Lepromatous leprosy presenting as generalized cutis laxa – a rare presentation

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Introduction

Leprosy is a common disease in the tropics. It has a diverse clinical presentation. Lepromatous leprosy is a multi system disease that develops in individuals who are unable to mount a cell mediated immune reaction against *Mycobacterium leprae*. There is massive multiplication of bacilli that infiltrate predominantly the skin and nerves. Lepromatous leprosy also affects reticuloendothelial system, upper respiratory tract, eyes, testis and adrenals.

Clinically, it presents with multiple symmetrical erythematous nodules, plaques or skin infiltration and can result in severe functional disability. Atypical presentations of leprosy is not uncommon. Rarely cases have been reported presenting with peripheral hypoesthesia with imperceptible or apparently no skin manifestations. Cases have also been reported as panniculitis. Atypical forms are a diagnostic challenge and a source of infection. Lepromatous leprosy still remains a major clinical challenge.

Cutis laxa is a congenital or acquired disorder of connective tissues in which skin becomes inelastic and hangs loosely in folds. Histologically it shows elastolysis. The patients with cutis laxa appear prematurely aged. Acquired cutis laxa has been reported following urticaria, angioedema, multiple myeloma, drug hyper-sensitivity reactions, amyloidosis, sarcoidosis and inflammatory conditions of the skin like SLE and RA. Localized anetoderma has been seen in increased frequency following lepromatous leprosy. Acquired cutis laxa is thought to be a variant of anetoderma.

We report a case with peripheral neuropathy and progressive generalized cutis laxa who was ultimately found to have lepromatous leprosy. He did not have any clinical evidence other than neuropathy to suggest leprosy. We report this case to emphasize that leprosy can present in atypical forms making clinical diagnosis difficult. Even though localized anetoderma had been reported following leprosy there were no reports of generalized cutis laxa reported following leprosy.

Case report

50-year old male was referred from the surgical ward for assessment of chronic leg ulcers. He had a history of recurrent painless leg ulcers for six months duration. He also complained of progressive numbness in both hands and feet. During past one year he developed progressive laxity of the skin. The lax skin was first noticed on face and it gradually spread to the rest of the body. There was no history of hypoesthetic skin patches, nodules or plaques. He did not have a history of loss of appetite or weight. No family history of similar disease.

On examination the patient had generalized lax skin with loss of elasticity. There was marked loss of elastic recoil. Blepharochalasis was noted bilaterally and it was marked on right side. He did not have hypoesthetic skin patches or inflammatory nodules. There were no thickened nerves. He had enlarged firm lymphnodes in axillae. There was no palor or hepatospleenomegaly. Neuropathic ulcers were noted in both feet.

Granulomatous slack skin syndrome, multiple myeloma or atypical presentation of lepromatous leprosy were suspected.

His full blood count showed Hb 10.3 g/dl, white cell count was 10200/mm^3 with N 55% and L 40%. Platelet count was 413 × 10^3. Blood picture showed normocytic normochromic picture. There was no evidence of malignancy or nutritional deficiency. Initial ESR was 102 mm/hr and repeat ESR was 42 mm/hr. Fasting blood sugar was 89 mg/dl. His liver and renal function tests were normal. Serum protein – electroporesis, urine Bence Jones proteins and x-rays did not reveal evidence of multiple myeloma. Skin biopsy showed evidence of lepromatous leprosy. Staining for Acid fast bacilli showed bundles of bacilli within the macrophages. Skin biopsy did not show evidence of malignancy. Slit skin smear for acid fast bacilli came positive. Lymphnode biopsy was positive for acid fast bacilli. The patient was started on multibacillary antileprosy treatment. His leg ulcers were treated in the Surgical Unit.

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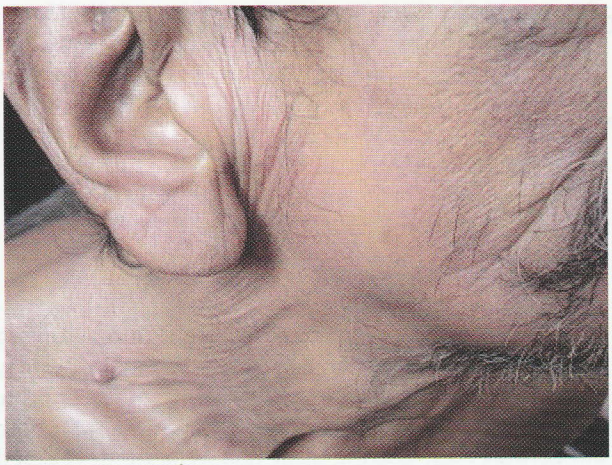
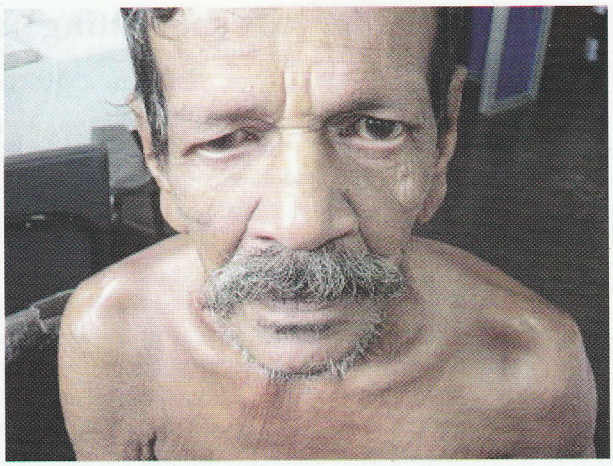
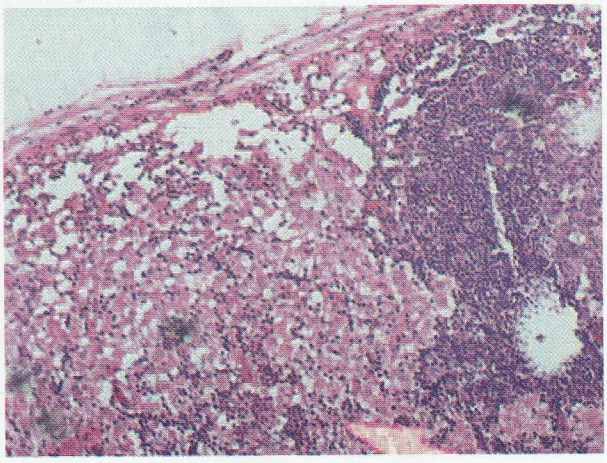


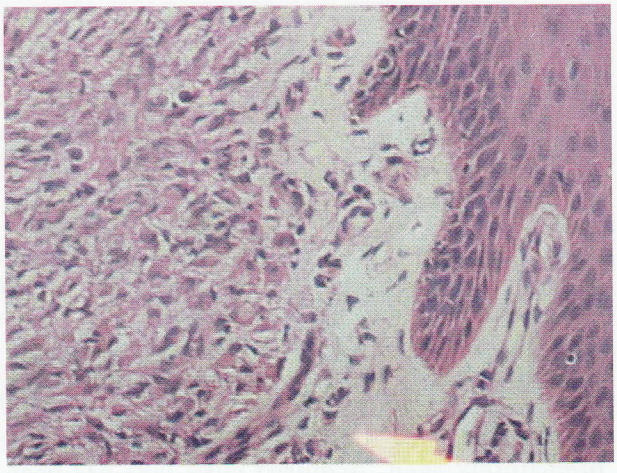
Lax pendulous skin



Blepharochalasis



Lymph node biopsy showing foamy macrophages



Skin biopsy showing Grenz zone and the dermis filled with foamy macrophages

Discussion

Cutis laxa is a rare inherited or acquired disorder of elastic tissue. Clinical presentation and mode of inheritance show considerable heterogeneity. The precise etiology is unknown but thought to be due to abnormal elastic metabolism resulting in markedly reduced elastin content or abnormal morphology. This causes degenerative changes in elastin fibers resulting in loose pendulous skin. Underline cause is variable. Abnormal Cu + metabolism, reduced serum elastase inhibitors, increased elastase activity, post inflammatory elastolysis and immune mediated mechanisms are reported in literature. Cutis laxa secondary to inflammatory disorders like SLE, RA, urticaria or drug hypersensitivity reactions has been reported. Inflammatory cells, neutrophills, macrophages and their mediators are thought to damage elastic tissues. In lepromatous leprosy massive infiltration of macrophages could be the reason.

Granulomatous slack skin syndrome, Marfans syndrome and mid dermal elastolysis should be considered as differential diagnosis. Our patient was a case of lepromatous leprosy presenting as cutis laxa and peripheral neuropathy. He did not have clinical evidence of leprosy. We suspected leprosy due to its common association with peripheral neuropathy. Localised anetoderma associated with leprosy had been reported in the literature but so far generalized cutis laxa has not been reported. We report this case due to its rare association. It is important to have knowledge of atypical and rare association of acquired cutis laxa.

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