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Case Report

Sarcoidosis Treated as Hansen Disease - A Case Report

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ABSTRACT

In a country like Nepal; where leprosy is prevalent, when a patient comes to us with nodular lesions, the first diagnosis is leprosy. A case of sarcoidosis with cutaneous and pulmonary involvement is reported here. In this patient initially the diagnosis of Hansen disease was made and was treated accordingly. As there was no improvement, a clinical suspicion of sarcoidosis. Presence of non-caseating granuloma in histopathology report and findings in chest X-Ray helped to reach in the diagnosis of sarcoidosis and was treated accordingly after which the patient showed significant clinical improvement. This case is presented here to emphasize that though in our part of the world and cutaneous sarcoidosis is rare; Sarcoidosis should be kept as one of the differential diagnosis, if clinically suspicious.

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INTRODUCTION

Sarcoidosis is a systemic granulomatous disease of unknown etiology involving the skin and several organs. The most frequently affected organs are the lymph node, lungs, liver, spleen, skin, eyes, small bones of hands and feet and salivary glands.1 It usually presents between the ages of 20 – 40 years and has a slight female preponderance.[1] Cutaneous involvement is seen in 20 – 35% of patients with sarcoidosis.[2]

CASE REPORT

A 38-year-old male, married, a contractor by profession from Terai region (Saptari) came to our outpatient department with the chief complaints of skin rashes all over the body for 15 months. He was apparently well 15 months ago when he noticed small skin colored lesions 2-3 in numbers over the face. He neglected them for 2-3 months but over the next 5-6 months, he noticed that the initial lesions increased in size and he developed multiple similar lesions over the trunk, upper limb, and lower limbs. They were asymptomatic without itching and pain. There was rapid growth in size as well as the number of lesions over the next 6 months. He did not give the history of any other skin problem before this and he did not have any other types of lesions. There was no history of loss of sensation either over the growths or any other part of the body. There was no history of fever, cough, malaise,

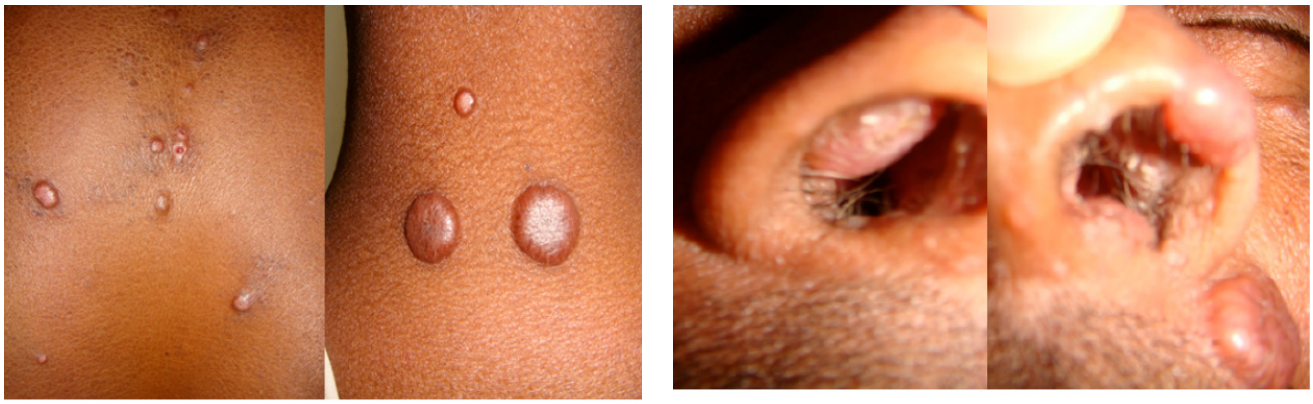

Figure 1A: Multiple skin-colored papules and nodules distributed over back and neck. B. Similar types of papules and nodules inside the nose.

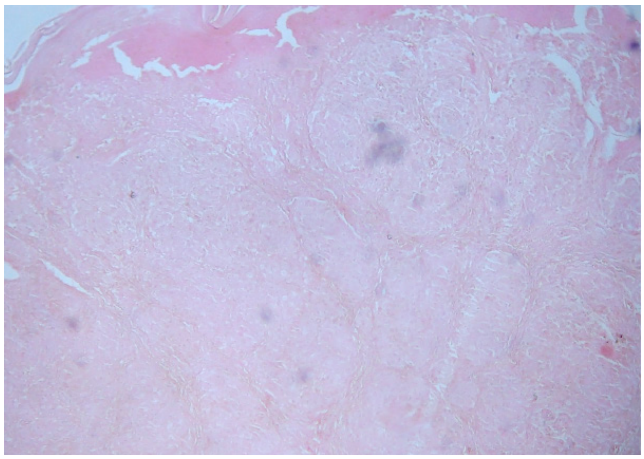

Figure 2: Photomicrograph showing multiple well formed epithelioid cell granuloma without caseation necrosis (HE stain; X50).

weight loss, anorexia, joint pain, epistaxis, pedal edema or visual disturbance. In the family, his father had pulmonary tuberculosis 5 years ago for which he had taken treatment for 6 months. For this problem, this patient had consulted many doctors. Biopsy of one of the nodules was done 5 months ago and histopathologically it showed features of Borderline Tuberculoid Hansen disease. Slit Skin smear from bilateral earlobes, bilateral eyebrows and lesions were negative for acid-fast bacilli. Slit skin smear for LD bodies and RK 39 were negative. With the diagnosis of Hansen disease, he was advised to take Multibacillary Multidrug Therapy (MB MDT). He took the drugs regularly for 4mths without any improvement. Instead, the growths increased in size and number. So, he stopped the medicines and visited some other center. Slit skin smear was repeated again there which again didn't show any acid-fast bacilli. In spite of that, he was advised to take MBMDT again. The patient was not satisfied so he had come to our hospital for further opinion.

On examination, the patient was well built, well nourished, with stable vitals. Examination of skin showed multiple skin-colored papules and nodules varying in size from 3mm to 5cm scattered all over the body. (fig. 1A). These papules were discrete, firm and non-tender. Some nodules were present even inside the nasal cavity (fig. 1B). There were no hypo or hyperpigmented macules or plaques anywhere in the body. The lesions were not anesthetic. Over the hands and feet, glove and stocking anesthesia was not present. Examination of the oral cavity, genitalia, hair, and nail did not reveal any abnormality. On examination of the nerves, they were neither enlarged nor tender.

A skin biopsy was taken from one of the nodules and slit skin smear was also taken from bilateral earlobes, bilateral eyebrows, and skin lesions. The histopathological examination of the sample showed multiple well-formed epithelioid cell granulomas without caseation necrosis (fig. 2). Slit skin smear was negative again.

As we could not come to a definite diagnosis with the above report we advised the patients to get a Chest X-Ray done. The postero-anterior (PA) view of the film showed multiple nodular opacities in both the lung fields. Right hilar and paratracheal lymphadenopathy was also present (fig. 3A). Differential diagnoses were sarcoidosis and tuberculosis. Basic blood investigations, liver function test, kidney function test, serum electrolytes, urine routine and microscopic examinations all were within normal limit except for slightly decreased serum calcium and slightly raised ESR. Mantoux test was also negative. In spite of severe lung involvement in the Chest X-Ray, there were no clinical symptoms of pulmonary tuberculosis. So the provisional diagnosis of Sarcoidosis was made. Pulmonary function test and Electrocardiography was normal. Ophthalmic examination was advised and no abnormalities were detected. With Sarcoidosis as our diagnosis, we started the patient on Tab Prednisolone 40 mg once daily in the morning after food with Omeprazole 20mg once daily on empty stomach.

The patient came for follow up after 45 days when to our surprise all the skin nodules had resolved with only post-inflammatory hyperpigmentation. A repeat Chest X-Ray was done which showed faint reticulonodular shadows in bilateral upper and mid zones with normal hilar shadow and free cardiophrenic angles (fig.3B). Compared with the previous film there was marked the reduction in lung opacities and no evidence of hilar adenopathy. The patient was asked to gradually taper the dose of steroid over next 2 months and stop it. The patient visited regularly for follow up once every 2 months for next 12 months without recurrence of the lesion.

DISCUSSION

Sarcoidosis can affect any organ besides the skin and is capable of imitating a variety of diseases. So it is often called " The Great

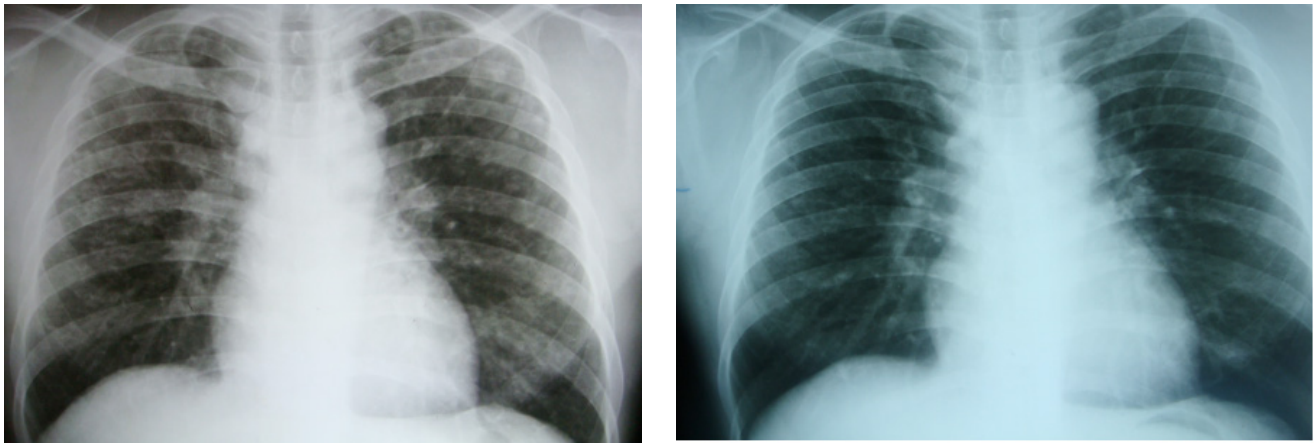

Figure 3A: PA view of Chest X-Ray showing multiple nodular opacities in both the lung fields with right hilar and paratracheal lymphadenopathy. B. Same patient's Chest X-ray after 1.5 months of treatment. X-ray shows faint reticulonodular shadows in bilateral upper and mid zones with normal hilar shadow

Imitator" or "Clinical Chameleon".[3] Sarcoidosis in the skin can present with various clinical manifestations like lupus pernio, subcutaneous nodular, papular sarcoidosis, scar sarcoidosis, plaque sarcoidosis, ulcerating sarcoidosis, small disseminated nodular sarcoidosis, erythema nodosum, ichthyosiform sarcoidosis and erythrodermic sarcoidosis.[3] There are reports of skin sarcoidosis mimicking seborrheic dermatitis, porokeratosis of mibelli and many other skin disorders.[4-6] So diagnosing it is not always easy. Cutaneous sarcoidosis is notably rare in Asian population.[4] Besides, as other granulomatous disorders like leprosy and tuberculosis are more common in Indian subcontinents sarcoidosis is underdiagnosed due to lack of an index of suspicion for this disease.[7] In our patient also that could be the cause of delay in diagnosing and persistently asking him to take treatment for leprosy.

Recognizing skin lesions is important in diagnosing sarcoidosis as they can provide a visible clue to diagnosing sarcoidosis and skin is an organ which is very accessible for histologic examination.[2] Histopathologically the specific lesion of sarcoidosis shows aggregates of epithelioid histiocytes with few or no inflammatory cells and these are the so-called naked or sarcoidal non caseating granulomas.[8] However, these findings are not specific for sarcoidosis as it can be present in many other conditions. This may be the reason why it may be difficult for the pathologist to make a definite diagnosis of sarcoidosis only with histopathological findings as in our patient.

There have been few reports from India where sarcoidosis was masqueraded by Tuberculosis, Borderline Tuberculoid Leprosy, and Borderline leprosy.[9-11] So when we come across cutaneous lesions infiltrated with granulomas we need to keep our eyes open and not only think about the infective causes but also rule out diseases like sarcoidosis. Besides this case report is an example to emphasize the fact that skin could be a marker of internal diseases.

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