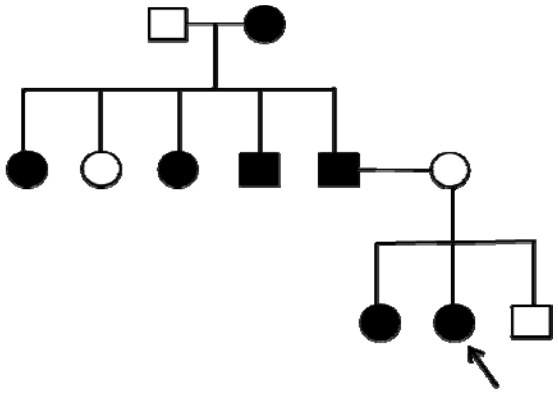
Case 8.1

A 17-year-old Thai female from Bangkok

Chief complaint: Multiple wide spread comedone-like lesions for 5 years

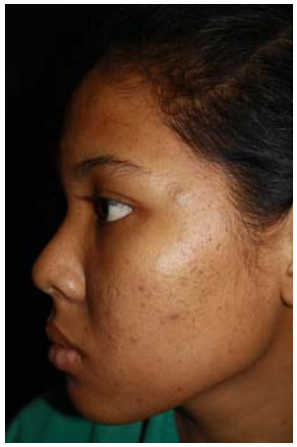
Present illness: The patient presented with recurrent multiple cysts at face, posterior neck, chest, shoulders and upper back. She came to the outpatient clinic of surgical department and was diagnosed as recurrent epidermal cysts. She was treated with multiple courses of antibiotics and surgical drainage. She was sent to dermatological department because one of the doctors obtained her family history of identical lesions. Additional history taking revealed that she had multiple asymptomatic blackish comedone-like papules since twelve, some papules become progressively larger, inflame and pain, leaved discrete pock-like scars especially on her face and back.

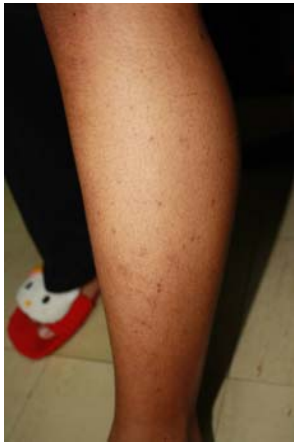
Family history: Many members of her family had similar lesions to the patient which all started around the age of twelve.

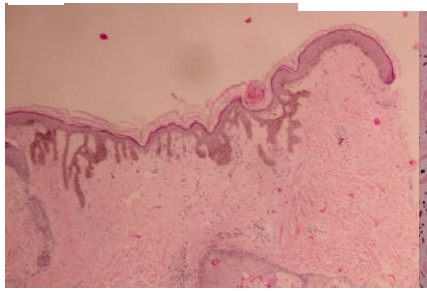


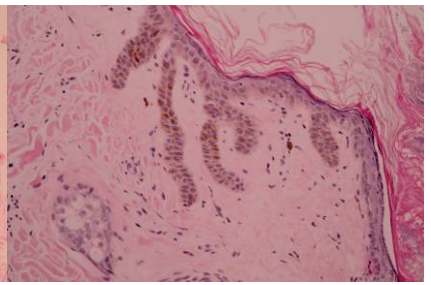
Skin examination: Widespread, symmetrically scattered, comedone-like papules at face, trunk, arms and legs with discrete inflamed nodules and comedones over the back . Mutiple pock-like scars were seen over the face and the back.

Histopathology (S10-15163) follicular dilatation and delling of parafollicular epidermis lined by atrophic epithelium thin, with branching, pigmented, elongated and interconnecting strands infiltrate of lymphocytes melanophage and fibrous in the underlying dermis









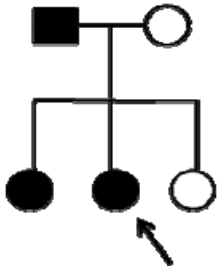
Case 8.2

A 16-year-old Thai female from Patumtanee

Chief complaint: blackish papules and inflamed nodules for 2 years

Present illness: The patient presented with two-year history of multiple widely spread comedone-like papules, in which she thought they were severe acnes. Her lesions initially appeared at her face and subsequently spread to involve upper chest, upper back both upper arms and also both legs. Apart from these skin lesions, multiple pock-like scars were seen over the face and the back. The lesions started as pinpoint dark papules, which gradually increased in number and extent of involvement as the patient grew older. When fully formed, the lesions measured around 0.5 to 1 cm in size, sometimes formed painful swelling inflamed nodules over the face and back. The patient had not received any treatment and was in good health and otherwise normal

Family history: Similar lesions appeared around puberty were noted in her father and older sister.

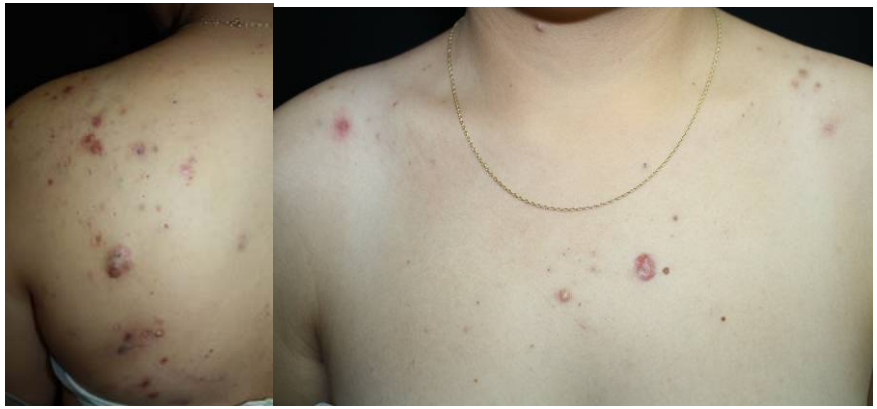


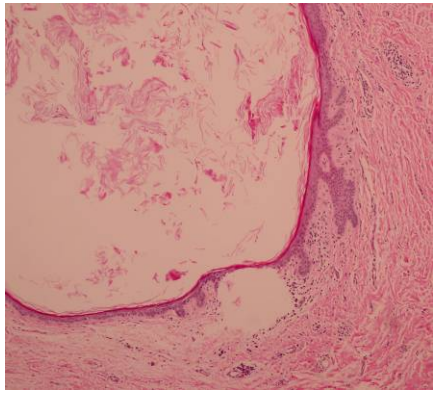
Skin examination: There were multiple dilated follicular openings filled with blackish plugs diffusely involving the face, chest, upper back and upper arms. Some lesions developed inflamed erythematous nodules with central ostia. Discrete pock-like scars are seen.

Histopathology (S11-04331)

keratinous follicular plugging, and cystic dilatation of follicular epithelium lind by thin epithelium with branching and elongated rete ridges

infiltrate of lymphocytes and fibrosis in the underlying dermis





Diagnosis: Familial diffuse comedones
Treatment: Isotretinoin (10 mg) 1 tab once daily.
Presenter: Thanya Techapichetvanich
Consultant: Chanitwan Wichayachakorn

Discussion:

Disorders of inherited widespread comedones may present in two clinical manifestations. The first is called *familial diffuse comedones*. The comedo-like lesions appear progressively and in a diffuse bilateral distribution. The latter, which is more common, is called *familial dyskeratotic comedones*, the comedones are less extensive and demonstrated acantholytic-dyskeratotic changes in the wall of follicular-like structure on histopathology.

Familial diffuse comedones is very rare inherited dermatosis. To the best of our knowledge, there are only two families were reported in the English language medical literature.

In 1967, Rodin et al [1] described a 42-year-old woman who had widely distributed multiple comedones since adolescence; her mother and grandmother also were affected. In 1978, Cantu' et al [2] postulated an autosomal dominante inheritance. He described a family in which 16 members had multiple relapsing comedomes distributed on face, neck, anterior and posterior thorax, abdomen and extremities. The onset was as early as 10 years of age, increased activity during adolescence. The pedigree analysis showed a definite autosomal dominant pattern of inheritance since both male and females members of four consecutive generations were affected including two instances of male-to-male transmission.

The results of histopathological study disclosed follicles filled with keratin plugs. The keratin was lamellar without parakeratosis and the follicular ostia was dilated. No dyskeratotic changes were found. The sporadic case, which has been reported under the term "extensive nevus comedonicus" also occur.[3-4]

Here, we described two adolescent female patients from unrelated families presented with multiple asymptomatic symmetrical wide spread black head comedo-like lesions. The dermatosis usually appear around puberty and show a predilection for the trunk, arms, legs and face, sparing the palms and soles. The lesions gradually increase in number and size with time and frequently causing painful erythematous inflame nodules with central dilated keratin-plug follicular ostia. Patients often complaint history of recurrent swelling nodules which often misdiagnosed as severe nodulocystic acnes or epidermal cysts. Finally, lesions healed and leaving permanent scars that were randomly distributed in all affected area.

The histopathological findings in both cases are similary as previous described.[1-2] The characteristic lack of acanthotic dyskeratotic changes distinguished familial diffuse comedones from infrequent seen, but more common familial dyskeratotic comedones.

In summary, we reported two patients diagnosed as familial diffuse comedones, a very uncommon inherited dermatosis. Our case reports emphasize the autosomal dominant mode of inheritance in these rare genodermatosis. The characterized widespread lesions might probably result from gonadal mosaicism, as yet unidentified mutation. Overall prognosis is favorable as the patients remain in good health except disfiguring scars that may cause social problems especially in the adolescent female population. The treatment of familial comedones has not yet been established due to the scant of reported cases. In our patients, oral isotretinoin 10 mg once daily were given, unfortunately, data regarding the long-term follow-up are lacking.

References

1. HH R, JL B , G B. Diffuse familial comedomes. Arch Dermatol 1967;95:145-46.
2. Cantu JM, Gomez-Bustamente MO, Gonzalez-Mendoza A , Sanchez-Corona J. Familial comedones. Evidence for autosomal dominant inheritance. Arch Dermatol 1978;114:1807-9.
3. MH B , VK D. Extensive nevus comedonicus. Arch Dermatol 1980;116:1048-50.
4. NP. A. Comedonicus nevus of extensive distribution. Arch Derm Syphiolol 1946;53:433.