Case 7

A 53-year-old Thai female from Bangkok province

Chief complaint:

Skin lesions on arms and trunk for 10 years

Present illness:

The patient presented with 10 years history of asymptomatic brownish macules on both arms.The lesion have been spreading to her neck and trunk for 4 months. Her sister has similar skin lesions.

Past history:

Spinal stenosis

Family history:

Her sister has similar skin lesions.

Physical examination:

Multiple discrete well demarcated brownish macules with thread-like border and furrow, diameter 5-10 mm on arms, V-shape of neck and few discrete lesion on trunk.

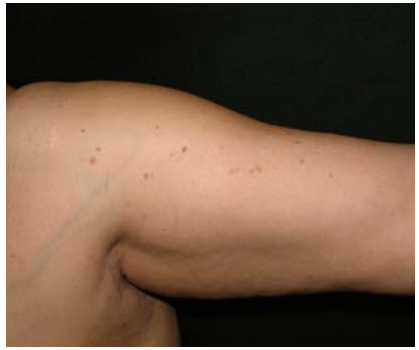

Fig. 7.1

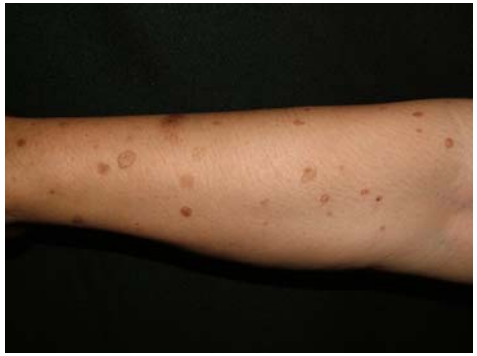

Fig. 7.2

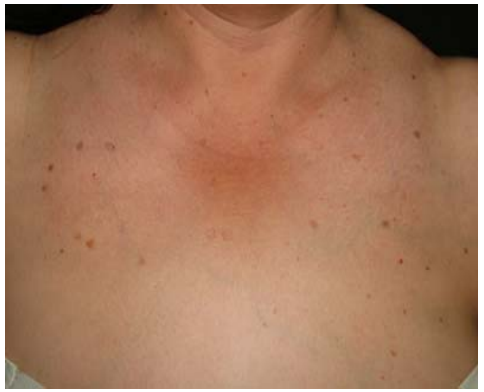

Fig. 7.3

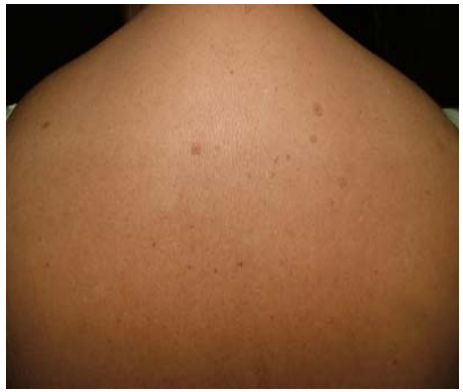

Fig. 7.4

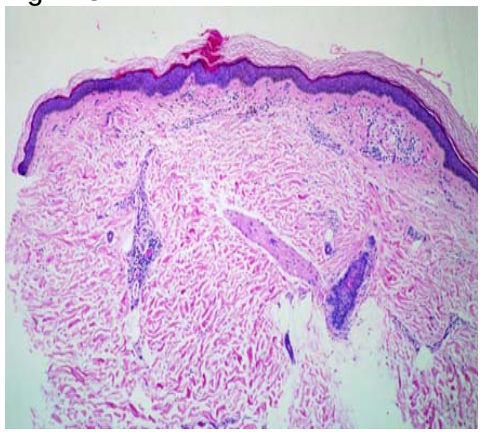

Fig. 7.5

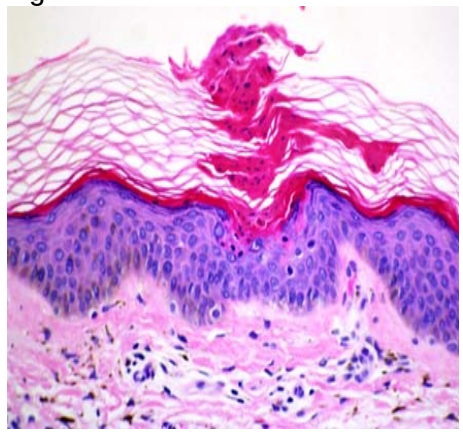

Fig. 7.6

Histopathology (S08-017039) (Fig.7.5, 7.6)

- Thin and tall columns of parakeratosis plugged on the epidermis with focal hypogranulosis and dyskeratotic cells.
- Superficial infiltrate of lymphocytes with melanophages in the upper dermis.

Diagnosis: Disseminated superficial actinic porokeratosis
Presenter: Silada Kanokrungsee
Consultant: Penpun Wattanakrai
Treatment: Sun protection and topical 0.05%tretinoin.

Discussion:

Porokeratosis is a group of genodermatoses consisting of keratotic papules or plaques which enlarge centrifugally, usually leaving a somewhat depressed or atrophic center surrounded by a raised hyperkeratotic, thread-like border[1]. Porokeratosis can be classified as porokeratosis of Mibelli (PM), disseminated superficial porokeratosis (DSP), disseminated superficial actinic porokeratosis (DSAP), porokeratosis palmaris et plantaris disseminata (PPPD), porokeratosis punctata palmaris et plantaris (PPPP), linear porokeratosis (LP), and craniosynostosis, anal anomalies, and porokeratosis (CAP syndrome) [2,3].

DSAP is the most common porokeratosis, autosomal dominant, characterized by small numerous superficial annular, keratotic and slightly atrophic centers lesion surrounded by elevated ,furrowed hyperkeratotic borders , ranging from 2 to 5 mm in diameter usually occurs on the sun-exposed areas.Generally,the cutaneous lesions begin to develop in teenagers of affected families with near complete penetrance by the third to fourth decades of life.

To date ,locus heterogeneity encountered in DSAP which three different chromosome loci have been reported to implicate in etiopathogenesis of DSAP : 6.4cM interval in 15q 25.1-26.1(DSAP 2) ,18p11.3 and 9.6 cM interval in 12q23.2-q24.1(DSAP1) ,the latter chromosome region harbors several genes,of which some were reported a candidate gene to be associated with DSAP including SSH1 ,ARPC3 ,SART3. Immune suppression and excessive sun exposure are known precipitants that may lead to more severe disease[4].

The cornoid lamella is the histopathologic hallmark of all forms of porokeratosis. It is essential that the specimen be taken from the peripheral, raised, hyperkeratotic ridge to demonstrate this finding.The cornoid lamella consists of a thin column of tightly packed parakeratotic cells within a keratin-filled epidermal invagination. The parakeratotic column extends at an angle away from the center of the lesion and develops from the interfollicular epidermis. In the epidermis beneath the parakeratotic column, the keratinocytes are irregularly arranged and have pyknotic nuclei with perinuclear edema. No granular layer is seen within the parakeratotic column, while the keratin-filled invagination of the epidermis has a well-developed granular layer. The papillary dermis beneath the cornoid lamella contains a moderately dense, lymphocytic infiltrate and dilated capillaries. Dermal amyloid deposits have been described in some cases of DSAP[4].

Course and prognosis are chronic and slow progression with lesion increasing in size and number occurs over decades. Malignant transformation has been reported, squamous cell carcinomas and Bowen's disease,although this is still a matter of debate.Unfortunately, there is no cure for DSAP, and current therapies are often ineffective, painful, or unappealing. The most commonly used treatments include cryosurgery, topical 5-fluorouracil, topical vitamin D-3 analogues, and retinods[4-7]. Some studies suggest a potential for diclofenac sodium 3% gel as a possible treatment for DSAP[8]. Protection from the sun, use of emollients, and observation for signs of malignant degeneration may be all that is needed for many patients.

References

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