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A case of ichthyosiform sarcoidosis with unusual localization

Case Report

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Abstract: Sarcoidosis is a systemic granulomatous disease of unknown cause that commonly involves the lungs, lymph nodes, bones, liver, spleen, or skin. Cutaneous findings of sarcoidosis occur in 20% to 35% of patients with systemic disease. The recognition of cutaneous lesions is important, because it gives important clues to diagnosis and also allows for easy biopsy. We report a 71-year-old Turkish woman with erythematous lesions, which included widespread, erythematous macules of various sizes on the chest, abdomen, and back on both sides of her body. Fine white scales covered some of the lesions. Hepatomegaly and bilateral hilar lymphadenopathy were also observed. Biopsy specimens of the skin showed dermal, noncaseating, epitheloid granulomas. The diagnosis, based on correlation of the clinical presentation and histopathological findings, was ichthyosiform sarcoidosis with systemic involvement. The difference between our case and other cases of ichthyosiform sarcoidosis described in the literature is that ours involved only the trunk and not the extremities. We are presenting this case because it is an interesting and rare variant of ichthyosiform sarcoidosis.

Keywords: *Sarcoidosis • Ichthyosiform eruption • Skin lesions*

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1. Introduction

The cutaneous manifestations of sarcoidosis are classified as specific or nonspecific. Specific lesions are noncaseating granulomas in the dermis. They are variable but classically include small papules, plaques, lupus pernio, macules, subcutaneous nodules, and infiltration of old scars. Nonspecific lesions do not show granulomatous inflammation in the dermis. The most common nonspecific cutaneous lesions are erythema nodosum [1-3]. The clinical presentation of sarcoidosis varies to such an extent that it is called “the great mimic” of disorders. Up to now, there have been only a few case reports of ichthyosiform sarcoidosis in the literature.

2. Case Report

A 71-year-old Turkish woman with the suspected diagnosis of lung cancer was referred to our hospital. Two months previously, she had experienced fatigue, weight loss, loss of appetite, shortness of breath, and skin eruptions. On physical examination, hepatomegaly was detected, and widespread, erythematous macules of various sizes were seen on the chest, abdomen, and back on both sides of the body. In some areas, these macules were united and formed a reticular shape. Fine, white scales covered some of the lesions. The patient’s skin was atrophic and had a “cigarette paper” appearance, even in the healthy areas. Some of the lesions had areas of induration that could be palpated with difficulty (Figure 1). Skin-biopsy specimens from both the macular and ichthyosiform lesions showed

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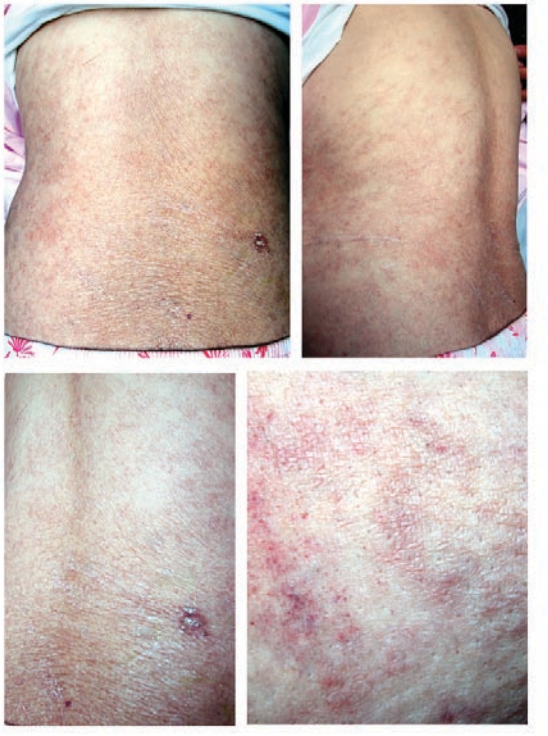
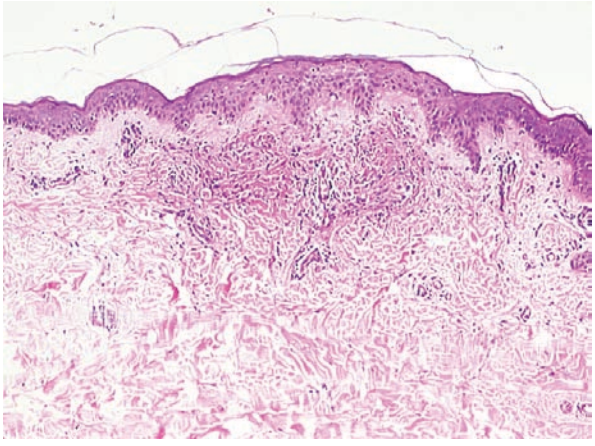
Figure 1. Lesions on the posterior and anterior trunk show an ichthyosiform pattern that is characterized by fine, white scales.



dermal noncaseating epitheloid granulomas consistent with sarcoidosis (Figure 2). There were no polarizable foreign bodies. Periodic acid-Schiff and Ziehl–Neelsen stains were negative for both mycobacteria and fungal organisms. Polymerase chain reaction for mycobacterium also produced a negative result. Laboratory data showed mild anemia (hemoglobin, 11 g/dl). The serum chemistry panel was unremarkable, except for hypoalbuminemia (1.9 g/dl; normal, 3.5–5.5) and an elevated lactate dehydrogenase level of 881; (normal, 250–450 U/L). The serum angiotensin-converting enzyme level was elevated at 136.7 U/L (normal, 8-52). Intradermal skin testing showed a negative reaction to purified protein derivative. Ocular examination was unremarkable. Both a radiograph and computed tomographic (CT) scan of the chest showed bilateral hilar lymphadenopathy. Laboratory analysis showed transudative ascites and cytologic examination was negative. The results of clinical, histological, and laboratory investigation indicated that the patient had systemic sarcoidosis, with acquired ichthyosiform erythroderma. After diagnosis, she was treated orally with prednisone (60 mg/day). Her pulmonary status, skin lesions, and general condition began to improve, and she was discharged from the hospital.

Figure 2. Dermal noncaseating epitheloid granulomas.



3. Discussion

Sarcoidosis is a systemic disease of unknown cause characterized by formation of noncaseating granulomas. Cutaneous lesions, depending on the absence or presence of noncaseating granulomas, are classified as specific or nonspecific [2].

Ichthyosiform sarcoidosis is an uncommon specific cutaneous manifestation of sarcoidosis. The appearance of ichthyosiform lesions may occur concurrently with, follow, or precede the diagnosis of systemic sarcoidosis [2,5]. Ichthyosiform sarcoidosis generally involves the extremities, especially pretibial areas of the lower extremities [2-6,9-12]. Involvement of the trunk may occur rarely [4,5,7]. The lesions are papules and plaques of various sizes [3-5,11]. However, macular lesions may also occur [3,7]. Polygonal, pigmented, adherent scales are generally seen on the lesions [4,5,7], and fine, white scales may rarely occur [7]. In our case, erythematous macules of various sizes were found on the whole body. These macular lesions combined to form reticules in some areas. There were fine, white scales on some lesions. The differences between our case and other cases of ichthyosiform sarcoidosis described in the literature are that our case involved only the patient's trunk and not the extremities and only macular lesions (no papules or plaques) were observed. In addition, these macular lesions formed large patches. However, the squames were not in classic polygonal or rhomboid lamellar shape, but in fine, white scales.

Acquired ichthyosis can be associated with systemic illness, including lymphoma, especially Hodgkin's disease and T-cell lymphoma; leukemia; carcinoma of the breast, lung, and cervix; leiomyosarcoma; Kaposi's sarcoma; and intestinal sarcoma. Other systemic diseases associated with acquired ichthyosis are

systemic lupus erythematosus, hypothyroidism, chronic malnutrition, leprosy, renal disease, and vitamin A deficiency [2,6].

Medication that interferes with sterol synthesis in epidermal cells, such as nicotinic acid, triparonal, butyrophenone, dixyrazine and nafoxidine, can be implicated in the pathogenesis of acquired ichthyosis [6]. Rarely, ichthyosis has been shown to occur in patients receiving high doses of clofazimine and cimetidine. Asteatosis induced by the oral retinoids etretinate and isotretinoin may resemble ichthyosis [5,6].

In addition, in children, juvenile rheumatoid arthritis must be considered in the clinical differential diagnosis for acquired ichthyosis [7,8].

In conclusion, ichthyosiform sarcoidosis is an unusual cutaneous manifestation of sarcoidosis. We emphasize that ichthyosiform lesions may be the only cutaneous manifestation of sarcoidosis. Therefore, cutaneous biopsy is necessary for a histologic confirmation of the diagnosis and for exclusion of other systemic illnesses and internal cancers. Recognition of cutaneous lesions is important because it provides a visible clue to the diagnosis, and those lesions are an easily accessible source of biopsy material for histologic examination.

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