CASE REPORT

A Rare Case of Disseminated Sarcoidosis Presenting with Neurosarcoidosis: A Case Report

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ABSTRACT

Sarcoidosis is characterized by formation of inflammatory granulomas affecting all over the body, with pulmonary predilection (1). Neurosarcoidosis is a rare but potentially dangerous manifestation of sarcoidosis. We report a case of disseminated sarcoidosis presenting with a neurological diagnostic dilemma. Worsening mediastinal lymphadenopathy, together formation of lung and liver nodules making a sarcoidosis diagnosis favourable. Histology from these lesions showed non-caseating granulomatous inflammation. She was treated as a rare case of disseminated sarcoidosis. To date, there is no specific or clear guideline on the management of disseminated sarcoidosis.

Keywords: Disseminated sarcoidosis, Neurosarcoidosis, Sarcoidosis

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INTRODUCTION

Sarcoidosis is a rare, multi-system condition that affected mostly the pulmonary and lymphatic systems. The hallmark of sarcoidosis is relapsing chronic inflammation by formation of non-necrotising granulomas that can sometimes congregate and formed inflammatory bodies or masses. Eventually these inflammations will almost lead to fibrosis, without treatment. When affecting the nervous system, the disease is coined as neurosarcoidosis. The remitting and relapsing inflammation is causing intermittent symptoms, making it difficult to diagnose as there are plenty other disease that of neural tissue origin presented similarly. Tissue histopathological examination is the gold-standard for diagnosis however it is almost clinically impractical to biopsy central nervous tissues. A combination of clinical acumen, expertise, and experience should help to diagnose neurosarcoidosis affecting central nervous system.

Disseminated sarcoidosis is when the inflammation occurs throughout the systems. Currently there is a debate whether treatment should be escalated in this type of sarcoidosis in cases without symptoms.

CASE REPORT

A 43-year old teacher presented with recurrent fleeting lower motor neuron facial nerve palsies initially on the right side, resolved, then on the left side, bulbar weakness with swallowing difficulties, and impaired hearing of the left ear with left-sided balance problem. Multiple cranial nerve palsies involving bilateral facial, vestibulocochlear and glossopharyngeal nerves suggestive of either myasthenia gravis, multiple sclerosis, or neurosarcoidosis. She received treatment for myasthenia gravis and benign paroxysmal positional vertigo, to which she responded.

Repetitive nerve stimulation and nerve conduction studies were normal. Serum acethycholine receptor antibody was not detected. Serum angiotensin converting enzyme (ACE) level was normal. Brain magnetic resonance imaging revealed non-specific foci in both parietal lobes convexities. Serial chest x-ray showed worsening widened mediastinum (Fig. 1). Serial thorax computed tomography (CT) scan showed enlarged mediastinal lymphadenopathies over the course of 3 (Fig. 2) years with new development of bilateral lung nodules, and left breast and liver lesions. Echocardiogram was normal. A positron emission tomography – computer tomography (PET-CT) scan highlighted hypermetabolic foci in lung, mediastinal nodes and liver (Fig. 3). Despite this, she had no gastro-intestinal symptoms. Later, her serial lung function test showed reducing forced vital capacity and

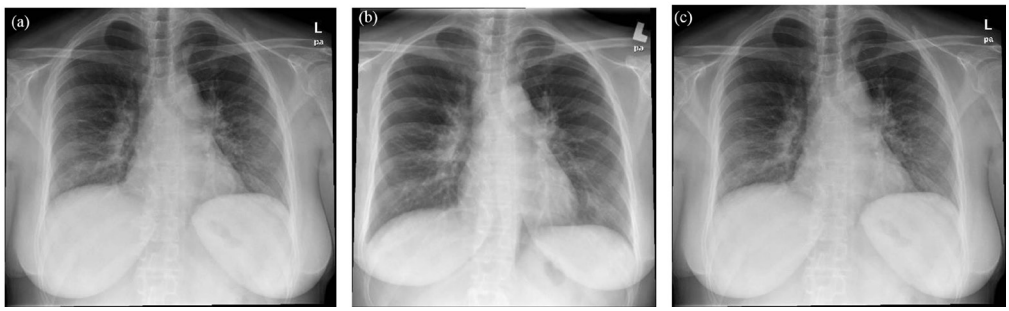

Figure 1: Serial chest X-ray. 2014 (left), 2016 (middle) and 2018 (right) chest X-ray showing worsening mediastinal widening.

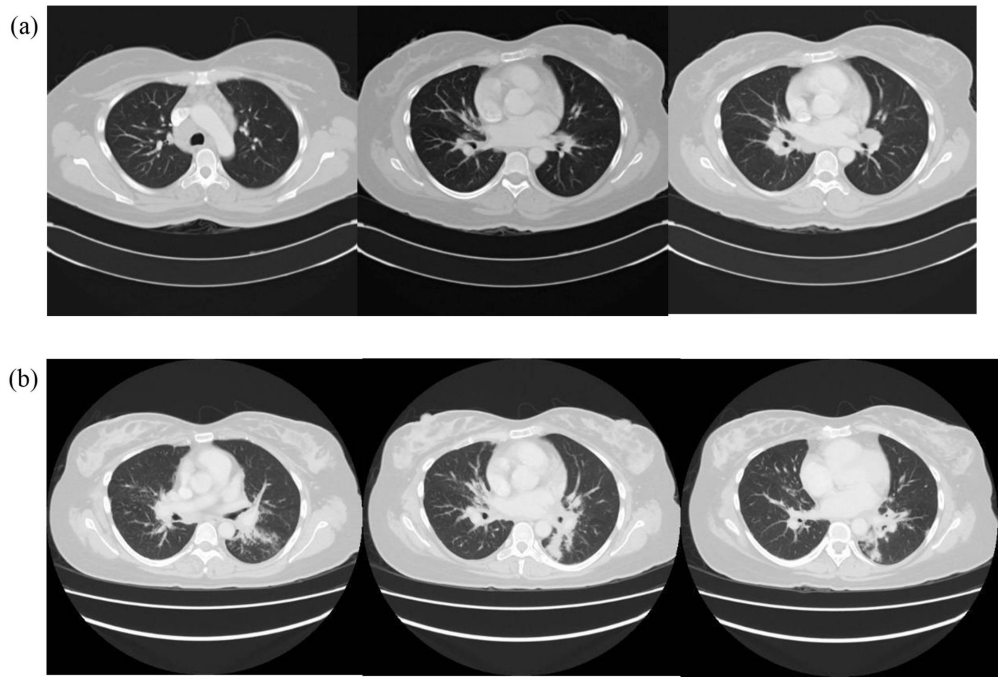

Figure 2: Serial CT thorax. 2014 (above) and 2018 (below) CT thorax showing worsening mediastinal lymphadenopathy.

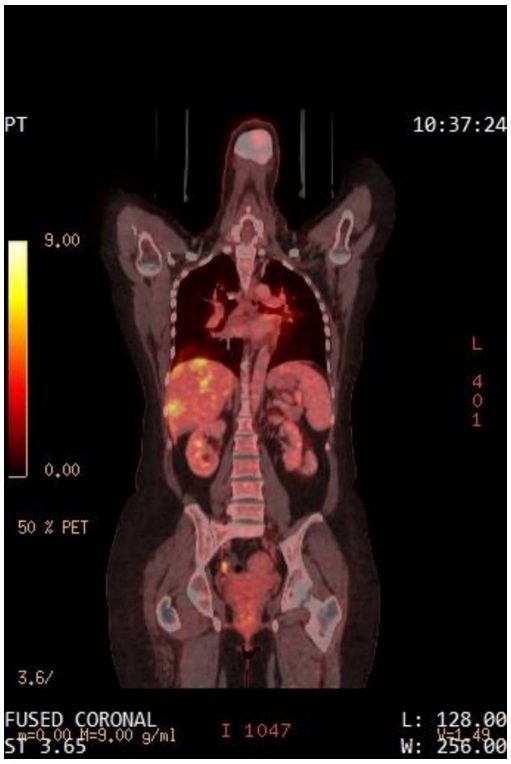

Figure 3: Coronal plane whole body PET-CT. The Multiple hypermetabolic foci over the liver, lung parenchymal, and mediastinal lymph nodes.

diffusing capacity and developed an obstructive airway disease picture.

Endobronchial ultrasound transbronchial node aspirations were done twice, to which both yield inconclusive results. CT-guided lung and liver biopsy showed non-caseating granulomas (Fig. 4). Excision biopsy of left breast lump revealed fibroadenoma. Tuberculosis investigations were negative.

High dose prednisolone was started, which slowly wean down later together with commencement of azathioprine as steroid-sparring agent. She tolerated the regime well. Her lung functions were improved. However, her chest radiograph remained the same.

DISCUSSION

The presence of non-caseating granulomas is characteristics of sarcoidosis that may involve multiple sites in the body (1). Neurosarcoidosis has a variable presentation that can either occur in isolation or together with other sarcoidosis clinical features. It can affect at any level of central and peripheral nervous system, including peripheral nerves and even muscles (2). Spontaneous remission is expected in up to 60% of the patients but relapsing episodes are well-documented.

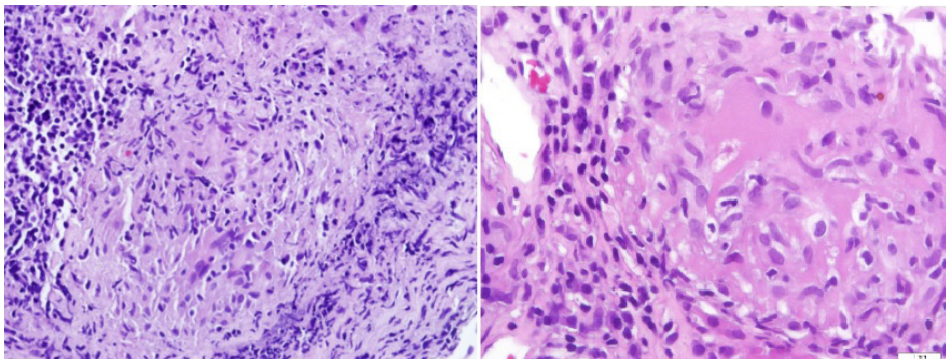

Figure 4: Histopathological examination of lung and liver. Non-caseating granuloma from lung (left) and liver (right) biopsies.

Hepatic involvement in sarcoidosis is common, amounting up to 60% of the patients. Many are symptomatic with on-specific abdominal and gastro-intestinal symptoms, but a good portion of patients are asymptomatic – in which case, no treatment is advocated (3). Liver biopsy, like in our patient, is usually required to arrive at the diagnosis. Hepatic sarcoidosis may result in portal hypertension and liver cirrhosis.

In pulmonary sarcoidosis, a restrictive defect with reduced lung volumes is seen, however diffusing capacity for carbon monoxide (DLCO) is the more sensitive pattern. Airflow obstruction occurs in 30 to 50% of patients. One study highlighted that up to 57% of patients may have a decrease of FEV1:FVC ratio at presentation, followed by reduction of DLCO (27%) and rather surprisingly, restrictive pattern only seen in 6% (4). This finding is in keeping with our patient.

Regarding treatment, there is no specific guideline for management of disseminated sarcoidosis (5). Corticosteroid is the mainstay of treatment. Steroid-sparring agents should be considered in such cases with steroid-resistant disease, intolerable adverse corticosteroid side effects, or patient desire not to take corticosteroids. Example of these agents are methotrexate, chloroquine and hydroxychloroquine, azathioprine, and chlorambucil.

The decision of treatment in our patient was based on worsening physiological lung functions. As there is no specific guideline for the management of disseminated sarcoidosis, we faced a challenge to start treatment initially. Finally, she responded well to steroid therapy with a combination of steroid sparing agent.

CONCLUSION

Our patient presented initially with an uncommon presentation of sarcoidosis (neurosarcoidosis) which subsequently progressed into disseminated sarcoidosis. Clinical and radiological features of tuberculosis and presence of breast lump was making it difficult to reach the final diagnosis. She responded well to steroid therapy and we manage to reduce the steroid to a low maintenance dose with an add-on one steroid-sparing agent.

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