Case Report

Multifocal micronodular pneumocyte hyperplasia: not all miliary nodules are due to infection / granulomas / metastases: a case report

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Abstract

Multifocal micronodular pneumocyte hyperplasia is an unusual pulmonary manifestation in tuber sclerosis, and is due to hamartomas of the type II pneumocytes in the alveolar septa. It manifest as multiple miliary nodules in the lungs, with a predominant upper lobe distribution. Further work up and treatment for these nodules is unnecessary as these nodules don't progress to cause clinical disease. Here, we present a 13 year old girl with tuberous sclerosis who presented with on and off breathlessness for 2 month duration and miliary nodules on Computed tomography of the thorax, which was diagnosed as Multifocal micronodular pneumocyte hyperplasia.

Keywords: Tuberous sclerosis [TSC], Multifocal micronodular pneumocyte hyperplasia [MMPH], CT thorax, miliary nodules, diffuse micronodular lung pattern.

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INTRODUCTION

Tuberous sclerosis [TSC] is an autosomal-dominant disorder that manifests clinically as a triad of seizures, mental retardation and adenoma sebaceum. The classical vogt's triad is seen only in 33% of the patients. This disorder is characterized by the presence of hamartomas in the skin, brain, heart and kidneys. Hamartomas may also be seen in the lungs, retina, gastrointestinal tracts and bones.[1] Although hamartomas in the brain and kidneys are common, hamartomas in the lung are rare[2]. Amongst the hamartomas in the lungs, lymphangiomyomatosis [LAM] is a more commonly reported radiologic manifestation than multifocal micronodular pneumocyte hyperplasia [MMPH][3,4]

CASE REPORT

A 13 year old girl presented with complaints of breathlessness on and off for the past 2 months. Patient was diagnosed as tuberous sclerosis at young age [patient had seizures from birth and also had multiple skin lesions]. Chest radiograph showed multiple nodular densities in both lungs and thus computed tomography of the thorax was requested for. CT of the thorax showed multiple randomly scattered micronodules [2-4 mm] in both lungs, with a predominant upper lobe distribution [figure 1 (a-d)]. CT of the brain showed multiple calcified subependymal nodules along the walls of the lateral ventricles [figure 2 (a, b)].With the background clinical diagnosis of tuberous sclerosis and the CT findings of characteristic diffuse micronodular disease pattern, the diagnosis of multifocal micronodular pneumocyte hyperplasia was made.

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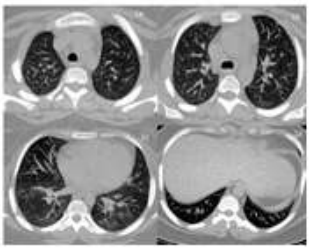

Figure 1 (a-d): CT sections of the thorax show multiple r andomly distributed micronodules in both lungs, more in the upper

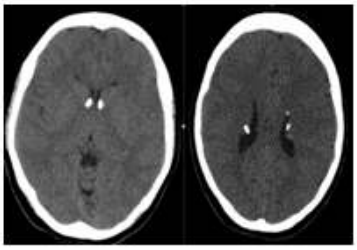

Figure 2 (a-b): CT sections of the brain show multiple calcified subependymal nodules along the walls of lobes the lateral ventricles.

DISCUSSION

MMPH is a rare lung manifestation in TSC. It was first described by popper at al[5]. It presents on CT as multiple tiny randomly distributed pulmonary nodules [diffuse micronodular pattern]. Differential diagnosis for diffuse micronodular pattern in the lungs is given in table 1.

Table 1

Infections	Miliary tuberculosis
Fungal infections	histoplasmosis, blastomycosis,cryptococcosis, coccidioidomycosis, Varicella pneumonia
Pneumoconiosis	Coal worker's pneumoconiosis, silicosis, berylliosis
Inflammatory	Sarcoidosis
Malignancy	Metastasis (thyroid carcinoma, osteosarcoma)
Others	Alveolar microlithiasis, amyloidosis

In MMPH, the nodules are characteristically distributed randomly in the secondary lobules with a higher predilection for the periphery of the lungs and upper lobes of the lungs[6]. LCH is the closest differential. in the early phase of LCH, centrilobular nodules are the dominant presentation. The nodules are seen on the inferior margins of the lung. Micronodules in the MMPH are benign hamartomas of the type II pneumocytes along the alveolar septa. They exhibit fibrous thickening, increased elastic fibers and aggregated alveolar macrophages[7,8]. There is no known malignant potential in the nodules[8].

CONCLUSION

MMPH should be considered in the differentials for diffuse micronodular lung pattern on CT thorax in patients known to have TSC. Recognition of this entity will avoid unnecessary further work up and wrongful treatment, as it is unlikely for the nodules to progress to clinically significant disease[9.10,11].

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