

CASE REPORT

Two sarcoidosis cases with lupus pernio and their clinical follow up

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Abstract

Lupus pernio is a characteristic cutaneous involvement forms of sarcoidosis. It can be accompanied by bone cysts, upper respiratory tract involvement and pulmonary fibrosis. It is a chronic form of sarcoidosis and it is one of the poor prognostic factors. In this case report two patients with lupus pernio were presented. In the first case with stage two sarcoidosis there was no other system involvement, whereas in the second case there was bone cysts, upper respiratory tract involvement, and also clubbing and hypopigmented skin lesions which are rare findings of sarcoidosis. In both of cases there was a chronic course and one of them was treated for three years and the other was under treatment for four years.

Key words: Sarcoidosis, lupus pernio, clubbing, hypopigmented area

Lupus pernio bulunan iki sarkoidosis olgusu ve klinik takipleri

Lupus pernio, sarkoidosisin karakteristik cilt tutulum formuna verilen isimdir. Buna kemik kistleri,

üst solunum yolu tutulumu ve kistik fibrozis eşlik edebilir. Sarkoidosisin kronik formudur ve kötü prognostik faktörlerinden biridir. Bu olgu sunumunda lupus pernio bulunan iki olgumuzu sunuyoruz. İlk olgumuz, evre 2 sarkoidosis olup diğer sistemlerde tutulum yoktu. İkinci olgumuzda ise kemik kistleri, üst solunum yolu tutulumu ve sarkoidosisin nadir bulgular olan clubbing, ciltte hipopigmente lezyonlar bulunmaktaydı. İki olguda kronik seyirli olup biri 3 yıl tedavi görmüş, diğeri ise 4 yıldır tedavi altındaydı.

Anahtar kelimeler: Sarcoidosis, lupus pernio, clubbing, hipopigmente alanlar

Sarcoidosis is a disease with unknown etiology involving many organ systems and characterized by non-caseating epithelioid granulomatosis. Skin involvement is present in 25% of cases. (1,2)

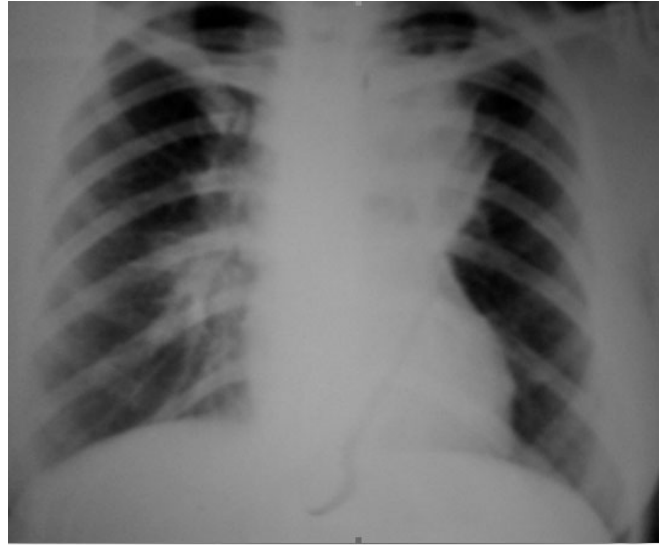
Lupus pernio is a finding of chronic sarcoidosis. Clinically lupus pernio was first described by Besnier in 1889 and in 1892 French investigators defined it as a lesion characterized with typical sarcoidosis histology showing epithelioid cell infiltration(3). It is more common in African Americans and females.(4,5). It begins as pink-red or violet colored infiltrations on nose, cheeks, forehead and lips and when it involves the respiratory tract it may infiltrate through pharynx and larynx mucosa. It is commonly associated with bone cysts and pulmonary fibrosis. Spontaneous remission is rare (6). It is thought to be poor prognostic finding of sar-

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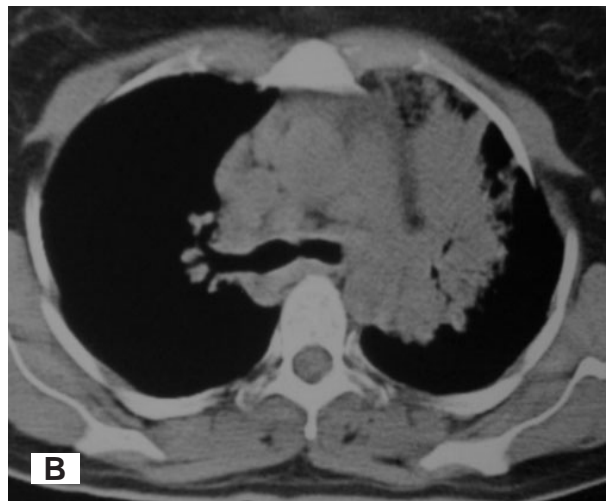
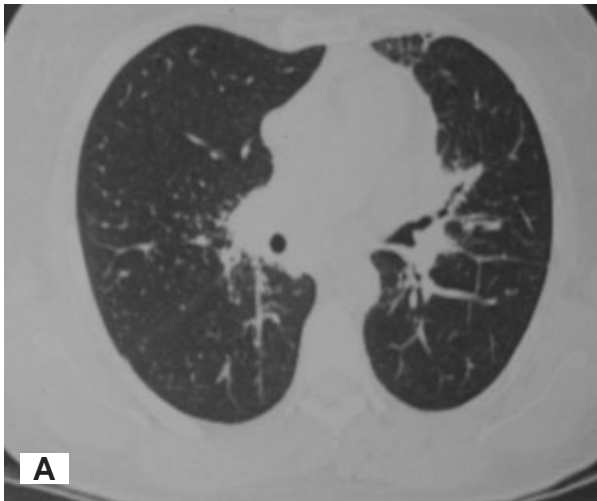
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Picture 1: Red, purplish lesion on face and telangiectatic lesions on cheeks and forehead.



Picture 2: Pretreatment chest radiograph of case I.



Picture 3a-b: Pretreatment thorax CT of case I.



Picture 4: PA chest radiograph of case I in the sixth month of therapy.

coidosis.

Case I:

A 43 years old, house-wife, admitted to our clinic with symptoms of fatigue, and signs of red and purple skin lesions on cheek and nose persisting for 6 months and non-productive cough, sometimes coming with attacks, persisting for 3 months. She has never smoked and she has been using insulin due to type II diabetes mellitus for 3 years.

In her physical examination blood pressure was 110/70 mmHg, pulse rate was 80 beats/min rhythmic, respiratory rate was 14/min and body temperature was 36.7°C. Other than the red-purple painless nodular lesions on the face, cheeks and forehead with telangiectasis over it (picture 1), her physical examination, was normal.

Her laboratory evaluation revealed erythrocyte sedimentation rate of 40mm/h, leucocytes level of 6.4

/mm³, hemoglobin level of 11.5gr/dl and hemotocrit level of 35.5. Routine biochemical analysis were in normal levels and tuberculosis skin test was negative. Her blood angiotension converting enzyme (ACE) level was 65 U/L and the calcium levels in blood and 24 hours urine were normal.

Pulmonary function tests results were FEV1: 2.21 (93%), FVC: 2.66 (96%), FEV1/FVC: 83 % and DLCO:15.9mL/mmHg/min (69%), DLCO/VA: 4.10 mL/mmHg/min (76%) .

Her chest radiography revealed bilateral hilar enlargement, a non homogenous consolidation area with approximately 4x5 cm in diameter with specular extensions and borders that could not be separated from left arcus aorta (picture 2). The patient was considered as stage II.

Thorax computerized tomography (CT) demonstrated bilateral hilar and multiple mediastinal lymph nodes and a mass lesion with diameter of 4x5cm at the anterior segment left upper lobe (picture 3a,b). In her fiberoptic bronchoscopy stenosis and mucosal edema at the entrance of superior segment of left upper lobe and mucosal irregularities at the entrance of left upper lobe were observed. Histopathology of biopsy material obtained by forceps biopsy from entrance of left upper lobe was considered as granulomatous inflammation. Skin biopsy obtained from the skin lesions present on the face were reported as non caseating granulomatous inflammation including epithelioid histiocytes at large areas and multinuclear giant cells at some areas (consistent with sarcoidosis).

Galium 67 total body scanning, abdominal ultrasonography and technetium 99m myocardial perfusion scanning and ophthalmologic examinations were normal.

Due to the skin lesions causing disfigurement corticosteroid therapy was indicated and metil prednisolone 32mg/day (discontinued in 8 months by tapering the dose), chloroquine 500mg/day (for 2 weeks) and 250 mg/day (for 5.5 months) were initiated. Even though no change was observed in skin lesions, chest radiography of patient improved (picture 4). Because radiological progression was detected on chest radiograph of the patient obtained four months after the treatment stopped, corticosteroid was restarted. With retreatment the radiology of the patient improved and minimal changes were observed in skin lesions. She is still receiving corticosteroid 8 mg/day.

Case 2:

A forty seven years old male, housepainter was admitted to our clinic with the complaints of redness on face and painful swelling at his hands. He had no smoking history.

In physical examination blood pressure was 110/70 mmHg, pulse rate was 80 beats/min and rhythmic, respiratory rate was 16/min. Erythematous, painless, elevated lesions including telangiectatic and nodular areas

with undefined borders was present on the right zygomatic area and there were three hard, elevated nodular lesions with undefined borders on his chin (picture 5). Painless, mobile lymphadenopathies with diameters of 1x1 cm at right and left supraclavicular areas and diameter of 1x2 cm at right and left submandibular areas were palpated. Respiratory system examination was normal, but extremity examination revealed large depigmented areas at the flexor surface of both arms (picture 6) and swelling at third and fourth interphalangeal joints and clubbing at right hand fingers (picture 7).

In his laboratory findings erythrocyte sedimentation rate was 60 mm/hour and total blood count, routine blood biochemical evaluations were in normal ranges. His PPD test was 12mm and blood ACE level was 103U/L. Blood and 24 hours urine calcium level was normal. In spirometric evaluation FVC: 5.47 lt (104%), FEV1: 4.22 lt(100%), FEV1/FVC: 77%, DLCO: 33.7mL/mmHg/min (96%), and DLCO/VA was 96 mL/mmHg/min .

His chest radiography revealed bilateral hilar adenopathy (Picture 8). He was considered as stage I. His thorax



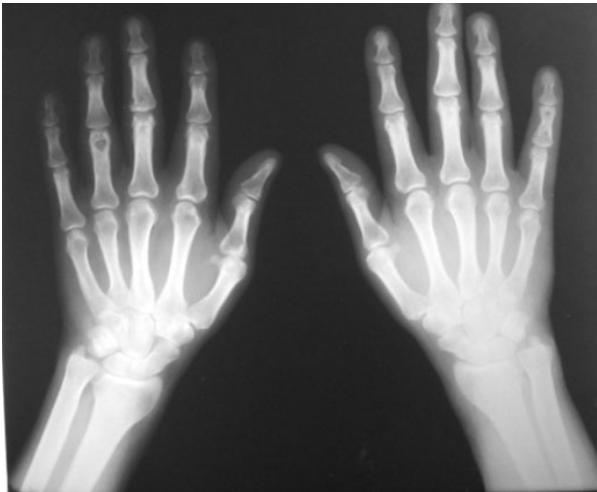
Picture 5-6: Case II: Erythematous and telangiectatic lesions on the cheeks of and large depigmented area on the forearm flexor surface.



Picture 7: Case II clubbing and swelling at the 3. and 4. interphalangeal joints.



Picture 8: Case II PA chest radiograph



Picture 9: Case II hand radiograph.

CT demonstrated bilateral hilar and multiple mediastinal lymph nodes, nodular lesions with diameter of millimeters at medial segment and with 1 cm diameter at superior segment of right lower lobe

There was no finding consistent with sarcoidosis in ophthalmologic examination whereas ear-nose and throat examination revealed granulomatous appearance at nasal septum and mucosa of right posterior mucosa of the nose.

Bilateral hand radiography revealed cystic areas at distal metaphysis of proximal phalanges of fourth digit, distal metaphysis of proximal phalanges of third digit, proximal metaphysis of middle digit of right hand and middle phalanges of fifth digit of left hand, and periarthritic soft tissue swelling at both hands (picture 9).

Skin biopsy was reported as granulomatous inflammation including multiple epithelioid histiocytes consisted of langhans type giant cells in the dermis (consistent

with sarcoidosis). Being considered as stage I, because of skin lesions causing disfigurement and extrapulmonary involvement corticosteroid was indicated. Metil prednisolone 32 mg/day and chloroquine 500mg/day were started. The steroid dose was continued by tapering the dose to 8 mg/day and chloroquine 500 mg/day was used for 2 weeks and was continued with a maintenance dose of 250 mg/day. Because ophthalmologic complications were developed in the maintenance dose of the chloroquine, it was discontinued in the second month. The patient is still receiving 8 mg/day corticosteroid. Radiologically no difference was observed in the pulmonary lesions and bone cysts.

Discussion:

In sarcoidosis cutaneous involvement is observed in 25% of cases. Of the skin lesions macule, papule, plaque, subcutaneous nodules, lupus pernio, lesions developing at areas of scars are the commonly seen pathologies. Rarely erythroderma, ulcerative, varicous, and hypopigmented lesions are also seen. Lupus pernio is a characteristic skin lesions of sarcoidosis. It may be observed in any stage of the disease. One of our cases was considered as stage I and the other was stage II.

Lupus pernio is often accompanied by bone cysts associated with sarcoidosis, upper respiratory tract involvement and pulmonary fibrosis. In our female case there was no involvement of other systems, but in male case beside skin involvement bone cysts, nodular lesions at the nasal septal mucosa were observed. In sarcoidosis bone cysts are usually more commonly seen in phalanx of hands than foot and it is accompanied by skin lesions over it (7). In our second case bone lesions were seen also at the phalanx, but skin lesions were absent. In this case also hypopigmented lesions, an unusual skin lesion of sarcoidosis, on the forearm of the patient and club-

bing, a very rare manifestation of sarcoidosis, were present.

Characteristically skin involvement of sarcoidosis is asymptomatic and is not life threatening. Even if it does not cause disfigurement, it is not present between the indications of systemic corticosteroid. The main therapy is local or systemic steroids. Also chloroquine, hydroxychloroquine, methotrexate, azathioprine, chlorambucil, cyclophosphamide and cycloserine are the drugs that may be used. Recently hopeful results with infliximab which is a monoclonal antigen inhibiting specifically TNF-alpha, in complicated cases was reported. In a study done with few cases demonstrated that infliximab was well tolerated and showed significant improvement in chronic sarcoidosis (8). Among alternative agents chloroquine, methotrexate and azathioprine are appropriate drugs that may be used in refractory cases or in cases whose steroid dose was needed to be reduced (9). With chloroquine in cutaneous sarcoidosis a response rate of nearly 35% was reported (10). In both of our cases corticosteroid was administered with chloroquine and both of them are still under treatment.

Lupus pernio is one of the poor prognostic factors of the sarcoidosis. In one series 22% of cases with cutaneous involvement radiological improvement of pulmonary lesions were observed, but none of them has lupus pernio (6). In our first case due to radiological improvement in the beginning treatment was stopped, but because radiologically the lesions reoccured, corticosteroid was restarted and radiological stability was achieved. In the second case eventhough clinical improvement was achieved with treatment, there was no changes in chest radiograph and bone cysts. This case has been receiving corticosteroid for four years.

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