Letter to the Editor
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A case of sarcoidosis diagnosed via skin involvement

Deri tutulumu ile tanı alan sarkoidoz olgusu

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To The Editor,

Sarcoidosis is a systemic granulomatous disease that is characterized by non-caseating granulomas. Approximately 20 to 30% of patients with systemic sarcoidosis have skin lesions of various morphology, leading to the designation "great imitator". Studies suggest that genetic, immunologic, and environmental factors interact together to cause sarcoidosis[1,2]. It primarily involves the lungs and lymphatic system as well as skin, bones, eyes, spleen, and parotid gland[3]. Cutaneous involvement may be the first manifestation of systemic disease, so patients with skin involvement should be investigated for systemic disease[2-4]. The diagnosis is made by radiological and clinical findings supported by histopathology, and by ruling out of other possible differential diagnoses[5,6]. Here we present a case presenting with skin findings and diagnosed as sarcoidosis as a result of histopathological and systemic research.

A 35-year old male patient presented to our outpatient unit with complaints of mildly pruritic reddish skin lesions of the face and scalp. The patient reported that the red colored skin lesions had first appeared 1 year ago on the face and scalp and he had used a number of different creams with partial improvement and re-occurrence of lesions. He also had swellings in the neck. In his dermatological examination showed elevated erythematous annular plaques, the greatest being 5-6 cm, in periorbital area, temporal area, left dorsolateral aspect of the nose, and in the cheek. These lesions were accompanied by occasional telangiectasias (Figures 1, 2). Similarly, erythematous plaques were also present in the neck and scalp. There were multiple mobile unpainful lymphadenopathy (LAP) approximately 1.5 to 2 cm in diameter, were present in the cervical and pre-auricular regions.

The family history, past medical history, and organ system examinations were unremarkable. Laboratory tests showed normal complete blood count and biochemistry values. Blood calcium levels were normal, angiotensin converting enzyme (ACE) level could not be studied. A punch biopsy was performed with pre-diagnoses of granuloma annulare, annular elastolytic giant cell granuloma, discoid lupus erythematosus, porokeratosis, actinic lichen, annular lichen, granuloma faciale, cutaneous metastasis, or sarcoidosis.

Chest x-ray and thoracic computed tomography imaging were performed, which showed bilateral hilar LAP and 4 to 5

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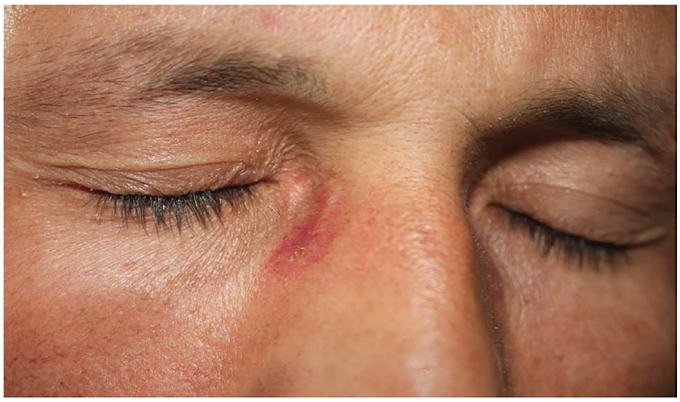

Figure 1. Erythematous plaque on the side of the nose

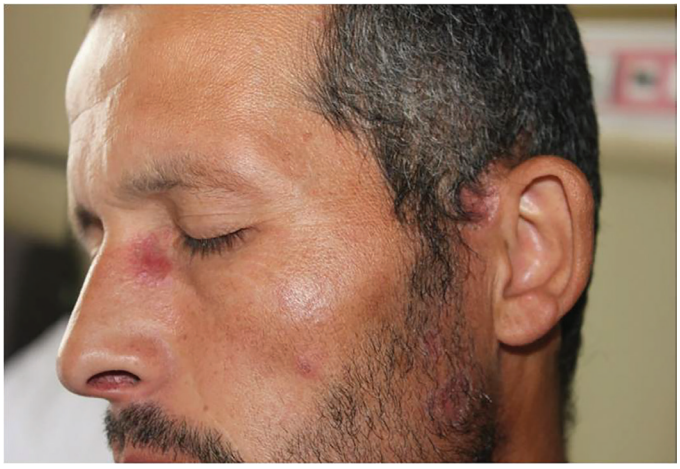

Figure 2. Erythematous plaques with peripheral elevation on the side of the nose, cheeks, ears, and temporal side of the face

lymph nodes as well as LAP (the greatest being 22x17 mm in size) with calcifications in the central lung.

An excisional biopsy for the cervical LAP was performed, which showed non-caseating granulomatous changes (Figure 3). A fundoscopic examination for possible uveitis showed normal findings. PPD was anergic. The histopathological assessment of the skin biopsy showed fibrotic scar-like areas in the collagen tissues as well as non-caseating granulomatous changes accompanied by giant cells in the dermis. Based on the clinical and histopathological findings, a diagnosis of cutaneous sarcoidosis with stage I lung involvement was made. The patient referred to department of pulmonary medicine. No treatment was recommended for lung involvement. Methylprednisolone aceponate ointment was given to the skin lesions.

Sarcoidosis was first recognized in 1877 as livid papillary psoriasis by Dr. Jonathan Hutchinson, an English dermatologist[2,3,5]. As in our case, it occurs more frequently in subjects under 40 years of age, with two age peaks, one between 20-29 years of age, and one above 45-55 years, particularly among women[2,6]. Lung involvement may occur in 90 to 95 of the cases (most frequently with bilateral hilar and mediastinal LAP, similar to our patient)[1,5]. Peripheral lymph nodes may be palpated in 10 to 15% of the patients. Our patient also had several palpable lymph nodes in the cervical region. Skin lesions of sarcoidosis are classified as specific or non-specific based on the presence or absence of non-caseating granulomas[1,3].

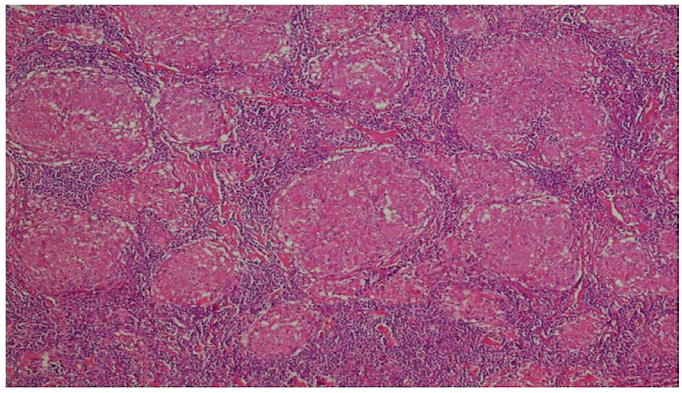

Figure 3. Histopathological view (hematoxylin and eosin&100): Non-caseating granulomatous inflammation abolishing the normal structure of the lymph node

Specific lesions are in the form of infiltrating plaques (sometimes hypopigmented), maculopapular eruption, ulceration, infiltration in an old scar, ichthyosis, alopecia, lupus pernio, or subcutaneous lesions. On the other hand non-specific lesions include erythema nodosum, erythema multiforme, calcification, prurigo, clubbed finger, and Sweet's syndrome[3,4,6]. The most common non-specific lesion is erythema nodosum[1,3,6], while the most common specific lesions are maculopapular lesions. These lesions may enlarge, transforming into plaques or annular lesions. Most of the plaques of sarcoidosis are infiltrated and reddish-brownish in color, and plaques are associated with chronic forms of the disease, are bilateral, symmetrical[6]. Diascopy shows the apple jelly sign in the form of papillae and plaque, which may also occur in other granulomatous diseases[1]. The most characteristic skin lesion in chronic cutaneous sarcoidosis is the so-called purplish "lupus pernio" with telangiectasia, while subcutaneous nodules may be a harbinger of systemic involvement. Pruritus is reported by nearly 15% of the patients[6]. Our patient also had mild pruritus in the erythematous annular plaques in the face, neck, and scalp.

Increased activity of macrophages and T-helper cells together with cytokine release play a major role in pathogenesis[4,5]. The classical pathological sign of sarcoidosis is the non-caseating granuloma with central epithelioid histiocytes, or more rarely Langerhans giant cells, surrounded by lymphocytes, macrophages, and fibroblasts[2]. From a histopathological viewpoint, differential diagnosis with other granulomatous disease should be made (e.g. tuberculosis, histoplasmosis, coccidioidomycosis, lepra, syphilis, and granuloma annulare)[1,2]. Laboratory tests may show hypercalcemia, hypercalciuria, and ACE elevation, more likely in acute sarcoidosis. Complete blood count may show anemia, lymphophenia, eosinophilia, and increased erythrocyte sedimentation rate[1]. Lung involvement does not always necessitate treatment[5,7]. In this case, regular follow-up at 6-month intervals was recommended for lung involvement, since no indications for systemic steroids could be identified. Treatment options include immunosuppressive agents such as topical or systemic steroids, azathioprine, and infliximab or others such as hydroxychloroquine or doxycycline[1,4]. Due to variable response rates to standard treatment with anti-malarial agents or methotrexate alternative therapeutic agents are being warranted. The aim of treatment in sarcoidosis is to achieve symptomatic relief,

improvement in objective test results indicative of disease activity, and to prevent chronic disability caused by disease progression[5,7]. Consent form was filled out by participant.

Ethics

Informed Consent: Consent form was filled out by participant.

Peer-review: External and internal peer-reviewed.

Authorship Contributions

Concept: G.Ö.Y., Design: G.Ö.Y., İ.H.Y., Data Collection or Processing: S.G.B., G.B., Analysis or Interpretation: İ.H.Y., G.Ö.Y., Literature Search: R.A., S.G.B., Writing: G.Ö.Y., R.A.

Conflict of Interest: No conflict of interest was declared by the authors.

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