CASE REPORT

ACQUIRED CUTIS LAXA WITH RECURRENT URTICARIA

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ABSTRACT: A 30 year old male patient presented with progressive laxity and wrinkling of skin over the face for past 10 years, patient also gives history of recurrent urticaria since 12 years. Skin biopsy using Verhoff Van Gieson stain suggestive of cutis laxa. We are reporting a rare case of acquired cutis laxa with recurrent urticaria.
KEYWORDS: Cutis laxa, Urticaria.

INTRODUCTION: Cutis laxa is a heterogeneous group of disorder characterized by wide spread laxity of skin resulting in a premature aged appearance.(1) It may be congenital or acquired, pathology being deficiency of elastic fibres. Organs other than skin may be involved, resulting in emphysema, aortic aneurysm, intestinal diverticulosis and multiple hernias.(2)

CASE REPORT: A 30 yr old male patient presented to our OPD with complaints of loose and wrinkling of skin, for the past 10years which initially started over the face gradually progressed to involve neck and arms. Associated with this patient gives history of recurrent urticaria for the past 12 years, resulting in formation of red raised lesions that would persist for few hours and subside on taking medications. Urticarial lesions aggravated with intake of egg, non- vegetarian and spicy food. No history of dental anomalies/Upper Respiratory Tract Infections. Patient had no history of dyspnoea, chest pain, palpitations, and joint pain. There was no history of similar complaints in family and there is no parental consanguinity.

On examination the patient had an 'old man' appearance. There were folds of lax and wrinkled skin over the face, neck, around eyelids and arms. Sagging of ear lobules was noted. Skin could be easily stretched and showed delayed recoiling. Urticarial wheals were noticed. Ophthalmic examination showed Blepherochalasis and entropion of upper eyelid, folds of loose hanging skin over the upper eyelid. There were no hernias, no hyper-extensibility of joints. Oral cavity, genitalia, hair, nail and systemic examination was normal.

Blood sugars, liver function tests, renal function tests, lipid profile, urine analysis were within normal limits. ANA was negative, no thyroid abnormality. Chest x-ray (PA), ECG, Echocardiography was normal. Serum IgE and AEC levels were elevated.

Skin biopsy with special stain Verhoff Van Gieson showed very few attenuated fragments of elastic fibres in dermis. There was no evidence of inflammation, morphological features was consistent with cutis laxa.

DISCUSSION: Cutis laxa (synonym: dermatochalasia, dermatomegaly) (CL) is a rare connective tissue disorder caused by defects in the elastic fibre network and can affect multiple tissues, predominantly the skin.(3) It is a heterogenous condition, which may be inherited as a dominant,

recessive or x-linked recessive disease, or it may be acquired. The face and neck are often affected which produces a 'bloodhound' appearance of premature ageing.[4] The acquired form is even rarer than the inherited form and is often associated with some form of preceding or accompanying cutaneous eruptions like eczema, urticaria, erythema multiforme, Sweet's syndrome or multiple myeloma.[5] Cutis laxa may be caused by mutation in genes ELN3-, ATP6VOA24-, ATP7A5-, BLN46- and PYCR8-.[6]

Cutis laxa syndromes and clinical subtypes:

Subtype/OMIM	Clinical features	Inheritance
1. Autosomal dominant (ADCL)/123700[7]	Late onset. Primary cutaneous lesions. Rare vascular complications. Some clinical variability	AD
2. X linked recessive/ (304150)[8]	Congenital skin laxity-often distal characteristic facies. Hoarse cry- vocal cord redundancy. Brittle hair, joint laxity-mild occipital exostosis develops after second year of age. Skeletal abnormalities, failure to thrive, diarrhoea and gastro intestinal diverticula.	XLR
3. Autosomal recessive cutis laxa (ARCL) (1219100)	Congenital, severe loose hanging redundant skin. Early lethality, Emphysema, Cor Pulmonale, Hernias and cardiovascular abnormalities.	AR
4. ARCL II Debre type/(219200)	Clinical variability Microcephaly, developmental delay, hypotonia and progressive neurological abnormalities.	AR
5. ARCL with progeroid features(612940)	Progeroid appearance Osteopenia, connective tissue weakness and finger contracture.	AR
6. Genodermia Osteodysplasia (GO)	Progeroid features. Malar mandibular	AR

(2B1070)[8]	hypoplasia. Joint laxity. Short stature.	
7. Cantu syndrome (114620)	Cardiac abnormalities. Osteochondrodysplasia.	AR
8. Wrinkly skin syndrome(WSS) (278250)	Mild microcephaly. Mental retardation.	AR
9. Latent transforming growth factor binding protein 4 deficiency	Cutis laxa. Growth delay. Gastro intestinal and genito urinary abnormalities	AR
10. Transaldolase deficiency	Congenital cutis laxa. Hydrops fetalis. Dystrophic features.	AR

Review of the world literature suggests only around 200 cases being reported and importantly only a striking minority of these patients has a known underlying molecular cause. Psychological and emotional aspects of patients to be addressed. Plastic surgery according to patient's will. Dapsone is reported to control acute swelling, which may support a role for neutrophil elastase. Botulinum toxin has been used to improve facial cosmesis.[6]

Our patient was referred to Department of Plastic Surgery for facial uplift procedure and he was treated with Tab. Levocetirizine 5mg and moisturizers.

We are reporting a rare case of acquired cutis laxa due to urticaria. Urticara being the etiology in our case.

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8. Online 'Mendelian Inheritance in Man' (OMIM) Cutis Laxa, X-Linked -304150.

Fig. 1: Photograph of the patient, when he was 20 years old

Fig. 2: Sagging of Ear lobule

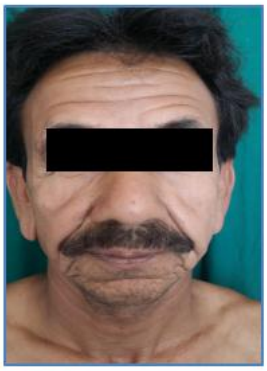

Fig. 3: Old man appearance, patient is presently 30 years old

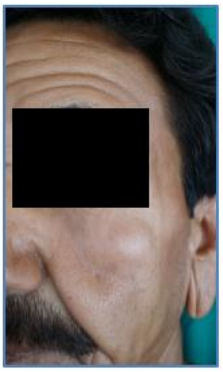

Fig. 4: Blepherochalasis

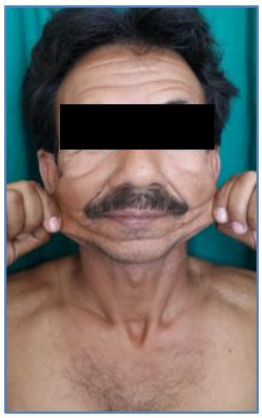

Fig. 5: Easy stretchability of skin with delayed elastic recoiling

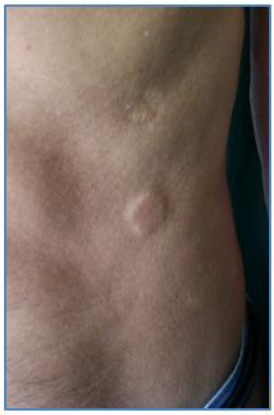

Fig. 6: Urticarial Wheal

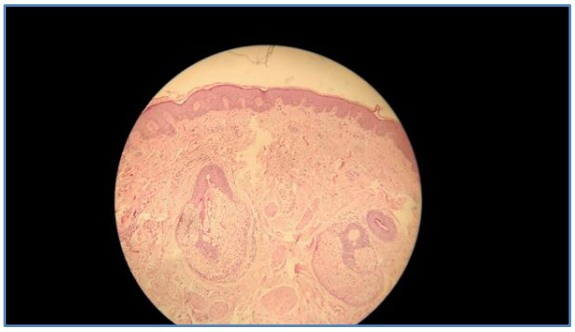

Fig. 7: Histopathology with 'Verhoff Van Gieson stain' showing very few attenuated fragments of Elastic fibres

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