Case Report

Leprosy or sarcoidosis? A diagnostic dilemma!

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ABSTRACT

There are two important differentials for non-caseating granulomatous inflammatory tissue—leprosy and sarcoidosis—which presents a diagnostic challenge due to their histological similarities and specific geographical distribution. This article describes a rare presentation of systemic sarcoidosis called Heerfordt’s syndrome with the triad of parotitis, uveitis, fever, and optional paralysis of facial nerve. This case was initially diagnosed as leprosy.

Keywords: Leprosy, non caseating granuloma, sarcoidosis

Introduction

Non-caseating granulomatous inflammatory tissue has two important differentials—leprosy and sarcoidosis. Due to this shared similarity between the two, they often get misdiagnosed as one for the other.¹⁻⁴ The two diseases also have specific geographical prevalence and are often misdiagnosed in the countries where they are not prevalent. We present a rare case of systemic sarcoidosis in the form of Heerfordt’s syndrome, which initially misdiagnosed as tuberculoid leprosy.

Case Report

In 2011, a 28-year-old male patient presented with asymptomatic pigmented lesions on dorsum of the left foot since 3–4 months. He gave a history of a solitary lesion that gradually progressing in number with no itching, burning, pain, or oozing from the lesions. There were no associated systemic complains. The cutaneous lesion on biopsy showed a non-caseating tuberculoid granuloma suggestive of tuberculoid leprosy and was, thus, diagnosed as tuberculoid leprosy on the basis of the clinical picture, burden of new leprosy cases in India,⁵ and the biopsy report. He was started on paucibacillary leprosy treatment.

After anti-leprosy treatment, the patient seemed alright for 2 years but came back in February 2014 with symptoms of three cranial nerve involvement—difficulty in swallowing, suggestive of nerve IX and nerve X involvement, and drooping of right eyelid, suggestive of nerve V involvement. The patient also had hypotension—probably suggestive of X nerve involvement. He also had bilateral parotid swelling with fever and conjunctival redness. The fluorodeoxyglucose-positron emission tomography (FDG-PET) scan showed metabolically active bilateral, level IV cervical, mediastinal, hilar, and peribronchial lymph nodes with multiple lung nodules and left common iliac adenopathy [Figure 1a-c]. On the basis of this report, a differential diagnosis of sarcoidosis or lymphoma was given. The excision biopsy of the cervical lymph node showed as previously discrete and confluent typical tubercles with no caseous necrosis. Ziehl-Neelsen (ZN) stain was negative for acid fast bacilli (AFB). However, Fite-Faraco stain showed one stout bacillus. MRI BRAIN showed prominence of lateral ventricles with no other significant pathology identified. Due to the positive Fite-Faraco stain and previous diagnosis of leprosy, the patient was restarted on anti-leprosy treatment. The patient was continuously on anti-leprosy treatment (as prescribed by the treating neurophysician) due to the reporting of AFB in histopathology. However, the patient did not get better and came back in October 2014 presenting with headache associated with nausea and vomiting since a month and a half, fever with chills for 8 days, and cough since 5 days. Patient was investigated and CT scan of

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chest showed right-sided basal pleural effusion and lower lobe subpleural consolidation with plate-like atelectasis with extensive mediastinal lymphadenopathy. The laboratory tests showed an elevated serum angiotensin-converting enzyme (SACE) level 62 U/L (normal range 8–53 U/L), raised serum calcium level 12.5 mg/dl (normal range 8.5–10.2 mg/dl), and urinary protein 976 mg/24 hours (normal range >80 mg/24 hours). The rest of the laboratory reports including peripheral smear for Malarial parasite, Dengue Serology, Widal, Mantoux, and Blood culture were negative. As the patient did not get better on anti-leprosy drugs and taking into consideration the triad of b/l parotitis swelling, uveitis, fever along with neurological involvement, incriminating FDG-PET scan, and elevated SACE levels, the final diagnosis of systemic sarcoidosis in the rare form of Heerfordt’s syndrome was made and patient was started on steroids.

Discussion

India has been saddled with leprosy for a long time and is one of the countries in which the highest number of new leprosy cases is detected every year. It is often the first diagnosis considered when the pathology of non-caseating granulomas is detected. Similarly, in our case, the patient was diagnosed with leprosy the first time as he presented no other symptoms or signs other than asymptomatic pigmented skin lesions, which on biopsy showed non-caseating granulomas. The patient came back after 3 years with symptoms of fever, b/l parotitis, uveitis, and neurological involvement. It is quite possible that this was a rare presentation of systemic sarcoidosis called Heerfordt’s syndrome. That this was systemic sarcoidosis, was also supported by the presence of b/l hilar lymphadenopathy with nodes displaying non-caseating granulomas, on biopsy. When the tissue was subjected to Fite-Faraco stain, the pathologist felt he saw a single leprosy bacillus. In view of the fact that this patient was initially diagnosed as leprosy, and in view of the report of the pathologist authenticating a bacillus on Fite-Farraco stain, the neurophysician, the PETSCAN physician, felt convinced about the diagnosis of leprosy, the patient was restarted on leprosy treatment. The dermatologist, at that time, had categorically conveyed to the neurophysician, after consulting other colleagues expert in the knowledge of leprosy, that this case is unlikely to be leprosy, cranial nerves involvement and multiple glands in the chest seen on PETSCAN go against the diagnosis of leprosy. The patient came back the same year with headache associated with naussea and vomiting, weight loss, since a month and a half, fever with chills for 8 days, and cough since 5 days. This time the SACE levels were done and they were found to be very high. Thus, taking into consideration the previous history of cutaneous lesions showing tuberculoid granulomas, the cervical gland biopsy showing tuberculoid granuloma, the PETSCAN showing peribullary glands, the triad of b/l parotitis, uveitis, and fever along with neurological involvement and with the biochemical reports, a diagnosis of Heerfordt’s syndrome which is a rare presentation of systemic sarcoidosis was established. He was started on steroids after which he showed continual improvement. Thus, this is a cautionary case history where the pathological evidence, if unsupported by clinical evidence, should not be taken as the sole basis of diagnosis. Pathology alone should not dictate the entire picture and a clinicopathological correlation should be made.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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