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Cutaneous Sarcoidosis in a patient with left Hilar calcification of the lungs - A Case Report

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Abstract

Sarcoidosis is an acquired idiopathic granulomatous disease, which is characterized by noncaseating epithelioid granulomas in organs and tissues. Most frequently it affects the lungs, liver, lymph nodes, skin, eyes and other organs. The cutaneous lesions appear in 20 - 30% of patients with systemic manifestations, and in 25% of them they appear without systemic manifestations. Based on the histopathological characteristics, cutaneous lesions are divided into specific, characterized by cutaneous granuloma, and non-specific, which are not granulomatous. Moreover, they can be classified as typical and atypical. We are presenting a female patient with unilateral hilar calcification of the lungs, who exhibited plaque skin lesions typical for sarcoidosis, with a specific granulomatous histology and a favorable response to corticosteroid and antimalarial therapy.

Key words

Sarcoidosis; Skin Diseases; Lung Diseases; Case Reports; Prednisone; Antimalarials; Treatment Outcome

Sarcoidosis is an acquired idiopathic granulomatous disease, which is characterized by noncaseating epithelioid granulomas in the organs and tissues (1). Most frequently it affects the lungs, liver and heart (1, 2). Sometimes, granulomas are large and numerous, damaging the function of the affected organs.

The first description of sarcoidosis was published by Hutchinson in 1877 (3). Lupus pernio was described in 1889 by Besnier (4), and Tenneson in 1892 (5). Hutchinson described the Mortimer's disease in 1898, which was probably sarcoidosis, and the term sarcoidosis was first used by Boek in 1899 (6). The Boeck's assumption that the disease affected the skin, but also internal organs, was confirmed by Schaumann (7).

The disease affects all races and all ages (8), but also both sexes, although it is more frequent in women. The highest incidence is seen in persons under the age of 40 years (peak age, 20 - 29). The other peak occurs in women over 50 (9). The highest prevalence of sarcoidosis is in Europe, in Sweden and Ireland (60 in 100.000) (10). In African Americans, the incidence is 35.5 - 64 in 100.000, in women even up to 107 in 100.000, and in Caucasians in America 10 -15 in 100.000. It is 3 to 4 times more frequent, acute and serious in Afro-Americans than in Caucasians, and it is more commonly asymptomatic in Caucasians.

Cutaneous lesions appear in 20 - 30% of patients with systemic manifestations, and in 25% of them they appear without systemic manifestations.

When accompanied by systemic manifestations, lungs and intrathoracic lymph nodes are most frequently affected. Therefore, cutaneous lesions can appear as the only, the first manifestation, usually in the first stages of the disease (11), or with systemic manifestations, simultaneously or later. Cutaneous lesions are divided, based on the histopathological characteristics, into specific, and non-specific (1, 12). Moreover, they can be classified as typical and atypical. One patient can have different lesions simultaneously. Cutaneous sarcoidosis is known as a great imitator in dermatology, or a "clinical chameleon" (13, 14, 15, 16), because the lesions can have a wide range of clinical presentations.

We are presenting a female patient with unilateral hilar calcification of the lung, plaque skin lesion typical for sarcoidosis that exhibited specific granulomatous histology and favorable response to corticosteroid and antimalarial therapy.

Case report

Medical History

A 68-year-old female was referred to our Department with a 15-year-long history of skin lesions that first appeared on her back. Although she claimed that skin

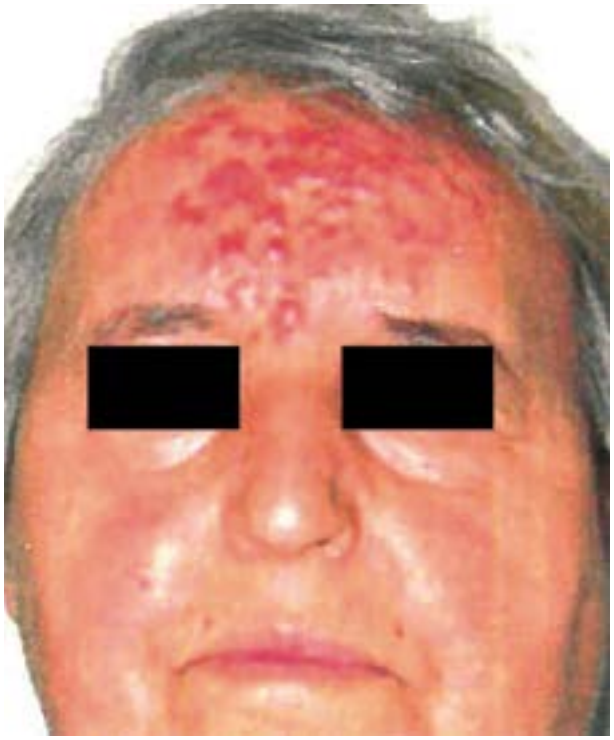


Figure 1. Multiple infiltrated plaques with distinct edges on the forehead and the apex, yellowish-orange to amber in color, in some places covered with crusts

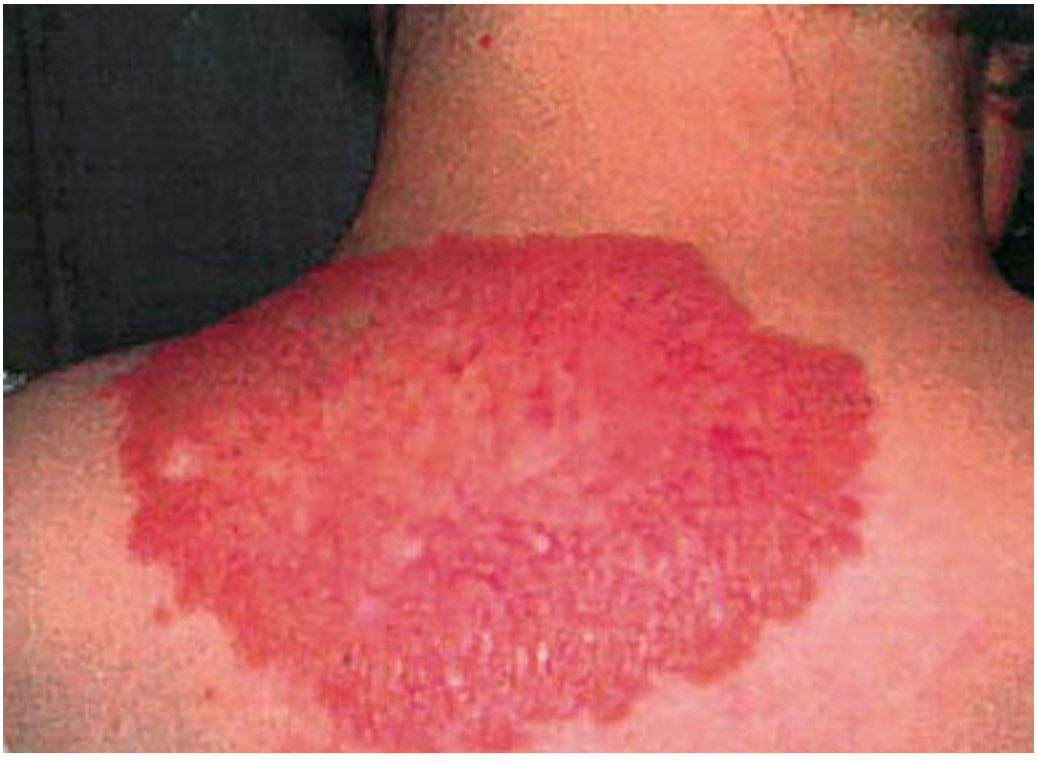


Figure 2. A lesion with a prominent edge, 12 x 10 cm on the back

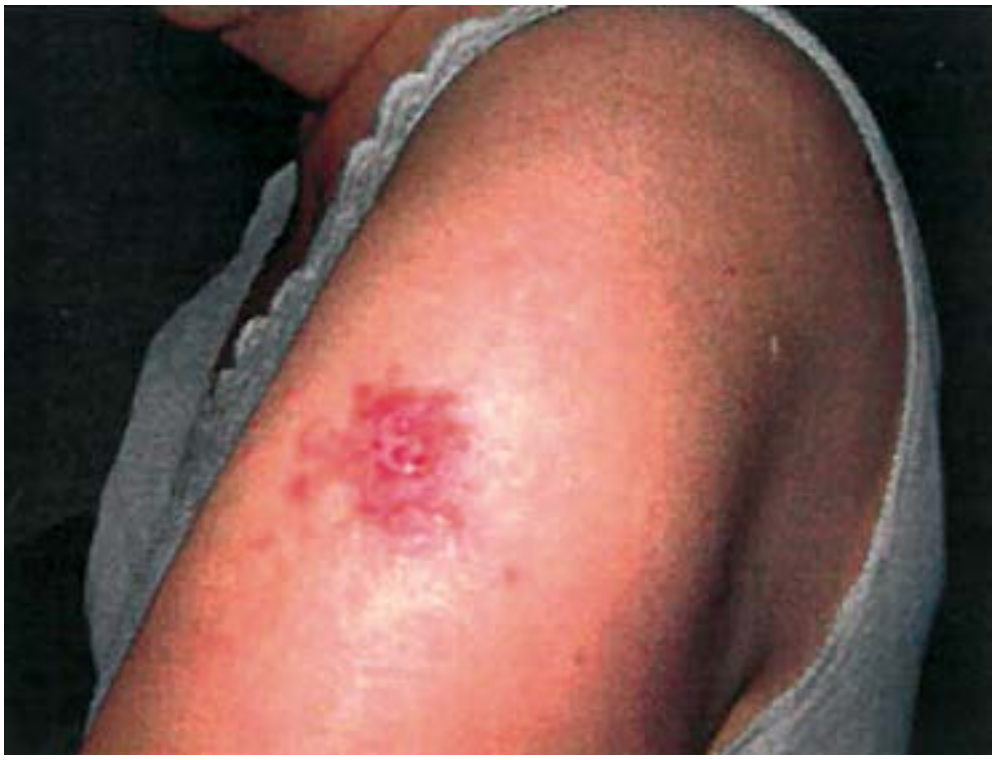
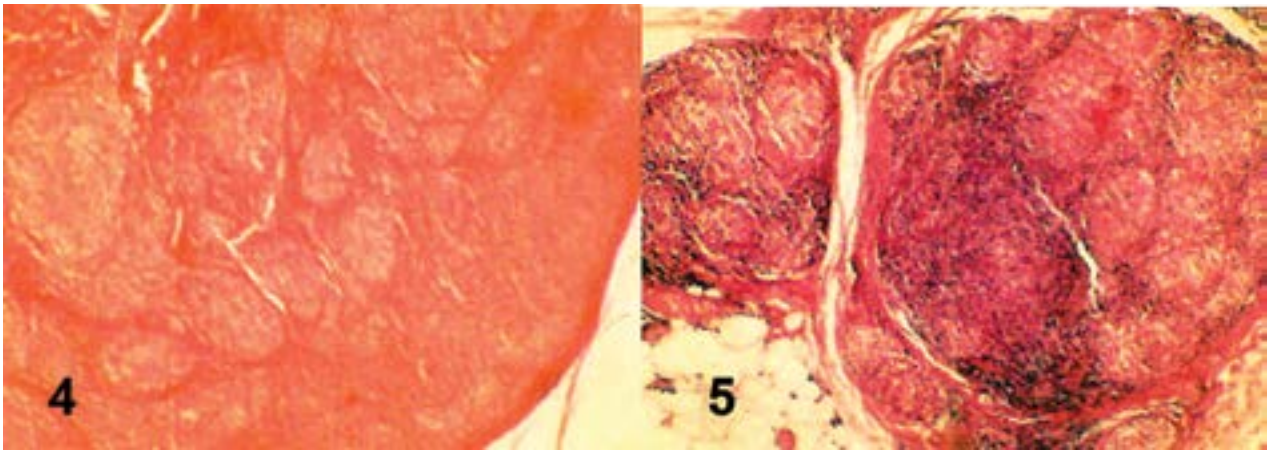


Figure 3. Nodular lesions, 2 cm in diameter, on both upper arms, similar in color, translucent

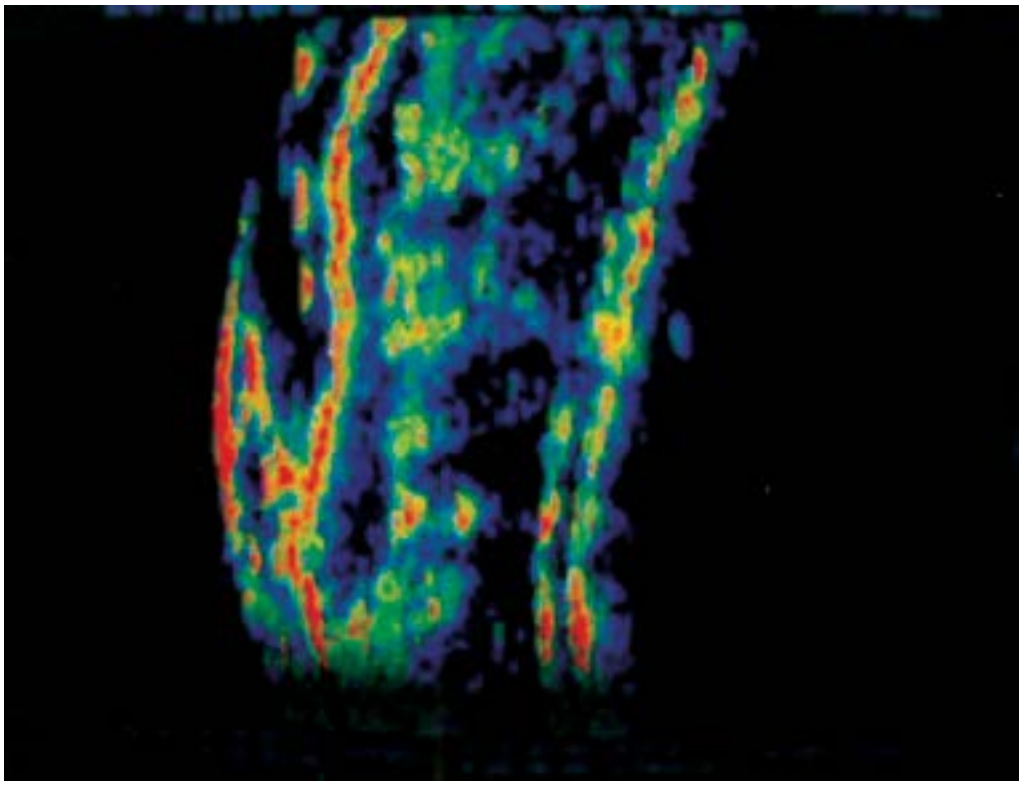
biopsy was taken 15 years before, unfortunately she did not have any medical reports. Three years ago, a new lesion appeared on her head and upper arms. There were no symptoms in other organs. She used various ointments, without any visible effect. Otherwise, she was in a good health and denied having any affected relatives.

Clinical examination

A clinical examination showed a lot of well defined plaques on the forehead and apex of the nose, yellowish-orange to amber color, some lesions covered with scabs (Figure 1). A lesion with prominent edge 12 x 10 cm (Figure 2) was visible on the back. Nodular lesions, 2 cm in diameter,



Figures 4. and 5. A strong granulomatous inflammatory reaction with numerous epithelioid and multinuclear Langerhans cells beneath the atrophic epidermis



Figures 6. Ultrasound of skin lesions on the forehead, above the root of the nose (at the beginning of treatment): A clearly limited lesion, different in thickness, resembling a string of beads. The thickest part of the skin lesion is 2.43 mm

were present on both upper arms, similarly colored, translucent (Figure 3).

Laboratory test results

All routine biochemical test results were within normal limits. Tuberculin testing was done by the Mantoux test with a 110 mm diameter of infiltration.

Histopathology

A strong inflammatory reaction of the granulomatous type with numerous epithelioid and multinucleated Langhans and/or foreign body cells were present beneath the atrophic epidermis (Figures 4 and 5).

X-ray and ultrasound imaging

Left hilar calcification was detected by chest X-ray. The ultrasound of the skin changes on the forehead, above the root of the nose, performed at the beginning of treatment revealed a well defined lesion without internal echo, different in thickness, looking like a string of beads. The thickest part of the skin lesion was 2.43 mm (Figure 6).

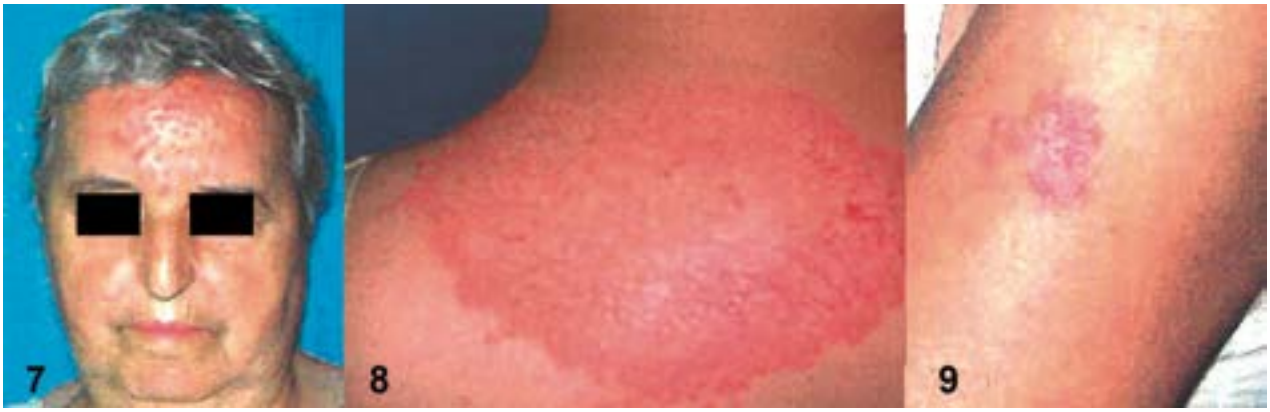
Therapy

Pronisone therapy, at a daily dose of 60 mg, slowly reduced to 20 mg per day, was initiated along with chloroquine phosphate (Delagyl) tablets, 250 mg twice a day. Betamethasone dipropionate ointment was applied topically twice a day, and cryotherapy with liquid nitrogen was commenced.

A significant improvement of the skin lesions was reported after three months of therapy: infiltration was paler, with less prominent (Figures 7, 8, 9) and reduced thickness (to 0,84 mm) identified by ultrasound (Figure 10).

Discussion

Sarcoidosis is an immune-mediated, complex inflammatory disease which represents abnormal immune response to various environmental factors in persons with genetic predisposition to granulomas (17, 18, 19, 20). It is characterized by activation of macrophages and CD4+ cells, and by accumulation of mononuclear phagocytes forming non-caseating epithelial cellular granuloma (21, 22).



Figures 7, 8 and 9. A significant improvement of skin lesions after three months: the lesions are paler, the infiltration is less prominent

Although the exact etiology of sarcoidosis has not been established, various agents which may act as antigens are believed to be responsible. These include viruses (Herpes virus, Epstein-Barr virus, Coxsackie B virus, Cytomegalo virus) and bacteria (Mycobacterium tuberculosis and other mycobacteria, Propionibacterium acnes (23), Borrelia burgdorferi, mycoplasma, Chlamydia (24), numerous nonorganic substances (aluminum, insecticides, lampblack, mold, titanium, silicon, iron, building materials, industrial dust) and organic substances (pine pollen, loam) (8, 24).

Genetics contributes a wide variety of clinical presentations and phenotypes noticed in sarcoidosis (18, 21).This has been confirmed by two findings - certain ethnicity was identified as an important risk factor, as well as the family history (19).

Antigen-presenting cells – macrophages and dendritic cells, interact with the antigen, which

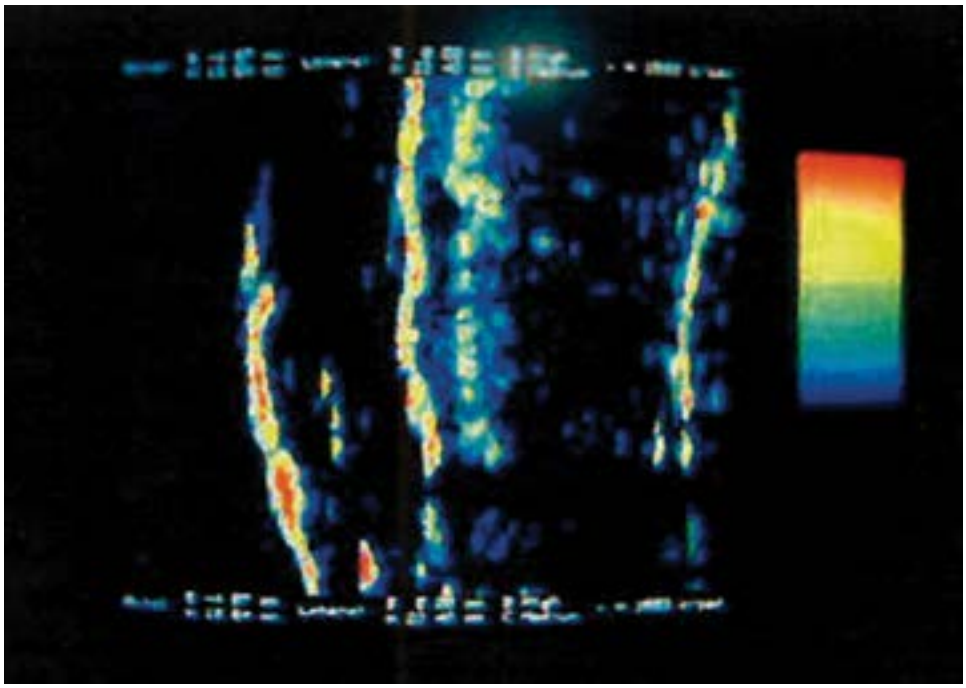


Figure 10. The ultrasound showed a reduced thickness - 0,84 mm

leads to activation and multiplication of CD4+ lymphocytes, initiating the immune process with Th1 cytokine profile. The produced cytokines: INF gamma (interferon gamma), TNF alfa (tumor necrosis factor alfa), IL-2 (interleukin-2), IL-12 (interleukin-12). IL-18 (interleukin-18), MIP 3 (macrophage proinflammatory human), GM-CSP (granulocyte-macrophage colony-stimulating factor) maintain the activation status and stimulate further mediator releasing and inflammation appearance, which are the characteristics of the disease. This occurs in various organs where the antigen is located, so due to the inability to remove the antigen, a typical epitheloid granuloma is created (25). In contrast to immune response at the site of inflammation in certain organs, state of energy is present on the periphery, which is characterized by decreased response when testing for delayed hypersensitivity and lower lymphocyte count in the peripheral blood in sarcoidosis patients, with unknown pathomechanisms (24).

Patients with sarcoidosis are at increased risk of malignant diseases, especially lymphoproliferative (26). There are a few mechanisms that can explain this phenomenon (27) – chronic inflammation, immune dysfunction, common etiological agents and genetic predisposition to autoimmune diseases and carcinoma (28, 29). Malignancies can develop after sarcoidosis, they can precede it, or, rarely, they develop simultaneously (30). Sarcoidosis can appear as a paraneoplastic syndrome. The antineoplastic treatment of either hematologic malignancies or solid tumors has also been observed to induce the initial onset or flare of sarcoidosis (31). The association between cutaneous sarcoidosis and malignancies is higher than with other types of sarcoidosis (29). Administration of immunosuppressants in sarcoidosis treatment increases the risk of infections, both fungal and bacterial. Association of sarcoidosis and HIV infection, which is rarely seen, is attributed to damaged immune system where CD4+ lymphocytes play the main role, causing granuloma formation in patients infected with HIV.

The cinical presentation of sarcoidosis may be quite peculiar, and it has been postulated that there is no disease with more varied manifestations (32). Cutaneous sarcoidosis without systemic involvement may occur in about one-quarter of all patients, and the same proportion of patients who develop a systemic disease may develop cutaneous sarcoidosis (32). When associated with systemic manifestations, lungs and intrathoracic lymph nodes are most frequently affected. Cutaneous lesions of sarcoidosis are generally classified as specific, if they are granulomatous infiltrates, and typical, if they appear as usual presentations of the disease.

Specific lesions include: papules or small nodules, plaques, nodules, subcutaneous and scarring forms, lupus pernio, angiolupoid and more rarely - scarring, but also non-scarring alopecia, erythroderma, ulcers, pustules, psoriasiform, ungual, and mucosal lesions (32, 33, 34).

Non-specific lesions include erythema nodosum, erythema multiforme, calcinosis cutis, ichthyosiform, as well miscelaneous, such as pruriginous varietas (35, 36).

Typical forms are most common and they are characterized by erythema nodusum, lupus pernio, nodular, papular, plaque forms, as well as subcutaneous and scarring forms (37, 38).

Atypical forms include erythrodermic, ichthyosiform, atrophic, ulcerous, verrucous, lichenoid, pruriginous, ungual, mocosal lesions, hyperpigmentations, and alopecia. Our patient had plaques on the forehead, on the capillitium and on the back (10 x 12 cm in diameter) and nodular lesions on the upper arms, up to 2 cm in diameter. The lesions were typical, both in localization and appearance.

Nodular lesions, including small nodular/papular lesions are most frequent (32). The lesions are localized on the face, extensor surfaces of the extremities, less commonly on the torso, gluteus, eyelids, and mucous membranes. The patients may have from several to hundreds of pale red, yellowish, and sometimes purple papules, 2 – 6 mm in diameter, with smooth surface, and sometimes with desquamation, up to larger, usually single or relatively few lesions, which remain circumscribed most often affecting proximal parts of the limbs, as in our patient.

Plaques manifest as red-brown, usually symmetrical circular or oval lesions, with elevated indurated edges, smooth, thin and paler skin in the center. Dilated blood vessels near the surface of the skin can be seen (angiolupoid), and annular forms are possible, most commonly on the face, forehead and neck. Plaques are, generally, most frequently

localized on the skin above the bones, on the forehead, shoulders, gluteus, thighs and ulnar aspect of forearms. Our patient had plaques on the forehead, on the capillitium and on the back (10 x 12 cm in diameter) and nodular changes on the upper arms, up to 2 cm in diameter. The lesions were typical, both in localization and appearance. The diagnosis of sarcoidosis was established based on the medical history, clinical presentation and typical histology with granuloma showing no caseation. Typical tuberculosis is usually distinguishable by histological features. On the other side, all attempts were made to delineate the full extent of the disease.

Any organ of the body may be affected by sarcoidosis, and the extent of cutaneous lesion does not correlate with the extent of the systemic disease.

The lungs are the most commonly affected organ (8) and more than 90% of patients with sarcoidosis present with lung symptoms. Based on chest radiography, lung sarcoidosis is divided into stages (32): stage 0 – normal chest x-ray (5 - 10%); stage I – bilateral hilar lymphadenopathy (only) (60 - 90%); stage II – bilateral hilar adenopathy with parenchymal changes (50 - 60%); stage III – diffuse pulmonary infiltration (less than 30%); stage IV – pulmonary fibrosis.

In our patient, the chest x-ray revealed a calcification of the left hilum, without other organ involvement. In sarcoidosis, atypical pattern of involvement of lymphadenopathy is not rare, which makes differentiation of sarcoidosis from other mediastinal diseases such as tuberculosis and especially Hodgkin's lymphoma, more difficult. Unilateral hilar lymphadenopathy is seen in less than 8% of sarcoidosis cases, but it is not uncommon in Hodgkin's lymphoma, where it accounts for 37.8% of cases. In sarcoidosis, unilateral hilar lymphadenopathy (if present) is approximately twice as common on the right side compared to the left side, and presence of lymph node calcification is more consistent with sarcoidosis. Contrast-enhanced CT scans may be helpful in differentiating intrathoracic sarcoidosis from Hodgkin's lymphoma based on the anatomical distribution of enlarged lymph nodes (39). Unfortunately, CT scan was not performed in our patient, and there was no other visible lymphadenopathy on the chest x-ray.

Treatment modalities for sarcoidosis are rather numerous, depending on the symptoms and the degree of systemic involvement (40), as well as on the possibility of spontaneous remission. In patients with isolated cutaneous forms, topical corticosteroids should be initiated, or even more effective intralesional corticosteroids (triamcinolone acetonide, 2 – 10 mg/ml in intervals of 2, 3 or 4 weeks). Application of topical tacrolimus has also showed satisfactory results. If this therapy fails to yield expected results, especially in chronic forms, like in plaque form, systemic therapy is applied (41).

Corticosteroids are usually prescribed in case of lung involvement, most frequently at initial dose of 20 – 40 mg/day of prednisone or equivalent doses, with gradual reduction during a couple of months.

Antimalarials – hydroxychloroquine, 200 – 400 mg/day, is the therapy of choice for mild isolated cutaneous forms over a longer period of time. Methotrexate can also be applied for pulmonary and extra-pulmonary manifestations (41) at doses of 10 – 25 mg per week, through an extended period, combined with low doses of corticosteroids.

Azathioprine has similar efficacy as methotrexate, and it can also be combined with corticosteroids.

Cyclophosphamide can be used orally and intravenously (50 – 150 mg/day - orally, or 500 – 2000 mg/week - intravenously), while chlorambucil in combination with low doses of prednisone.

Mycophenolate mofetil (45 mg/kg daily) combined with corticosteroids, showed significant improvements with no side effects in patients with pulmonary and extensively mucocutaneous sarcoidosis.

Thalidomide (42), tetracyclines (minocycline 200 mg/day, average period of administration 12 months, and doxycycline for relapse) (41), tumor necrosis factor–alfa (TNF–alfa) inhibitors (infliximab, etanercept) are also applied.

Ultraviolet therapy (UVA 1 and PUVA), showed good results with cutaneous forms of sarcoidosis. A favorable response to corticosteroid and antimalarial systemic therapy was achieved in our patient after three months.

The prognosis of sarcoidosis is generally better in females, in those with less severe pulmonary disease at the onset, and in patients with a positive tuberculin test (32).

Spontaneous remission occurs in 2/3 patients, while 1 - 3% of patients have a chronic course. Papular and nodular lesions resolve over the course of months or years, while the plaques are more resistant. The overall level of mortality from sarcoidosis varies from 1 - 5%, and with chronic liver disease the percentage increases to 12%.

Conclusion

This is a case of a female patient with unilateral hilar calcification of the lung, who exhibited plaque skin lesions typical for sarcoidosis, with specific granulomatous histology and a favorable response to corticosteroid and antimalarial therapy, confirmed by ultrasound skin imaging.

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Sarcoidosis cutis kod pacijenta sa kalcifikacijom u levom hilusu pluća - prikaz slučaja

Sažetak

Sarkoidoza je stečena idiopatska granulomatozna bolest za koju je karakteristično prisustvo nekazeifikujućih epiteloidnih granuloma u organima i tkivima. Bolest se javlja kod osoba svih rasa i u svim uzrastima, takođe kod oba pola, nešto češće kod žena. Na osnovu histopatoloških karakteristika, kutane manifestacije se dele na specifične, koje karakteriše sarkoidni granulom i nespecifične, koje nisu granulomatozne. Takođe se mogu klasifikovati kao tipične i atipične. Kada su u kombinaciji sa sistemskim zahvatanjem, najčešće se radi o plućima i intratorakalnim limfnim žlezdama. Kutana sarkoidoza je poznata kao jedan od velikih „imitatora" u dermatologiji ili kao „klinički kameleon", jer lezije mogu imati širok spektar poremećaja.

Prikaz bolesnice. Kod bolesnice od 68 godina prve promene su se javile na koži leđa pre 15 godina, a pre tri godine i na glavi i nadlakticama. Nema simptome od drugih organa. Klinički, na čelu i temenu prisutno više jasno ograničenih infiltrovanih plaža narandžastožućkaste do mrke boje, mestimično pokrivenih krustama. Na leđima plaža 12 x 10 cm, sa naglašenim rubom. Na obema nadlakticama nodularne promene do 2 cm u prečniku, slično prebojene, translucidne. Laboratorijske analize u granicama normale. Testiranjem na tuberkulin *Mantoux* metodom registrovana induracija od 10 mm. Patohistološki nalaz: ispod atrofičnog epiderma jaka inflamatorna reakcija granulomatoznog tipa sa brojnim epiteloidnim i multijedarnim ćelijama *Langhansovog* tipa. X-zracima registrovana kalcifikacija u levom plućnom hilusu. EHO promene na čelu iznad korena nosa (na početku lečenja): najveća debljina same promene 2,43 mm. Primenjena terapija: pronizon tablete u dnevnoj dozi od 60 mg do smanjenja na 20 mg dnevno, *chloroquin phosphat* (*Delagyl*) tablete a 250 mg 2 x 1 dnevno, betametazon propionat mast dvaput dnevno, krioterapija tečnim azotom. Posle tri meseca registrovano znatno poboljšanje promena na koži: promene bleđe, manje izražena infiltracija i EHO pregledom registrovano smanjenje debljine na 0,84 mm.

Diskusija. Sarkoidoza je imunoposredovana inflamatorna kompleksna bolest koja predstavlja abnormalni imunoodgovor na raznolike faktore iz okruženja (virusi, bakterije, razne organske i neorganske materije) kod osoba genetski predisponiranih da razvijaju granulome. Kod naše bolesnice promene su bile tipične po lokalizaciji i izgledu. Rtg pluća pokazao kalcifikaciju levog hilusa. Dijagnoza sarkoidoze zahteva određene postupke: iscrpnu anamnezu, kliničku prezentaciju i tipičnu histologiju granuloma bez kazeifikacije. Na osnovu ovih procedura, postavljena je dijagnoza kod naše bolesnice . Dodatno je urađen i EHO kožne promene

na početku i posle tri meseca primene terapije. Pored lokalne terapije kortikosteroidima, bilo je neohodno primeniti i sistemsku terapiju (kortikosteroidima i antimalaricima), što je dovelo do poboljšanja.
Zaključak. Prikazana je bolesnica sa unilateralnom hilarnom kalcifikacijom pluća, sa plak tipom kožnih lezija tipičnih za sarkoidozu, sa specifičnim histološkim nalazom i povoljnim odgovorom na terapiju kortikosteroidima i antimalaricima, što je potvrđeno ultrazvučnim pregledom kože.

Ključne reči

Sarkoidoza; Kožne bolesti; Plućne bolesti; Prikazi slučajeva; Prednizon; Antimalarici; Ishod terapije