

A Case of Generalized Lupus Vulgaris Leading to Joint Mutilation

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Abstract

Observations: Lupus vulgaris is a chronic progressive granulomatous bacterial disease and the most common cutaneous tuberculosis. It arises in individuals who were previously sensitized and have moderate immunity. Lesions usually begin on the nose and cheeks and slowly progress towards neighbouring sites in the disease that usually involves head and neck. In this paper, a 51-year-old subject who has many lupus vulgaris plaques on dorsum of the left hand, left arm and left gluteal region is presented. A contracture deformity had developed on the area of lupus plaques on 4. and 5. fingers of the left hand. The patient who had complaints for approximately 10 years was diagnosed as pulmonary and cutaneous tuberculosis as the result of clinical and laboratory examinations. The patient was treated with quartet antituberculosis treatment. The case was decided to be presented due to its atypical localisation and leading deformity.

Introduction

Lupus vulgaris is a granulomatous bacterial disease and the most common cutaneous tuberculosis [1]. Its incidence among all skin diseases is 0,37%. Lesions usually begin on the nose and cheeks and slowly progress to neighbouring sites in the disease that usually involves head and neck. Lupus vulgaris arises in individuals who were previously sensitized to the bacillus and developed moderate immunity. Trunk is involved in disseminated form of lupus vulgaris [1, 2]. The case that had an atypical location and led deformity was decided to be presented.

Case Report

A 51-year-old female patient applied to our clinic with complaint of a lesion that began on her hand approximately 10 years ago and spread to her arm

and hip and did not cause pruritus. Lesion on her hand had caused deformity. She was given different therapies with diagnosis of contact dermatitis and tinea corporis by the doctors she applied. The patient who did not have night sweating, cough, sputum, weight loss, stated that there were no tuberculosis patients around her. The patient did not have a familial or personal history of tuberculosis, her family history and medical history were unproblematic.

The patient had two lymphadenopathies of which the largest one approximately 2 cm in size in both axillary regions. On her dermatologic examination, there were well demarcated papules and plaques with yellowish white crust and thin squam on dorsum of the left hand, on flexor, extensor and medial condyle sites of left forearm, of which the largest was 8x10 cm in left gluteal area and the smallest was 1x1 cm on flexor surface of left arm (Figure 1). Atrophic areas had developed on hand dorsum. There



Figure 1. Typical lupus vulgaris plaques and joint deformities

were contracture deformities on medial interphalangeal joints of 4. and 5. fingers of left hand. Apple-jelly reffe was found positive on diascopy examination (**Figure 2**). Routine biochemistry tests, urogram, hemogram, peripheral blood smear, CRP, tumor makers, hepatitis markers, chest graphy were normal. Bacillus was detected in 300 areas in one of the AARB (Acid-Alcohol Resistant Bacteria) examinations in sputum done on 3 sequential days



Figure 2. Apple-jelly appearance on diascopy examination

however remaining two were negative. Growth was found in sputum culture for tuberculosis. On computed chest tomography, two lymphadenopathies of which the biggest was 2 cm in size were found in bilateral axillary regions and linear atelectasis was found in both lungs. PPD (Purified Protein Derivative) test was measured as 30x30 mm positive (**Figure 3**). On her skin biopsy, granuloma comprised of severe chronic mononuclear inflammation and epitheloid histiocytes was seen and interpreted as lupus vulgaris (**Figure 4**). The patient was diagnosed as pulmonary and cutaneous tuberculosis by the pulmonologists and isoniazid 300 mg 1x1, rifampicin 300 mg 1x2, ethambutol 500 mg 1x2 and pyrazinamide 500 mg 1x3 treatment was started. Plaques were seen to improve significantly and no new lesions were observed on her control one month later. The patient whose treatment is planned to continue for 12 months goes on her treatment and regularly comes to her controls.

Discussion

Lupus vulgaris arises from hematogenous, lymphatic or contiguous spread from a tuberculosis focus in anywhere of the body. It can rarely develop by exogenous inoculation or following BCG vaccination. It is most commonly seen in head and neck. It starts from the nose and the cheeks and slowly progresses to the neighbouring sites. It may lead cicatricial alopecia if it is in scalp. Only very small proportion of the lesions are seen on extremities. Involvement of the trunk is quite rare except for disseminated lupus vulgaris patients [1, 2, 5]. Lesions were located on gluteal area, arm and hands and were multiple in our case. It may last for long years unless it is treated as it is a quite chronic illness. Complaints of our



Figure 3. PPD test 30x30 mm

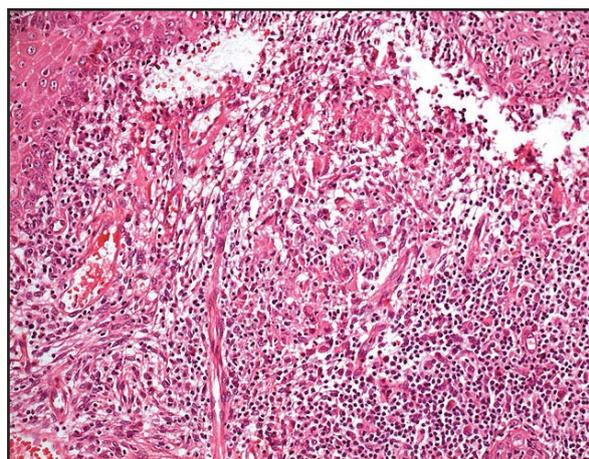


Figure 4. Granuloma in dermis comprised of severe chronic mononuclear inflammation and epithelioid histiocytes (HEx200)

subject began 10 years ago and did not heal as she was not treated, even new lesions had developed. The disease may lead permanent deformities unless it is treated. Contractions limiting the motion of the joint, mutilations in the cartilage of the face, ectropion, microstomy interfering with speech and feeding are the sequela of the disease [2, 5]. In our case, joint deformities and mutilation had developed in the fingers of the hand. Squamous cell carcinoma, basal cell carcinoma and plasmacytoid lymphoma may develop on lupus vulgaris scarring [5]. Lupus vulgaris is generally asymptomatic. Our subject did not have any prominent symptoms.

Lupus vulgaris is seen in individuals who were previously sensitized to the bacillus and developed moderate immunity. Lupus vulgaris cases are also present that develop from scrofuloderma scarring or following exogenous reinfection. It is 2-3 folds more frequent among females [1, 2, 3]. Our case was an immunocompetent female.

Lesions are characterized with red, soft, pin-head macules and papules. These papules that are called as lupom show mat yellowish apple-jelly appearance if they are touched with a lame (diascopy). This is pathognomonic for lupus vulgaris [3]. Diascopy was positive in our case. Lesions can easily be perforated with a stile due to the presence of underlying tissue harm. Plaques are formed by slowly peripheral enlargement and joining of the lupoms. Plaque, hypertrophic, ulcerative and vegetative forms are prominent clinical types [3, 4]. Lesions were hypertrophic big plaques expanding

to the periphery and atrophic in patches in our case.

Tuberculoid granuloma prominent especially in epidermis and little caseous necrosis is accompanied is seen in its histopathology. Secondary changes like thinning and atrophy in epidermis or extreme hyperkeratosis and acantosis or pseudo epitheliomatous hyperplasia may be seen [1, 2]. Skin biopsy was consistent with lupus vulgaris in our case.

Sarcoidosis, lymphocytoma, discoid lupus erythematosus, tertiary syphilis, lepra, deep mycotic infections, lupoid leishmaniasis should be taken into consideration in differential diagnosis [1, 2, 4]. Diagnosis of typical lupus vulgaris plaques is unproblematic.

Diagnosis can be made with the history of tuberculosis, clinical appearance, PPD, EZN (*Ehrlich Ziehl-Neelsen*) staining, histopathology, chest graphy, bacteriologic examination (i.e. seeking for bacillus in sputum on 3 sequential days), mycobacteriologic culture, molecular methods sensitive to nucleic acide (PCR). Making a diagnosis can sometimes be difficult due to different clinical appearances, failure to show mycobacteria in skin biopsies due to little bacillus count [4, 5]. Lesions' being present for a long time and apple-jelly appearance on diascopy are useful criteria for diagnosis [1]. In our case, diascopy was positive, acido resistant bacillus was detected in EZN staining of sputum and sputum culture was positive.

According to recommendations of World Health Organization, usually quartet antituberculosis drugs are given during the first two months: isoniazid (5 mg/kg), rifampicin (10 mg/kg), ethambutol (15 mg/kg) and pyrazinamide (25 mg/kg). Treatment should be continued with two drugs (isoniazid and rifampicin) for the following 4-6 months [3]. In our case, lesions significantly became thinner and thick squams decreased and new lesions did not develop with quartet therapy for the first 2 months and dual therapy for the following months.

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