Diseases of the skin

Keratosis follicularis, dyskeratosis congenita, white sponge nevus

Dept.of Oral Pathology & Microbiology

Learning Objectives

At the end of the lecture student should be describe

- Clinical features, Oral manifestations,& Histopathological features Acanthosis Nigricans, Ehlers-danlos syndrome, Focal dermal hypoplasia syndrome (Goltzs syndrome).

ACANTHOSIS NIGRICANS

Acanthosis nigricans is a rare cutaneous disease, which usually affects the flexural surfaces of skin and it has an oral mucosal component.

3 TYPES:

1.Benign type-

- Present at birth or during puberty.
- Inherited as autosomal dominant trait
- Never associated with internal malignancy.

2.Malignant type-

- Usually develops after the age of 40 years
- Invariably associated with internal malignancies like adenocarcinomas or lymphomas of the GI tract.

3.Pseudoacanthosis type-

- Most common form of acanthosis nigrican
- Lesions develop around body creases as a result of obesity.

CLINICAL FEATURES

- Pigmented, papillary or verrucous growths over the axilla, palms, soles & face, etc.
- Dorsum of the tongue exhibits hypertrophy of the filiform papilla with development of a shaggy appearance.
- Tongue also presents some areas of papillomatous growth.

- Lip is grossely enlarged (mainly the upper lip) and its surface is dotted with small papillo-matous nodules especially at the commissure.
- Givgiva also exhibits hyperplastic changes and the lesions are clinically similar to that of the fibromatosis gingivae.

- Buccal mucosa and palate show a velvety white appearance with papillary projections in some areas.

Histopathology

- Thickening of the epithelium with marked acanthosis.
- Hyperkeratinization of the surface epithelium.
- Malignant form of the disease shows marked epithelial hyperplasia and acanthosis.

EHLERS-DANLOS SYNDROME

- A group of hereditary disorders characterized by defective collagen synthesis in various body organs.

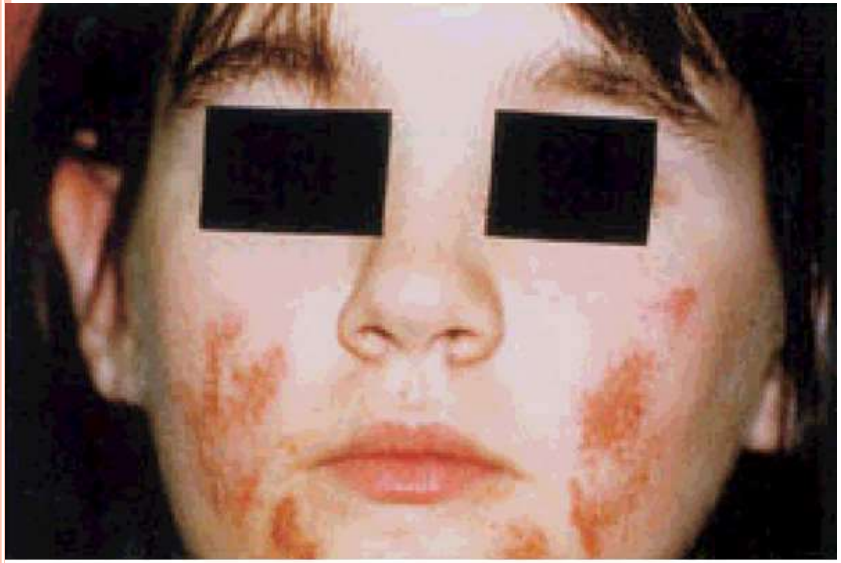
Hyper mobility of the joints & increased laxity of the skin, - **"Rubber man"**

Excessive bruising tendency & defective wound healing due to increased fragility of the skin & blood vessels.

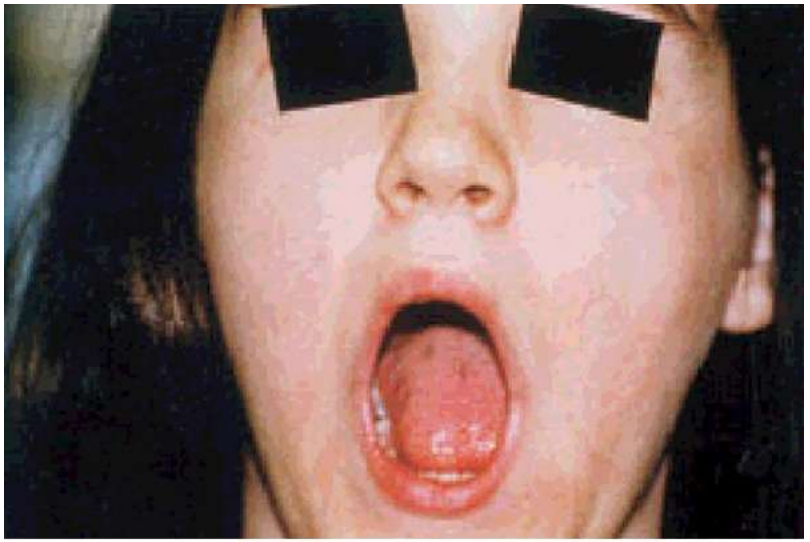
Oral Manifestations

- Increased fragility of the oral mucosa. Retarded wound healing
- Bleeding from the gingiva & oral mucosa ,Mobility of teeth.
- Enamel hypoplasia
- Loss of normal scalloping of the dentino-enamel junctions of the tooth

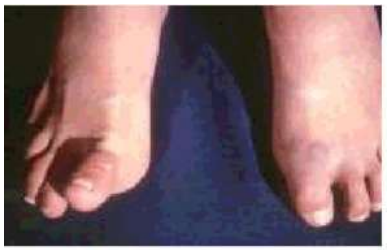
- Large pulp stones in the teeth & formation of irregular type of dentin
- Hypermobility of the temporomandibular joint.
- Difficulty in suturing the oral wounds.



Ehlers-Danlos Syndrome
19-41 A. Ehlers-Danlos Syndrome. Bilateral bleeding in the cheeks following episodes of luxation. Pg 1148



Ehlers-Danlos Syndrome
Figure 19-41 B. Ehlers-Danlos Syndrome. Luxation of the temporomandibular joint in a 12-year-old patient. p. 1148



A

B

C

Figure 19-42 A. Patient with Ehlers-Danlos syndrome.
Note the abnormal ability to elevate the right toe. Pg 1149

Figure 19-42 B. Girl with Ehlers-Danlos syndrome.
Dorsiflexion of all the fingers is easy and absolutely painless. Pg 1149

Figure 19-42 C. Patient with Ehlers-Danlos Syndrome mitis.
Joint hypermobility is less intense than with other conditions. (Courtesy of Enrico Ceccolini, MD). Pg

- **Differential Diagnosis**
- Hereditary benign intraepithelial dyskeratosis.
- Lichen planus.
- Hyperplastic candidiasis.
- Leukoplakia.
- Verrucous carcinoma.

Histopathology

- White sponge nevus microscopically presents mild to moderate hyper-parakeratosis of the epithelium with acanthosis and intercellular edema.

- There may be presence of some "vacuolated" cells in the spinus cell layer having pyknotic nuclei.

Focal dermal hypoplasia syndrome (Goltzs syndrome)

- Genetic disorder characterized by distinctive skin abnormalities and variety of defects affecting eyes , teeth, skeletal , urinary , gstrointestinal , cardiovascular and central nervous system.
- The skin lesions appear to evoke as accumulations of fat rather than hypoplasia of the dermis.

- Mnemoic FOCAL
- Female sex
- Osteopsthica striatia
- Coloboma
- Lobster claw deformity
- Absent ectodermis, mespdermis, neurodermis elements.
- Affected individuals are recognized at birth or prenatally.
- Also known as **GOLTZ** syndrome.

Etiology:

- X-linked dominant inheritance pattren, common findings of syndactyly, oligodactyly and polydactyly.
- Clinical features:
- Characterized by relative focal absence of dermis associated with herniation of subcutaneous fat into the defects
- Skin atropy, streaky pigmentation & telangiectasia
- Multiple papillomas of the mucosa.

Oral manifestations

- Papillomas of the lips is a striking feature.
- Teeth are commonly defective in size, shape or structure
- Microdontia is a common finding.
- Cleft lip or palate may also be present.

Histologic features:

- Attenuation of dermal collagen fibres with partial to complete absence of significant portions of dermis,
- Change in appearance of adipose in the cells in the dermis.
- If the accumulation of adipocytes is pronounced it may cause the apparent herniation of subcutaneous through the thinned skin.

SUMMARY

- Introduction, Clinical features, Oral manifestations,& Histopathological features of
- Acanthosis Nigricans
- Ehlers-danlos syndrome
- Focal dermal hypoplasia syndrome (Goltzs syndrome)

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Thank You