REVIEW ON LITEARTURE OF EPIDERMO REVIEW DYSPLASIA VERRUCIFORMIS

With 2 Case Reports

By

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Epidermo-Dysplasia Verruciformis literally means faulty (dys) growth or development (plasia) of epidermis in the manner of (formis) warty (verruci) excrescences. The title is applied to a characteristic genodermatosis in which the hands and feet particularly develop verrucous acanthomas

We were inspired to report these cases as to our knowledge there is lack of material on this disease in Indian literature, and both our patients manifested with not only papules, but also with macules Ordinarily epidermo-dysplasia verruciformis presents itself as tiny papules of about 2 mm. in diameter usually affecting the dorsa of the hands and feet, and later the face and neck and remaining parts of the body. The lesions on the face and hands resemble verruca plana, while on the trunk and extremities they are larger and firmer simulating verruca vulgaris. Herewith we are reporting 2 cases in both of which the patients presented with not only papules, but with macules which were hypopigmented.

Epidermo-dysplasia verruciformis of Lewandowski and Lutze is an autosomal recessive disease involving members of a family. In both our cases, the patients were the only affected in their families.

Case Report. I. A twelve year old female patient attended our out-patient department with the complaints of hypopigmented patches and minimal itching on both hands and legs since 2 years. To start with, the lesions were hypopigmented macules without scaling on the dorsa of hands. The macules on the hands increased in size and pari-passu similar new lesions started appearing on the legs. There were no lesions on the face or trunk. There was no history of taking any drugs, trauma or fever prior to the onset of lesions.

There was no similar history in the family General examination and systemic examination did not reveal anything abnormal.

LOCAL EXAMINATION

The patient had well defined, hypopigmented round and oval lesions on both hands, each about 1 inch in diameter. Some of the lesions were slightly raised above the surface. Borders were well defined and a few macules were surrounded by satellite papules. Similar lesions were also seen on both forearms and arms (picture No. 1). On the legs, the lesions were more localised around the knee joint, with tiny lichenoid papular border. There was no scaling and itching. No mucous lesions. (Picture No 2)

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ROUTINE INVESTIGATIONS

Hb: 12.0 gms.%. WBC: 7800/Cmm. Polymorphs-68%. Lymphocytes-25%. Eosinophils-3%. Monocytes-4%. Urine: NAD. Stool: NAD. Screening chest: Normal.

Biposy: It shows hyperkeratosis and acanthosis. At places there is basket-weave appearance (Picture No 3). Some of the cells in the rete malphigili show a vacuolated appearance with nuclei lying in the centre (Picture No. 4)

Case Report No. 2.: A 10 year old male patient came to our out-patient department with hypopigmented macules on both arms of one year duration. There was occasional itching, and the macules were slowly increasing in size. There was no history of drugs or trauma. Family history of similar lesions absent. General examination did not show anything particular.

On local examination, we found round regular macules of about 0.5 to 1 inch in diameter scattered irregularly on both the hands. There were no other lesions anywhere else on the body. Mucous lesions were absent.

Investigations: Urine: NAD. Stool: NAD. WBC – T. C. 5600/cmm Polymorphs: 65%. Lymphocytes: 27%. Monocytes: 2%. Eosiqnophils: 5% Hb: 13.0 Gms%. Screening chest: NAD.

Biposy: Showed features consistent with the diagnosis of epidermo dysplasia verruciformis.

DISCUSSION

According to Eller and Eller, epidermo dysplasia verruciformis has been considered an epithelial nevus, a hereditary type of dystropy, or an intensified form of verruca. It can be distinguished from verruca plana and other types of verrucous lesions by its history of early onset and parental consanguity, and its persistence throughout life.

According to Lutze, epidermo dysplasia verruciformis is only a generalised eruption of verruca plana which assumes a particular aspect in accordance with the terrain on which it develops. It may be possible that epidermo dysplasia verruciformis is a dermatosis in which verruca planae occur on a skin with certain anatomic or functional abnormalities.

There are many points in common between wart (virus) and epidermo dysplasia verruciformis:— (i) Familial occurrence determined by a special predisposition which is fundamental in infection with warts. (ii) Development of Bowen type lesions in epidermo dysplasia veruciformis may suggest an etiologic relationship between viral infection and Bowen's epithelioma. (iii) By electron-microscopy, the virus of epidermo-dysplasia verruciformis is found to be identical with or closely related to that of verruca vulgaris.

Since both patients had maculo-papular lesions with satellite papules, and histopathology of biopsy specimens from both the patients was identical with that of wart (verruca), we feel that epidermo-dysplasia verruciformis may be due to viral infection related probably to a generalised type of verrucosus.

SUMMARY

Two cases of epidermo-dysplasia verruciformis with not only papular lesions, but also with macular lesions are reported. In both the case reports, family history was negative. In one case, lesions were present for 2 years, and in the other, lesions were present for one year.

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