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Subcutaneous sarcoidosis mimicking tumor

Sonali Someek Basu, Pranav Kumar

ABSTRACT

Subcutaneous sarcoidosis is a multisystem granulomatous disease with cutaneous lesions present in about one fourth of patients. Diagnosis may require a high index of suspicion. A patient was referred to the respiratory clinic for evaluation of a firm, non-tender, subcutaneous, mobile mass on the left forearm with bilateral hilar lymphadenopathy on the CT chest. There is a strong association between subcutaneous sarcoidosis and bilateral hilar lymphadenopathy (72.7%). Clinical and histopathological reports have confirmed subcutaneous sarcoidosis, also radiological examination with the hilar lymph node involvement. About 15% of patients have in order of frequency uveitis, parotitis, arthritis, mucositis, dactylitis, neurological and renal involvement, hepatosplenomegaly. This is a rare form of the nodular cutaneous sarcoidosis.

Keywords: Corticosteroids, Epithelioid granuloma, Methotrexate, Sarcoidosis

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INTRODUCTION

Sarcoidosis is a multisystem granulomatous disorder of unknown aetiology, it affects individuals worldwide, characterized by the presence of non-caseating granulomas in involved organs and tissues [1]. Jonathan Hutchinson described the first case in 1869 [2]. It involves skin, lung, lymph nodes, eyes, joints, brain, kidneys and heart. Skin involvement is the second most common following pulmonary involvement in sarcoidosis.

Muscle involvement in sarcoidosis was first reported by Licharew in 1908. 25% patients of sarcoidosis presents with cutaneous lesions, it varies in morphologies, including papules, nodules, plaques and infiltrated scars versus subcutaneous sarcoidosis that occurs in 1.4 to 6% patients of systemic sarcoidosis [3]. Subcutaneous sarcoidosis affects women commonly, in their fifth and sixth decades. Lesions could be multiple, bilateral, asymmetrical, asymptomatic hard indurated mobile subcutaneous nodule/mass located in upper extremities, commonly involve forearm, without any changes in overlying epidermis [4]. Nodular muscular sarcoidosis often mimics a tumour. MRI and muscle biopsy are useful investigations for diagnosis [5, 6].

Biopsy and histopathological examination is the gold standard method of diagnosis which shows “naked” granuloma formation which has sparse lymphocytes at margin with epithelioid cells with little or no necrosis. Diagnosis is confirmed after ruling out other differentials that causes granulomatous lesion [7].

Initial treatment option is the steroid. Immunosuppressive agents have been used in corticosteroid resistant form [8]. We report this case of asymptomatic nodular sarcoidosis with pulmonary involvement showing bilateral hilar lymphadenopathy and small pulmonary nodules on imaging which was initially thought to be fibro-sarcoma.

CASE REPORT

A 61-years-old man with rapidly enlarging painless mass on his left forearm which is noticed two months ago (Figure 1). It is mostly painless and initially thought to be fibro-sarcoma. There is no history of trauma. Ultrasound repeated twice, the size has increased from 14.4 x 4.7 cm to 18 x 6 cm (Figure 2). The high signal intensity area was observed in MRI examination (Figure 3).

He has had multiple sebaceous cysts removed from his back. He has background history of Type II Diabetes and is on Lantus 20U nocte, Metformin 1 gm BD, Gliclazide MR 120 mg nocte and hyperlipidaemia treated with Atorvastatin 40 mg. He is a diesel mechanic, a non-smoker and a social drinker. He has family history of Type II Diabetes. There was no axillary or inguinal lymphadenopathy. He has unremarkable systemic signs. Serum Angiotensin Converting Enzyme level was 100 U/Land urine calcium excretion ratio is 7.8. Spirometry –Respiratory Function Test was suggestive of good inspiratory effort. The flow volume loops, lung volumes and CO gas transfer are all within normal limits. CT chest showed bilateral extensive hilar mediastinal lymphadenopathy with right upper lobe sub-pleural nodule and left lower lobe sub-pleural nodule. The distribution of hilar node did raise the possibility of sarcoidosis and lymphoma. There was no parenchymal lung involvement showing consolidation or fibrosis (Figure 4). His histopathology has been found to be a case of subcutaneous sarcoidosis involving muscle (Figure 5). As this is nodular form of muscular sarcoidosis, making it the least common subtype of specific lesion of this disease is often confused with a soft tissue neoplasm. Based on the clinical, radiological and histological findings, he was started on oral prednisolone and has regular follow up with respiratory physician.

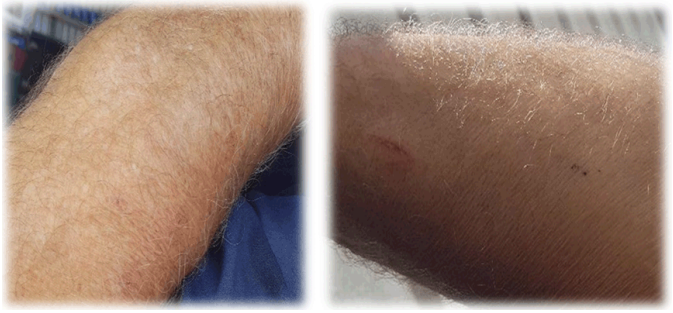

Figure 1: A firm, mobile, painless nodule, raised above skin with normal overlying epidermis.

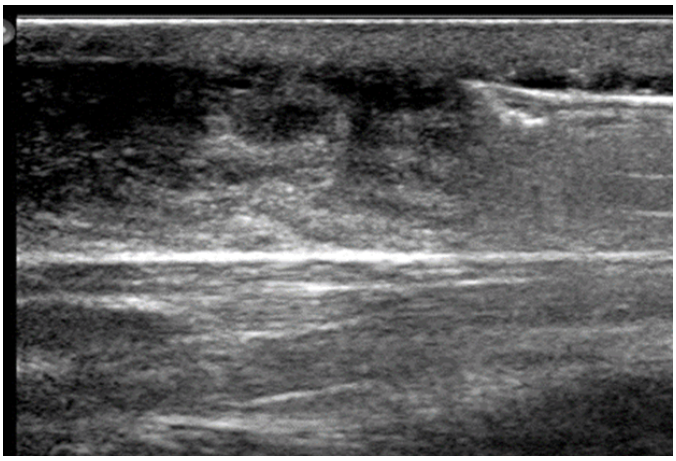

Figure 2: US suggestive of hypoechoic oval nodule underneath skin.

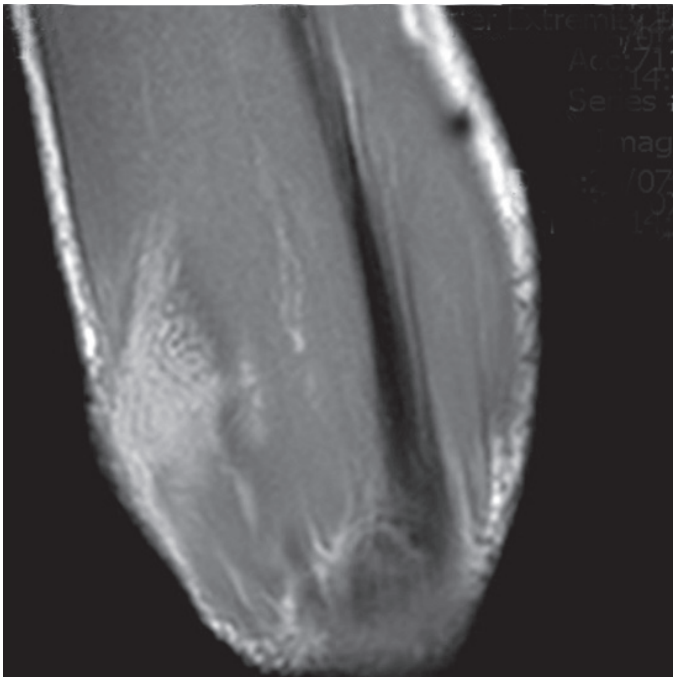

Figure 3: MRI of left forearm showed intramuscular involvement with homogenous high signal intensity area.

DISCUSSION

Sarcoidosis is a multisystem granulomatous disorder which can present as numerous skin manifestations such as cutaneous nodule. It mimics infections and malignancies.

In 1899, a Norwegian dermatologist Caesar Boeck, described sarcoid skin lesion “fleshy” meaning “sakodes” in Greek. Darier-Roussy described specific subtype of nodular cutaneous in 1904. Drier-Roussy sarcoidosis is now replaced by more accurate term ‘subcutaneous sarcoidosis’ [9–11]. Subcutaneous sarcoidosis is a nonspecific form of sarcoidosis, a non-severe systemic disease and is not associated with chronic fibrotic disease. Granuloma primarily involves subcutaneous tissue [12].

Subcutaneous sarcoidosis diagnosis is associated with extra-cutaneous systemic disease involvement, especially bilateral hilar adenopathy. It also has been described in the absence of systemic disease.

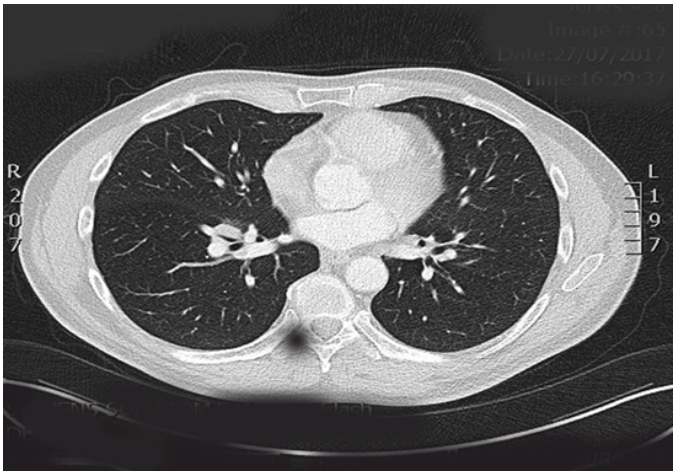

Figure 4: CT chest showed bilateral extensive hilar mediastinal lymphadenopathy with right upper lobe sub-pleural nodule and left lower lobe sub-pleural nodule. The distribution of hilar node did raise the possibility of sarcoidosis and lymphoma. There was no parenchymal lung involvement showing consolidation or fibrosis.

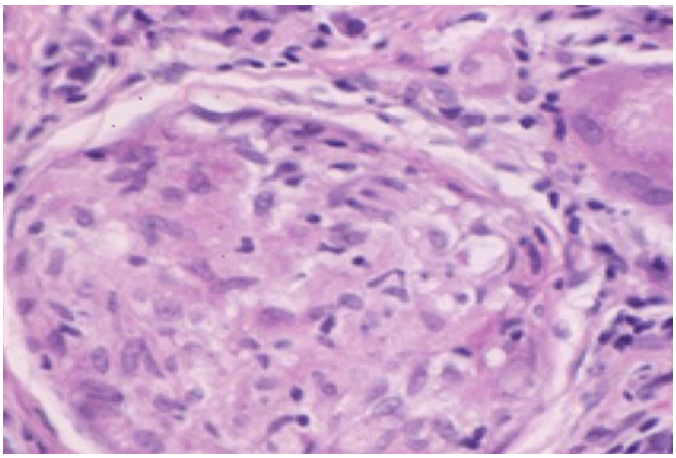

Figure 5: It shows a granulomatous inflammation composed of coalescent granulomata comprising giant cells and histiocytic cells. Background fibro-connective tissue with fibrosis can be seen. No necrosis can be identified. There is no atypia or evidence of malignancy. Special stains are negative for mycobacterial organisms. Impression is granulomatous reaction suggestive of sarcoidosis.

Subcutaneous sarcoidosis presents as painless, firm, mobile nodules without overlying epidermal involvement [11].

Normally, asymptomatic muscle involvement occurs in 50–80% of sarcoidosis patients, whereas symptomatic involvement is very rare [13].

Asymptomatic involvement has been reported in the early stages of the disease. Symptomatic involvement of muscles or myopathy electromyography abnormalities have been reported in chronic and usually the systemic form of sarcoidosis [14, 15].

Three different types of symptomatic muscular involvement in sarcoidosis have been reported which are nodular, acute myositis, and chronic myopathy. However, nodular form of sarcoidosis is a rare condition [5].

Clinically, it may appear single or as many lesions, fleshy, deep seated, non-tender, normally appearing epidermis or hyper-pigmented nodule, tend to coalse to form linear bands [16].

A very few reports shown evidence of a symmetrical muscle contracture that can occur in sarcoidosis [17].

Due to nodular nature of lesion, the differential diagnosis are giant cell tumour of tendon sheath, metastasis, foreign body granuloma, subcutaneous granuloma annulare, xanthoma, rheumatoid nodule, myxoid cyst, lipoma, erythema elevatumdiutinum, fungal or atypical mycobacterial infection, calcinosis, and gout [16].

MRI is helpful in detecting intramuscular nodular sarcoidosis which evaluates the extent and distribution of underlying muscular involvement to monitor steroidal response. Axial spin-echo T1 and T2 contrast enhanced images shows an oval nodule with a star-shaped area of low signal intensity surrounded by homogenous high signal intensity area [18, 19].

Chest Radiograph findings used for staging of pulmonary sarcoidosis based on The Siltzbach classification system. Stage 0 is normal appearance at Chest X-ray; stage 1 is lymphadenopathy only; stage 2 is lymphadenopathy and parenchymal lung disease; while stage 3 is parenchymal lung disease only and stage 4 is pulmonary fibrosis [20].

High-resolution computed tomography is the most sensitive and specific imaging method for the evaluation of sarcoidosis especially in cases where atypical presentation predominates. Nodular pattern is very common which varies in size small to large cavitary lesions mimicking neoplasm. Nodules are distributed peri lymphatic region, involving the peribronchovascular cuffs, interlobular septa, subpleural region, centrilobular areas and the entire length of the fissures.

Apart from nodules, there can be parenchymal or reticular opacity. Air trapping is relatively due to granulomas or fibrosis leading to small airway disease which correlates with restrictive pattern of pulmonary function test. Bronchiectasis is uncommon form of sarcoidosis [21]. Anaemia is uncommon but if present, it is usually anaemia of chronic disease or autoimmune haemolyticanaemia can occur [22]. There can be leukopenia, eosinophilia. Elevated CRP is more associated with more severe form of chronic sarcoidosis which is under research proving better response to infliximab [23, 24]. Hypercalciuria is more common.

Serum ACE levels is safe, simple, non-invasive test. It is elevated in 75% patients of sarcoidosis. It is nonspecific because serum ACE levels can be elevated in other conditions which mimics sarcoidosis. It's relevance in clinical practise to assess response to treatment or compliance is unclear [25].

Histology suggests inflammatory infiltrate of sarcoid granulomas, composed of epithelioid cells with

multinucleated giant cells with sparse lymphocytes limited to the hypodermis, sharply demarcated at the dermo-hypodermic junction. In low-power view, it resembles lobular panniculitis [26]. However, all lesions of sarcoidosis may or may not demonstrate the classic findings [27]. Subcutaneous sarcoidosis is differentiated from erythema nodosum due to several factors with main feature of predominantly lobular rather than septal involvement [28]. Erythema nodosum is distinguished clinically also due to painful, red, raised and non-ulcerative subcutaneous nodules [29].

Management plans varies from no treatment for benign course or not cosmetically displeasing, to specific topical or systemic therapy depending upon extra-cutaneous systemic involvement. Corticosteroids is the cornerstone of cutaneous sarcoidosis [15].

For most non-life-threatening sarcoid organ involvement, starting dose of corticosteroids is 20 to 40 mg for 6 months to 2 years can be weaned to lowest effective dose to avoid potential adverse effects.

If disease is recalcitrant or patient experience side effects of corticosteroids, additional immunomodulatory agents which are steroid sparing such as methotrexate, hydroxychloroquine, azathioprine, and cyclophosphamide can be used.

Methotrexate is 80% effective for skin lesions and has been well studied steroid sparing agent and relatively safe according to long term studies [29].

Hydroxychloroquine has been used with significant response rate in small case series of cutaneous sarcoidosis.

Subjective improvement in chronic cutaneous sarcoidosis is seen in patients who have has been prescribed Thalidomide. Infliximab is used as treatment option for refractory sarcoid patients [30].

There is little exiting data in literature to guide therapy for cytotoxic (i.e, mycophenolate and leflunomide) and biological agents (i.e, rituximab, adalimumab and abatacept) are likely to be tried in chronic sarcoidosis [29].

CONCLUSION

There are various differential diagnosis for this condition. Understanding and applying the process of diagnostic reasoning is important to make sure not to miss serious diagnosis. A variety of therapies have been used for cutaneous sarcoidosis. Therapy should be used to derive therapeutic decisions which maximise patient benefit and acceptance. However, high quality evidence to support the efficacy of these treatments is limited. This case report highlights a rare form of nodular cutaneous sarcoidosis. It will help physician, being a educator by recognising the importance of this rare case in the wider community to develop skills to undertake this role as after good understanding of patient with his illness.

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Author Contributions

Sonali Someek Basu – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Pranav Kumar – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor of Submission

The corresponding author is the guarantor of submission.

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Consent Statement

Written informed consent was obtained from the patient for publication of this case report.

Conflict of Interest

Authors declare no conflict of interest.