with an increased risk of malignancy. The aim of this article is to present this rare case of giant porokeratosis associated with squamous cell carcinoma.

Key Words: Porokeratosis of Mibelli, Squamous cell carcinoma

INTRODUCTION

Porokeratosis of Mibelli is a genetic disease transmitted as an autosomal dominant trait.[1] It is a specific disorder of keratinization, clinically characterized by irregular annular plaques with well-demarcated, raised, hyperkeratotic borders. The border is usually more than 1 mm in height and contains a thread-like groove. Giant porokeratosis is rare. The lesion may be 10 cm in diameter and has a high incidence of malignant transformation.[2]

CASE REPORT

A 45-year-old woman presented with a giant plaque on her back of 30 years' duration and ulceration since 5 months. The plaque was about 1 cm in diameter initially and gradually spread over a period of years to involve almost the entire back. It showed ulceration and bleeding since 5 months (Figure 1). A few similar lesions appeared during this time on both hands and face and those were associated with itching.

On examination, there was a 30 x 20 cm plaque on the back showing a well demarcated hyperkeratotic border with a thread-like groove. Overlying this was a 10 x 10 cm fleshy exuberant plaque which bleeds on touch.

Histopathology of the edge of the plaque was suggestive of porokeratosis of Mibelli. Fine needle aspiration cytology of the ulcerative lesion revealed features of squamous cell carcinoma. The patient refused any other investigation and treatment.

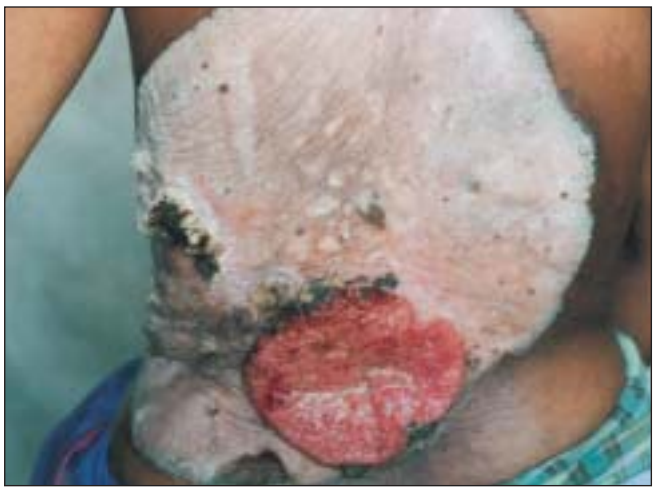

Figure 1: Fleshy exuberant ulcerated plaque superimposed over a large keratotic plaque with well demarcated hyperkeratotic border

On family screening, it was found that the patient's father and both her sisters (aged 40 years and 53 years) had disseminated superficial porokeratosis (the latter since 10-15 years of age), but not her mother or her 50-year-old brother. Hence the patient had disseminated superficial porokeratosis along with giant porokeratosis of Mibelli with squamous cell carcinoma.

DISCUSSION

Classic porokeratosis was described by Mibelli in 1893 as one or more localized, gradually progressive, hyperkeratotic, irregular plaques with central atrophy and a prominent peripheral keratotic ridge. A more superficial disseminated form was described by Respighi and later by Andrews.[1,2]

In the rare giant type of porokeratosis there is an increased risk of malignant transformation. Malignant changes can occur in long standing lesions of porokeratosis. Squamous cell carcinoma is the commonest type of malignancy.[3,4]

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