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Sarcoidosis: A rare disease in Malaysia

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Abstract

Sarcoidosis is a systemic granulomatous disease of unknown etiology that is rarely reported in Malaysia. Diagnosing sarcoidosis is challenging in this setting given the similar clinicoradiology presentation with more common diseases. This may in turn results in under-diagnosis of sarcoidosis. Here we report a case of 27-year old Malaysian Indian gentleman who was diagnosed with sarcoidosis after incidental findings of bilateral hilar lymphadenopathy on chest radiograph. We hope that this case report will help to raise the awareness of this disease in Malaysia. Though rare in our community, it should still be considered in the differential diagnosis of patients after excluding pulmonary tuberculosis which is endemic and lymphoma.

Keywords

sarcoidosis; non-caseating granuloma; bilateral hilar lymphadenopathy; pulmonary tuberculosis; lymphoma

Abbreviations

BHL: bilateral hilar lymphadenopathy; CECT: contrast-enhanced computed tomography; EBUS: endobronchial ultrasound guided; ACCESS: A Case Control Etiologic study of sarcoidosis

Introduction

Sarcoidosis is a systemic granulomatous disease of unknown etiology that may involve multiple organs. It occurs through out the world, affecting both sexes, all ages and races. The diagnosis is achieved when clinical and radiological findings are supported by pathological evidence of non-caseating epithelioid cell granulomas [1,2].

While it is a common disease in western countries [3], it is rarely being reported in Malaysia [4]. Diagnosing sarcoidosis is challenging in this setting given the similar clinicoradiology presentation with more common diseases in Malaysia such as pulmonary tuberculosis which is endemic and lymphoma. Here, we report a case of an asymptomatic Malaysian Indian gentleman who had incidental findings of bilateral hilar lymphadenopathy (BHL) on chest radiograph which was later confirmed to be sarcoidosis.

Case Presentation

A 27-year-old Malaysian Indian gentleman with no underlying medical illness and active

complain was initially presented to our center for a pre-employment medical examination. In general appearance, he was a healthy looking young man. The physical examination was unremarkable. Routine blood investigations (full blood count, renal profile and liver function tests) were normal.

A routine chest radiograph was also performed and he was found to have incidental findings of bilateral hilar lymphadenopathy (Figure 1). A contrast-enhanced computed tomography (CECT) of the thorax and subsequently CECT neck, abdomen and pelvis were performed for further evaluation, revealing symmetrical hilar lymph nodes enlargement as well as mediastinal lymphadenopathy (paratracheal, subcarina, aortopulmonary, precarina and preaortic regions) (Figure 2). There were also multiple small nodules noted along the peribronchovascular bundle in the posterior segment of both lower lobes (Figure 3). In addition, there were enlarged lymph nodes in the abdomen (porta hepatis, paraaortic and aortocaval regions) and cervical region (right level I). Hepatosplenomegaly and multiple ill-defined hypodense lesions were also seen at the spleen (Figure 4).

Tuberculosis work-up were negative. An endobronchial ultrasound guided (EBUS) transcarinal needle aspiration of the subcarinal lymph nodes was performed and the cytology revealed granulomatous inflammation with no acid fast bacilli or amyloid deposition seen.

Correlating the clinical, radiological and cytology findings, the diagnosis of sarcoidosis was established. In view that he was asymptomatic with no vital organ involvement (such as eyes, brain or heart), it was decided to follow up the patient with imaging in 6 months (in accordance with a consultant respiratory physician) to allow spontaneous resolution of the lesions. Corticosteroids are kept in view as first line therapy in case of disease progression.

Discussion

Sarcoidosis a common disease in the west. The ACCESS (A Case Control Etiologic Study of Sarcoidosis) study reported incidence of 35 to 80 per 100,000 among African Americans, 15 to 20 per 100,000 among Northern Europeans and 3 to 10 per 100,000 among European Americans [3]. In Asia, the incidence in Japan is 1 to 2 per 100,000 [3] and in Singapore, the estimated annual incidence is 0.56 per 100,000 [5]. Few studies have shown that there is a female predilection of the disease [1,3,6]. The disease is more common in adults less than 40 years of age, peaking in those 20 to 29 years old [1].

The epidemiology of sarcoidosis in Malaysia is largely unknown due to its rarity. Sarcoidosis is rarely reported in Malaysia. In a study by Liam CK et al in 1993, there was a total of 14 patients from 1976 to 1990 [4]. In another more recent updated study by Rahim ZAB et al in 2002 at the same center, there was a total of 36 patients from 1987 to 2001 [7]. Table 1 shows the comparison between the gender and ethnicity in these two studies which shows slight female predominance and more common in Indians.

This disease remains a radiological challenge in Malaysia due to its rarity. Imaging findings may provide clues to its diagnosis but may not clearly distinguish it from more common diseases such as lymphoma and tuberculosis which is endemic. In this part of discussion, we will focus on the typical radiologic findings in sarcoidosis, particularly the thoracic region as pulmonary and mediastinal involvement is seen in over 90% of patients [1,2,8].

Thoracic lymphadenopathy is the most common radiologic finding in sarcoidosis (85% of cases) [8]. Various studies have shown that the most common pattern of lymphadenopathy in sarcoidosis is

middle mediastinal lymph nodes (particularly left paratracheal, subcarinal, and aortopulmonary window region), prevascular nodes or both are involved in approximately half of the patients [11]. It is estimated that 95% of patients with sarcoidosis presents with the characteristic BHL with or without concomitant mediastinal lymphadenopathy [9,11].Isolated unilateral hilar lymphadenopathy (more commonly on the right) is only seen in less than 5% of cases [11]. Lymph nodes in sarcoidosis are usually non-necrotic and non-compressive [10]. Amorphous, punctate, or eggshell-like calcifications can be seen frequently in longstanding disease [8,10].

In a review by Winterbauer et al, symmetrical BHL was the mode of presentation in only 3.8% of lymphomas [13]. Isolated mediastinal enlarged lymph nodes without BHL is more suggestive of other diagnosis, particularly lymphoma. BHL in absence of clinical symptoms is strongly suggestive of sarcoidosis [10]. Pulmonary tuberculosis lymph node involvement is typically unilateral and right sided, involving the hilum and right paratracheal region although it is bilateral in about a third of cases. Lymph nodes often measures more than 2cm with a characteristic low-attenuation center with peripheral rim enhancement representing granulomatous inflammatory tissue with central necrosis [14,15].

The most characteristic parenchymal pattern in sarcoidosis is 2 to 4mm small rounded nodules in a perilymphatic distribution (along the subpleural surface, interlobular septa and peribronchovascular bundle) [8,10,11]. They are seen in 75 to 90% of patients with pulmonary sarcoidosis, usually in the upper and middle zones [8,11]. Frequently, these nodules cause irregular thickening of interlobular septa and peribronchovascular bundles [8, 11]. These pattern of distributions, in the presence of thoracic lymphadenopathy is strongly suggestive of sarcoidosis [8].

On the other hand, post-primary tuberculosis typically presents with 2 to 4mm centrilobular nodules with tree-in-bud appearance and cavitations [14,15]. Other parenchymal manifestations of pulmonary sarcoidosis includepatchy fibrotic changes(linear opacities, traction bronchiectasis, architectural distortion) predominantly in the upper and middle zone is usually seen in advanced stage of disease, however may also be seen in other diseases such as tuberculosis[8,10,11].

In this case, there is also an additional findings of splenic nodules. Splenic involvement in sarcoidosis is seen in approximately 40-80% of patients [2,16,17]. Splenomegaly is seen in about one third of patients with 15% having multiple small ill-defined hypodense non-enhancing nodules [16,17,18]. However, these findings are not specific as differential diagnosis may include lymphoma whereby spleen is involved in 30 to 40% of patients and tuberculosis [18,19]. As such, correlation with other clinical and radiological findings is important.

In our patient, given the radiological findings particularly homogeneous symmetrical bilateral hilar lymphadenopathy and perilymphatic distribution of micronodules, in addition to patient being asymptomatic, points more towards the diagnosis of sarcoidosis instead of lymphoma. Tuberculosis is less likely as the patient's tuberculosis workup is negative. Supported by the pathological findings of granulomatous lesion, the final diagnosis of sarcoidosis is established in this patient.

Conclusion

Sarcoidosis is a rare disease in Malaysia and is rarely reported. We hope that this case report will help to raise the awareness of this disease in Malaysia. In all probabilities, this disease is probably overshadowed by the presence of lymphoma and in particular tuberculosis. In such circumstances, though rare in our community, sarcoidosis should be kept in mind as one of the differential diagnosis.

Figures

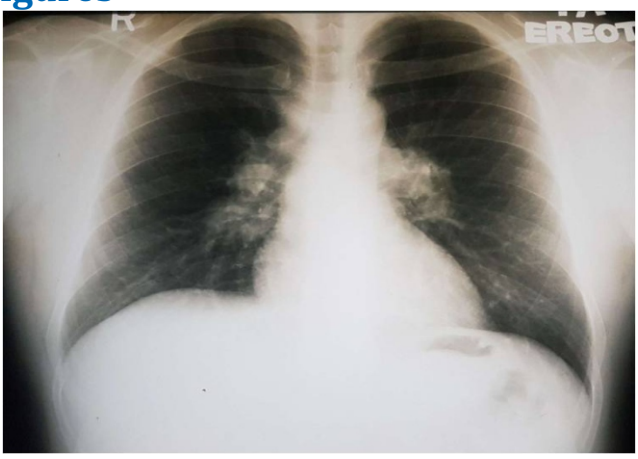

Figure 1: Chest radiograph (posteroanterior) showing bilateral hilar lymphadenopathy.

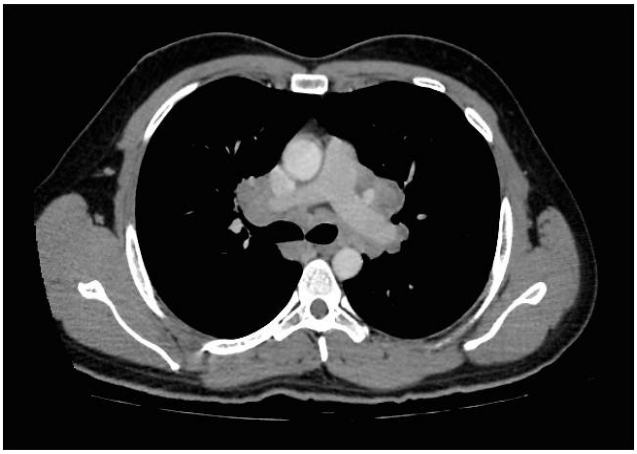

Figure 2: Contrast enhanced CT (axial view) showing bilateral hilar lymphadenopathy.

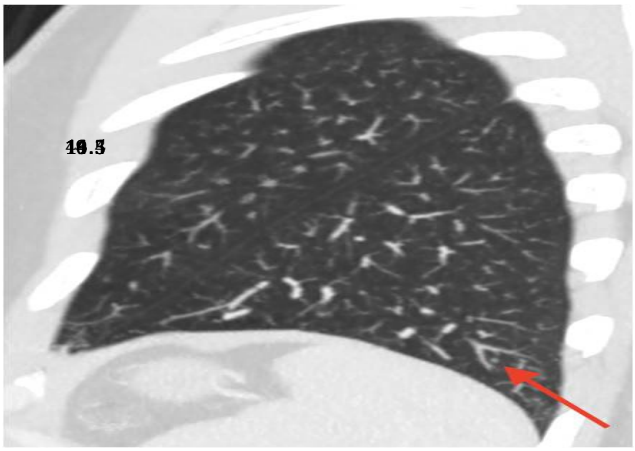

Figure 3: Contrast enhanced CT (sagittal view) showing a small nodule along the peribronchovascular bundle (red arrow).

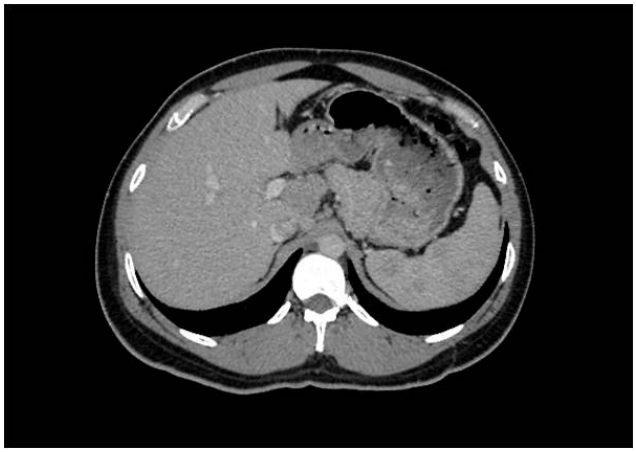

Figure 4: Contrast enhanced CT (axial view) showing multiple ill-defined hypodense lesions in the spleen.

Tables

Table 1: Comparison between gender and ethnicity of two studies

Study	Liam CK et al		Rahim ZAB et al	
	n = 14	%	n = 36	%
Sex				
Male	7	50	16	44.4
Female	7	50	20	55.6
Ethnic				
Malay	2	14.3	7	19.5
Chinese	2	14.3	4	11.1
Indian	10	71.4	25	69.4

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