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Journal of Skin and Sexually Transmitted Diseases

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Frontal swelling – An unusual presentation of sarcoidosis: A case report

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Received: 02 August 2021
Accepted: 26 September 2021
EPub Ahead of Print: 24 November 2021
Published: 09 April 2022

DOI
10.25259/JSSTD_66_2021

Quick Response Code:

ABSTRACT

Subcutaneous sarcoidosis is a rare, cutaneous expression of systemic sarcoidosis. It is observed as asymptomatic firm, nodules covered by normal-appearing skin, principally on the extremities, which show the typical histopathology appearance of non-caseating granulomas localized to the subcutaneous tissue. It may be associated with the early benign, hilar lymphadenopathy of sarcoidosis or with the later stages of progressive sarcoidosis. Here, we report a case of subcutaneous sarcoidosis, which presented as an asymptomatic, forehead swelling with involvement of skull bone and lung. We report this case since subcutaneous sarcoidosis on the face with involvement of skull bone is a rarity.

Keywords: Subcutaneous sarcoidosis, Darier-Roussy sarcoid, Bone sarcoidosis, Osteolytic lesions

INTRODUCTION

Sarcoidosis is a systemic granulomatous disease of unknown origin and a master mimicker that can affect any organ.[1] Skin lesions are heterogeneous and may be specific or nonspecific based on the presence or absence of typical sarcoidal granulomas.[2] Subcutaneous sarcoidosis is a rare, but specific subset of cutaneous sarcoidosis, first described by Darier and Roussy in 1904, and is also known as Darier-Roussy sarcoid.[3] It's diagnosis may require a high index of suspicion. Bone involvement is rare in sarcoidosis and when present, can manifest as an osteolytic lesion. Here, we report a case of subcutaneous sarcoidosis with involvement of lungs and frontal bone of skull.

CASE REPORT

A 50-year-old female, without any other comorbidities, presented with a single, skin-colored, asymptomatic swelling on the forehead. The swelling was about 1 × 1 cm in size initially and gradually increased in size over a period of 6 months. There was no history of any trauma or any systemic symptoms. The patient gave a history of a similar swelling 6 years back at the same site, which was treated with curettage and cementation. On clinical examination, a well-defined, mobile, firm to hard, non-tender nodule of size 2 × 2 cm was present over the right side of the forehead. Skin over the swelling was normal and not adherent to underlying structures [Figure 1]. Systemic examination was unremarkable.

Our differential diagnoses were foreign body granuloma, lipoma, and cutaneous metastasis. Complete hemogram, urine microscopy, and renal and liver function tests were within normal

limits except for an elevated erythrocyte sedimentation rate (40 mm/hour). Fine-needle aspiration cytology from the nodule was inconclusive. Radiography of skull showed a lytic lesion in the frontal bone [Figure 2]. Computed tomography (CT), magnetic resonance imaging (MRI), and positron emission tomography (PET) scans of skull and brain confirmed the presence of a solitary lytic lesion in the frontal bone with an extracranial soft-tissue swelling.

Histopathological examination of the forehead swelling showed presence of naked granulomas composed of epithelioid histiocytes and giant cells, in the dermis, which was consistent with sarcoidosis [Figure 3]. The specimen did not show any atypical cells. Wade-Fite stain for acid–fast bacilli and Giemsa stain for fungal elements were negative. Biopsy specimen from the underlying frontal bone showed lymphocyte-poor non-caseating granulomas with occasional fibrosis [Figure 4].

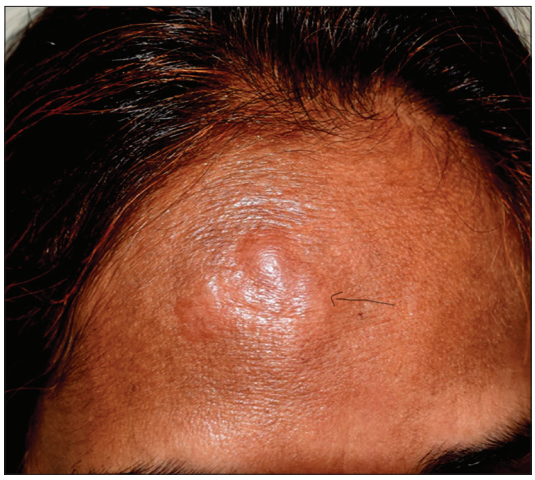

Figure 1: A well-defined, mobile, firm to hard, non-tender nodule of size 2 × 2 cm on the right side of forehead (black arrow).

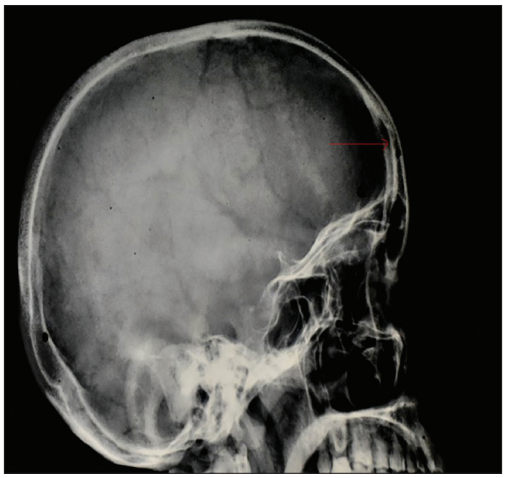

Figure 2: Radiography of skull showing a lytic lesion (red arrow) in the frontal bone.

Mantoux test with five units of purified protein derivative was negative. Serum calcium and angiotensin-converting enzyme levels were within normal limits. Chest radiography showed bilateral pulmonary reticulonodular shadows. High-resolution CT scan of the chest showed subpleural thickening with atelectasis involving lower lobes of both lungs. Ophthalmology evaluation did not show any abnormality. Electrocardiogram and a detailed cardiology evaluation did not show any abnormality.

Thus, we arrived at a final diagnosis of sarcoidosis with cutaneous, pulmonary, and osseous involvement and referred the patient to the physician. She was treated with methotrexate 10 mg once a week and hydroxychloroquine 200 mg twice a day. Subcutaneous nodule subsided within 3 months of treatment and the patient opted for further treatment from a nearby hospital.

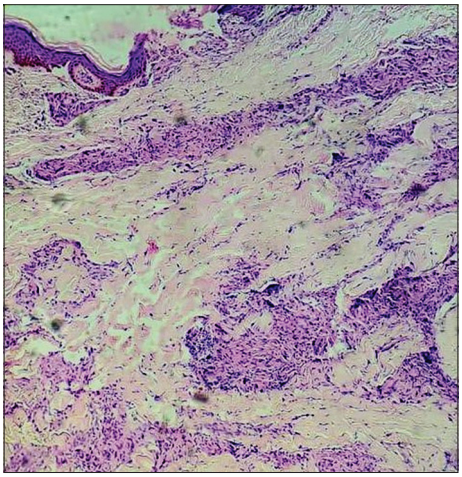

Figure 3: Histopathological examination of the forehead swelling showing the presence of naked granulomas composed of epithelioid histiocytes and giant cells in the dermis (H and E, ×100).

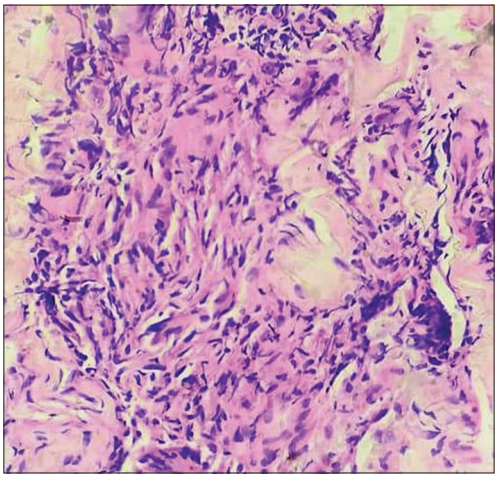

Figure 4: Histopathological examination of the specimen from frontal bone showing lymphocyte-poor non-caseating granulomas with occasional fibrosis (H and E, ×400).

DISCUSSION

Subcutaneous sarcoidosis is a rare, manifestation of sarcoidosis which has a female predilection. Mostly, the disease affects middle-aged women and favors the extremities.[4] Subcutaneous sarcoidosis is clinically characterized by asymptomatic skin-colored nodules and is seen in about 1.4–6% of patients with the disease.[2] In our patient, the site (forehead, which is not a common site for subcutaneous sarcoidosis), history of curettage, and cementation of the area and paucity of systemic symptoms, made us consider foreign body granuloma as the more probable diagnosis, while the osteolytic lesion observed in the skull radiography indicated the possibility of a malignant lesion. However, the histopathology examination of biopsy from the forehead swelling and curettage of the underlying bone was suggestive of sarcoidosis.

The uncommon features noted in our case were the site of the lesion (subcutaneous sarcoidosis is commonly seen on the extremities and rare on the forehead) and the involvement of frontal bone. Small bones of hands and feet are the common locations for bone sarcoidosis.[5] Cutaneous lesions can occur along with bone lesions, which manifest as well marginated areas of bone translucency in radiography.[6] CT scan of skull identifies sarcoid lesions as regions of enhanced uptake in subcutaneous area with areas of bone destruction and sclerosis.[7,8] MRI findings are high-intensity signals in T2-weighted images.[8] However, in our patient, these imaging modalities were inconclusive and histopathology revealed the typical sarcoidal granulomas.

Subcutaneous sarcoidosis may be associated with the early benign hilar adenopathy syndrome of sarcoidosis or with the later stages of progressive sarcoidosis.[9] Our patient had evidence of pulmonary and bone sarcoidosis at the time of diagnosis of subcutaneous sarcoidosis.

Bone sarcoidosis was described by Kreibich for the 1st time in 1904.[8] Even though, the disease most commonly affects hands and feet, other sites such as axial skeleton, skull, long bones, and ribs may also be affected occasionally. The diagnosis is often difficult since the disease when manifests as lytic lesions, may mimic metastasis or myeloma.[10] Targeted biopsy remains the most reliable diagnostic investigation. PET, CT, and MRI scans can provide supportive evidence for diagnosis. MRI is a very useful, second-line investigation when CT and PET scans are inconclusive. When metastasis or myeloma is a differential diagnosis, MRI can delineate the smooth margins of the lesion and indicate the benign nature of the disease.[7] However, in our patient, these radiological investigations were inconclusive. Our case highlights that bone sarcoidosis should be added to the list of conditions that can present as osteolytic skull lesions. Histopathological examination incorporating special staining techniques helps to rule out the histopathology mimics and to arrive at a correct diagnosis.

In sarcoidosis, systemic treatment is recommended, when a patient presents with disfiguring skin lesions and/or organ involvement. Systemic corticosteroids are the most effective treatment. Methotrexate, antimalarials, tetracyclines, and tumor necrosis factor-alpha antagonists have also been used with varying response.[11] In resistant forms of bone sarcoidosis, infliximab is found useful.

CONCLUSION

Cutaneous manifestations may be an early feature of sarcoidosis. Bone sarcoidosis, though rare, should be considered as a differential diagnosis of osteolytic skull lesions.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

Financial support and sponsorship

Nil.

Conflicts of interest

Dr. Mary Vineetha is on the editorial board of the Journal.

REFERENCES

1. Mohan P, Joy B, Anoop T, Sridharan R. Sarcoidosis-the master mimicker. J Skin Sex Transm Dis 2021;3:96-8.
2. Fernandez-Faith E, McDonnell J. Cutaneous sarcoidosis: Differential diagnosis. Clin Dermatol 2007;25:276-7.
3. James WD, Berger TG, Elston DM, editors. Andrew's Diseases of the Skin: Clinical Dermatology. 11th ed. Philadelphia, PA: Saunders, Elsevier; 2006. p. 700-5.
4. Dalle Vedove C, Colato C, Girolomoni G. Subcutaneous sarcoidosis: Report of two cases and review of the literature. Clin Rheumatol 2011;30:1123-8.
5. Zhou Y, Lower EE, Li H, Farhey Y, Baughman RP. Clinical characteristics of patients with bone sarcoidosis. Semin Arthritis Rheum 2017;47:143-8.
6. Teirstein AS, Wolf BS, Siltzbach LE. Sarcoidosis of the skull. N Engl J Med 1961;265:65-8.
7. Charifi Y, Dkhissi Y, Haloua M, Lamrani YA, Berrady G, Boubbou M, *et al*. Pelvic and lumbar pain revealing bone sarcoidosis: A case report. Clin Med Insights Case Rep 2021;14:11795476211014788.
8. Kemah B, Özturan B, Bilgic B, Özkan K, Akpınar F, Kılıc B. An unusual orthopaedic disease: Sarcoidosis-a case report. Surg J (NY) 2017;3:e75-8.

9. Vainsencher D, Winkelmann RK. Subcutaneous sarcoidosis. Arch Dermatol 1984;120:1028-31.
10. Zisman DA, Shorr AF, Lynch JP 3rd. Sarcoidosis involving the musculoskeletal system. Semin Respir Crit Care Med 2002;23:555-70.
11. Mana J, Marcoval J, Rubio M, Labori M, Fanlo M, Pujol R. Granulomatous cutaneous sarcoidosis: Diagnosis, relationship to systemic disease, prognosis and treatment. Sarcoidosis Vasc Diffuse Lung Dis 2013;30:268-81.

How to cite this article: Poulose L, Mathew S, Sahadevan G, Palakkal S, Vineetha M. Frontal swelling – An unusual presentation of sarcoidosis: A case report. J Skin Sex Transm Dis 2022;4:115-8.