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Medical Sciences **Case Report……!!!**

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FAMILIAL POROKERATOSIS OF MIBELLI WITH OVERLAP OF DISSEMINATED SUPERFICIAL ACTINIC POROKERATOSIS

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Porokeratosis of Mibelli, Disseminated Superficial Actinic Porokeratosis, Acetretin

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ABSTRACT

Porokeratosis of Mibelli (PM) is a rare disorder of keratinization with autosomal dominant mode of inheritance characterized clinically by marginate scaly lesions and a column of parakeratotic keratinocytes on histological examination. A 23 year old male patient of PM is reported with involvement of three generations in the family. Patient had overlapping features of PM as well as disseminated superficial actinic porokeratosis. He was put on oral Acetretin with good response within 4 weeks.

INTRODUCTION

Porokeratoses are group of disorders with defective keratinization characterized by marginate scaling lesions demonstrating column of parakeratotic keratinocytes (the coronoid lamella) on histopathology. [1] Various forms of porokeratoses have been described- Disseminated forms such as disseminated superficial actinic porokeratosis (DSAP), porokeratosis of Mibelli (PM), linear, giant and palmo-planter porokeratosis. Of these DSAP is the most common form with multiple small lesions while PM is the most classical variety with single or few but larger lesions. Porokeratosis of Mibelli (PM) is a rare genodermatosis, first described by Mibelli in 1893. Males are more commonly affected. Lesions are usually single or few well defined plaques with raised keratotic border. Usual sites are extensors of limbs but uncommonly mucosal lesions are also seen. [2] Various topical treatment modalities have been described for localized PM which include curettage, cauterization, CO2 laser, cryotherapy, 5-florouracil and 5% imiquimod cream among others.[3][4] Systemic agents like oral retinoids, dexamethasone pulse therapy and Grenz rays have been successfully used for treatment of disseminated forms.[5][6][7]

Case Report: A 23 year old male patient presented with multiple asymptomatic skin colored well defined plaques of various sizes distributed all over the body since childhood. First lesion appeared around the age of 5 years at left knee. Over the years patient developed new lesions over limbs, chest, back, neck and face. The lesions increased in size progressively over the years. Face was last to be involved, where small new lesions had appeared in last few months. Maximum numbers of lesions were present over the limbs and face. Two lesions were observed over right ear and one was seen left retro-auricular region. Largest plaque was on the left knee area, approximately 12x10 cm in diameter. It was well defined, annular in shape with hyperkeratotic border and slight atrophic center. Smallest lesion was found over left naso-labial area, which was about 4x3mm in diameter. Rest of the lesions were of intermediate sizes, all retaining the characteristic annular morphology.

Figure-1 Largest lesion on the knee

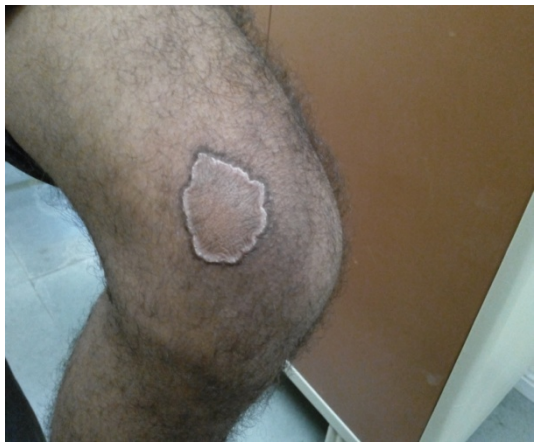
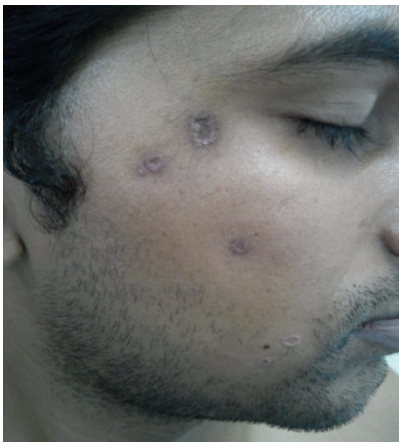


Figure-2 Lesions on the face



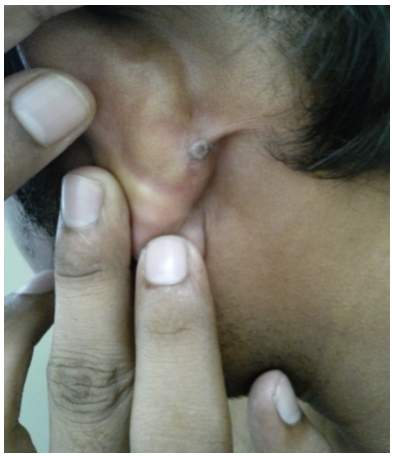
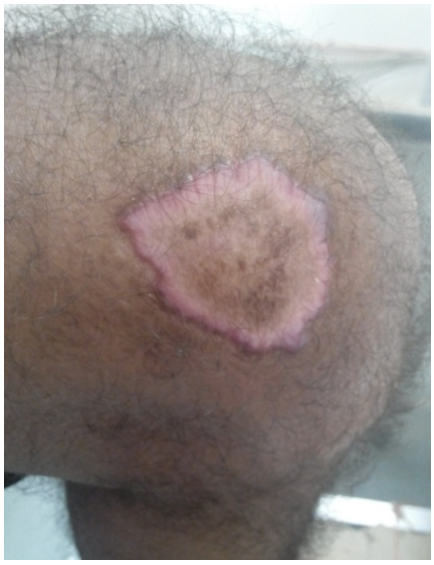
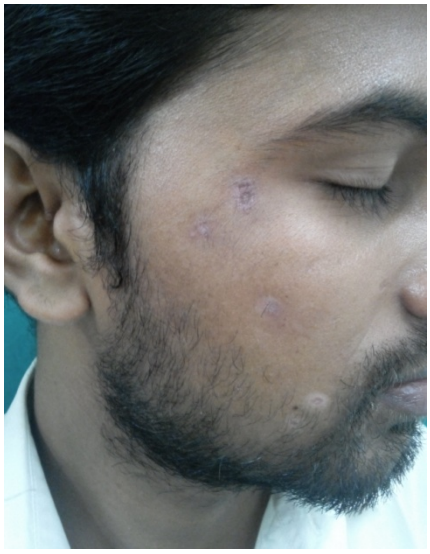


Figure-3 Lesion over retro-auricular area.

Histopathological examination from the knee lesion demonstrated focal invaginated epidermis showing parakeratosis while adjoining epidermis showed acanthosis and hyperkeratosis. Moderate lymphocytic infiltrate was found in dermis.

There was a positive family history of similar lesions in patient's father and grandfather. No other sibling was involved in any of three generations.Patient was put on oral acetretin 25mg after requisite tests. In 4 weeks patient has shown good response. Figure-4 and Figure-5 showing good response to acetretin therapy.





DISCUSSION

Classical PM is more common in males, with childhood onset, positive family history and distribution over limbs. These features were present in our patient. Though in classical PM lesions are generally few, our patient had more than 30 lesions with generalized distribution. Even uncommon areas such as ears and retro-auricular area were involved. This is not usually seen in classical PM. Presence of multiple small sized lesions and distribution over face and trunk suggests DSAP. Mehta and Balachandran also reported simultaneous co-occurrence of DSAP and PM in their case. [8] Though familial involvement in classical A rare varient of porokeratoses disseminated porokeratosis of childhood has also been described in the literature which may run in families. [1]

There have been occasional case reports of PM running in families from India. [9][10] Prasad and Singh reported PM in 18 and 4 members of two families. [9]Prasad et al also reported a case of familial PM in which two generations were involved.[10] In that case patients mother as well as all the siblings, 2 brothers and 3 sisters were involved. Our case also had unique inheritance pattern with only eldest sibling of three generations being involved.

As our patient had multiple disseminated lesions we used systemic treatment with acetretin as there have been enough evidence of successful treatment of porokeratosis of Mibelli with oral retinoids. A good response was observed within 4 weeks. Small lesions almost disappeared and large lesions with raised borders flattened with treatment. In our opinion, acetretin should be prescribed in patients with porokeratosis if the lesions are extensive.

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