DERMATO-VENEREOLOGICAL QUIZ

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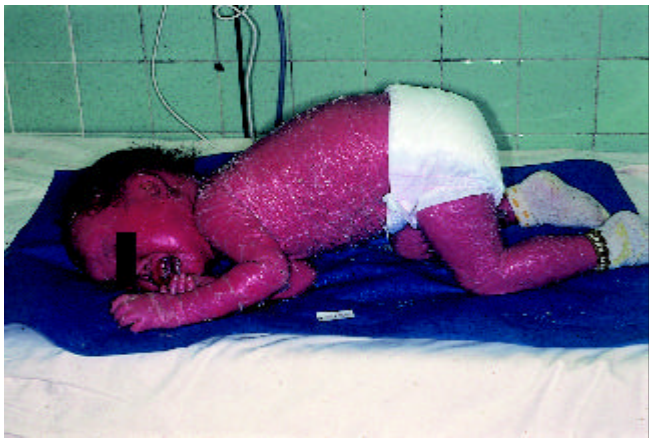

Figure 1.1

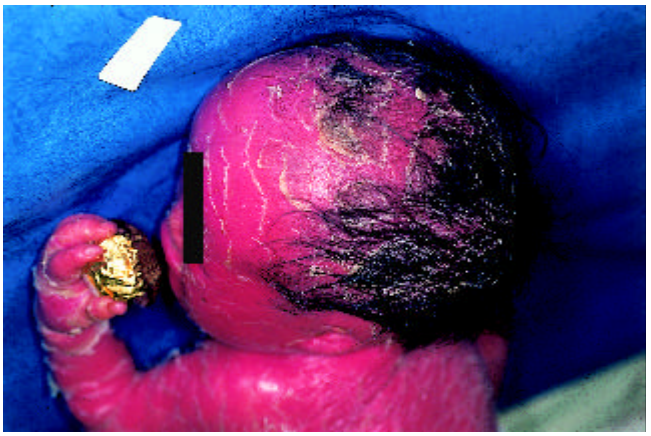

Figure 1.2

Question 1

This prematurely born infant presented with this severe skin disorder. She was also found to be of low birth-weight. There was no similar condition in the family or any history of severe skin diseases. There was no consanguinity of the parental marriage. She was subsequently treated and enjoyed reasonable good control of her skin condition.

1. *What severe skin condition did this baby suffer from?*
2. *What are the clinical features illustrated?*
3. *What was the medication that had been used to control her condition?*

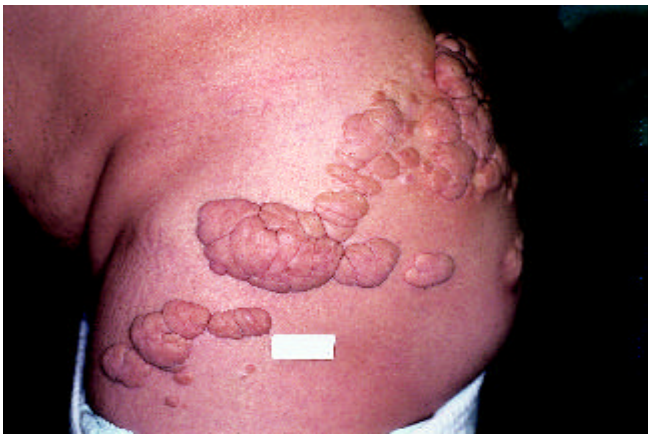

Figure 2.1

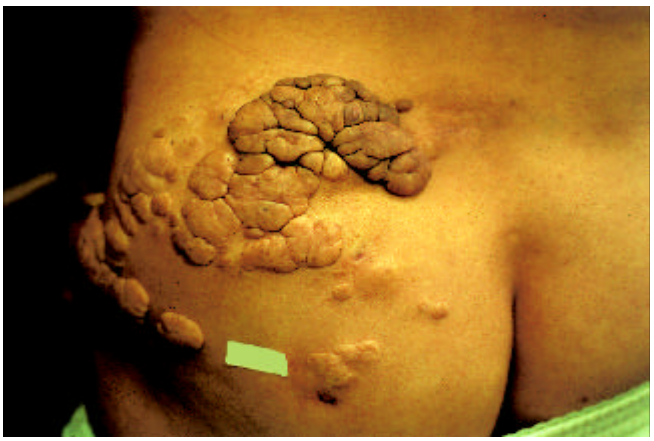

Figure 2.2

Question 2

This 40-year-old male patient presented to our dermatological clinic with this unilateral, soft, fleshy cerebriform yellowish plaque over his buttock and upper thigh. He had these lesions since early childhood and the condition had remained static. He was clinically asymptomatic and on physical examination, there was no other abnormality found. There was no significant family history.

1. *What is the dermatological diagnosis?*
2. *What are the main histological features?*
3. *Which rare genodermatosis needs to be differentiated from this condition?*

(answers on page 16)

Answers to Dermato-venereological Quiz on page 42

Answer (Question 1)

1. This condition is known as Harlequin fetus.

2. The clinical features illustrated are generalized erythroderma, severe skin fissurings, ectropion of the eyes, eclabium of the lips and deformed ears. The baby born was usually of low birth weight, prematurity and died shortly after birth.

3. Systemic retinoids like etretinate had been used successfully to control the condition. Our illustrated case had been treated with oral acitretin and liberal topical emollients. The side effects of systemic retinoids were regularly monitored.

   Harlequin fetus is a very severe and rare neonatal condition of the skin. Its mode of inheritance is thought to be autosomal recessive. The term Harlequin referred to the diamond-like costume of the fetal skin as a result of the severe fissures and hyperkeratosis. It is usually a fatal condition but systemic retinoids may control the condition. Nowadays, authorities believe that Harlequin fetus is a heterogeneous skin disorder. The baby if survive may evolve into an ichthyosis resembling congenital ichthyosiform erythroderma. Molecular studies suggested that the features of the condition may be due to a biochemical block in the conversion of filaggrin from profilaggrin. Hyperproliferative keratin 6 and 16 had also been identified in the abnormal keratinocytes.

Answer (Question 2)

1. The diagnosis is Naevus lipomatosis superficialis (classical type as described by Hoffman and Zurhelle).

2. Histologically, there is a proliferation of adipose tissues in the dermis of the skin. The adipose tissues surround the blood vessels and the vessels may rise up from the subcutaneous layer and spread out to form the subpapillary plexus.

3. Naevus lipomatosis superficialis should be distinguished from focal dermal hypoplasia. The latter condition is an important genodermatosis which can be associated with widespread ocular, dental and skeletal dysplasia.

   Naevus lipomatosis superficialis is a benign connective tissue naevus of the adipose tissues. It is divided into two types: the classical type is the Hoffman Zurhelle variant which is characterized by unilateral multiple skin colored cerebriform plaque, mostly occurred over the buttock and upper thigh of the patient. The second type is the solitary form in which the lesion appears as domed shaped, single papule. The site of predilection is the arm, axilla, head and neck other than the lower trunk. Both conditions are asymptomatic. No specific treatment is required except surgical excision for cosmetic reasons.