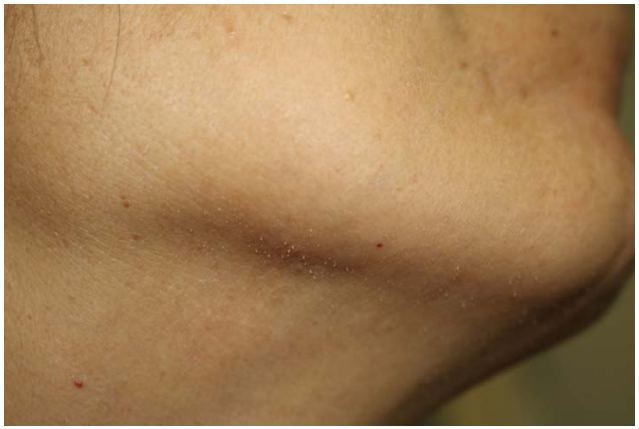
Case 20
A 54-year-old Thai woman from Bangkok

Chief complaint: multiple spiky spicules on her neck and face for 1 month


(Fig. 20.1)

Present illness: She developed asymptomatic multiple minute spiky spicules on her neck and face for 1 month. These lesions were easily removed by scratching or rubbing without bleeding and usually reappeared within few weeks. She was otherwise in good health.

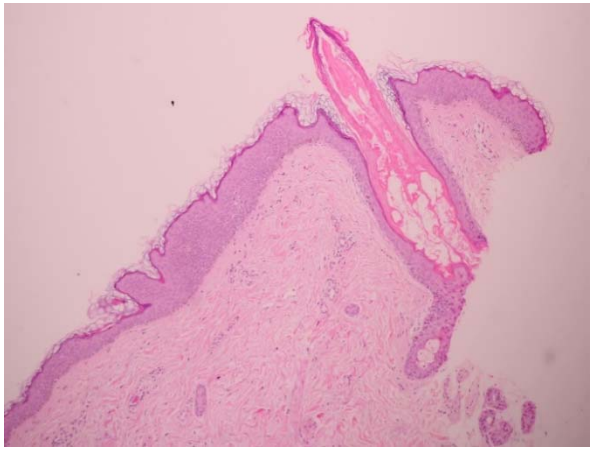
Past history: Her underlying disease was dyslipidemia. Her current medication is simvastatin 10 mg per day. Her previous operation was total abdominal hysterectomy with bilateral salpingo-oophorectomy 13 years ago from myoma uteri. She is in a surgical menopause condition.

Family history: There was no family history of similar cutaneous lesions, autoimmune diseases or malignancies

Dermatological examination: (Fig. 20.1)
Multiple discrete skin-colored tiny filiform hyperkeratotic papules on face and neck

Physical examination:
Physical examination other than skin revealed no abnormality.

Histopathology: (S16 - 26748, Submandibular area) (Fig 20.2)


(Fig. 20.2)

- Dilated follicles with digitate follicular hyperkeratosis and parakeratosis
- No trichostasis or koilocytes

Laboratory investigations:
- CBC: Hct 35.4%, WBC 4,990 cells/µL (N 51%, L 42%, Mono 6%, Eo 1%), Platelets 211,000 cells/µL

- BUN 16 mg/dL, Cr 0.65 mg/dL
- LFT: ALP 72 U/L GGT 49 U/L, AST 26 U/L ALT 24 U/L, TB 0.6 mg/dL DB 0.2 mg/dL, TP 78 mg/dL Alb 40.3 mg/dL
- Serum protein electrophoresis (SPEP): Normal

Diagnosis: Idiopathic follicular hyperkeratotic spicules

Treatment: 0.1% adapalene gel apply on lesions once daily

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Discussion:

Hyperkeratotic spicules is a rare cutaneous disorder characterized by follicular or non-follicular digitate keratosis of the face [1]. The disease has been reported in associated with various conditions such as hematologic disorders including multiple myeloma[2], paraproteinemia[3], cryoglobulinemia[4] and Sézary syndrome[5], as well as drug-induced including acitretin[6] and sorafinib[7]. A case with no associated condition was reported one time in the literature[8].

Various hypotheses for pathogenesis of spicule formation include a change in normal keratinization, or a particular autoantibody activity in the monoclonal immunoglobulin modified the keratinization process. In 1990, Bork *et al.*[9] firstly demonstrated that the follicular-plug component contained myeloma dysprotein and cryoglobulins precipitating into the follicular infundibulum. The present of lesions localized to cold exposed area suggested that immunoglobulin cryoprecipitation may play a role in the pathogenesis. However, not all patients showed spicules containing monoclonal dysproteinemia[2, 10] such in our case.

Histopathology revealed focal spike-like orthokeratotic or parakeratotic column fillng the infundibulum and protruding above the surface, sparse lymphocytic infiltrate in papillary dermis. Some reported cases with dysproteinemia revealed homogenous compact of eosinophilic inclusions (immunoglobulin) organized in protruded structure and intercellular space between keratinocyte[1].

Various treatments including 12% lactic acid cream, adapalene gel, tretinoin cream, fluocinolone acetonide oil, and antibiotics have previously been tried less effectiveness[6, 10]. Some cases with hyperkeratotic spicules associated with multiple myeloma who received chemotherapy showed a marked improvement of the skin lesions[2, 4, 10, 11].

In conclusion, we reported a case of idiopathic follicular hyperkeratotic spicules with partially improved after treatment by of topical adapalene for 2 months. Due to the reports of serious associated conditions, we suggest that finding the underlying disease in all cases is the most important strategy in management of patient and regularly re-evaluation should be performed in all cases with idiopathic hyperkeratotic spicules.

References:

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