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Case Report

Generalized Lymphangiomatosis: Rare Hepatic and Pulmonary Presentation

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Keywords

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Abstract

Lymphangiomas are rare benign neoplasms believed to be result of abnormal development of lymphatic system. They grow very slowly usually localized to one organ, but occasionally involve several organs in one part of the body. Reports of single organ involvement have been published relating to nearly all organs of the body except CNS with certain predilection tocervicothoracic region. Lymphangiomatosis or multifocal/ diffuse lymphangiomas presented during childhood mainly in the head and neck region; less than 5% are intra-abdominal. In one large series, lymphangioma of head and neck accounted for nearly 50% of cases in children, while 10% of cases had visceral disease including thorax. We report 4-year-old child with lymphangiomatosis having *cannon ball opacities* on chest radiograph and *simultaneous pulmonary and hepatic involvement* which has never been reported and is unusual presentation.

ABBREVIATIONS

CECT: Contrast Enhanced Computed Tomography, **CNS:** Central Nervous System; **CT:** Computed Tomography: **MRI: M**agnetic Resonance Imaging; **MUAC:** Mid-Upper Arm Circumference; **PT-INR:** Prothrombin Time-International Normalised Ratio; **SGPT**-Serum Glutamic Pyruvic Transaminase; **SGOT**: Serum Glutamic Oxaloacetic Transaminase.

INTRODUCTION

Lymphangiomas are rare slow growing benign neoplasms due to abnormal development of lymphatic system localized to one organ, but occasionally involve several organs in one part of the body except CNS [1-3]. The diagnosis of this condition may be achieved by CT scan without any need for biopsy due to characteristic findings [4].

The plain chest radiograph will reveal either a localized lung lesion, with solitary type, or diffuse pulmonary interstitial infiltrate with or without thoracic lymphadenopathy, pleural or pericardial effusion [1,5]. Parenchymal lung involvement with lymphangiomas has a characteristic CT appearance seen as lobular or cystic mass of heterogenous soft tissue density in the localized type or diffuse involvement, seen as patchy areas of ground glass appearance. Using conventional and high resolution CT scan, Swensen et al, reporting on eight patients with diffuse disease concluded that the appearance is distinctive and includes diffuse, smooth thickening of interlobular septa and bronchovascular bundles with extensive infiltration of mediastinal fat and associated perihilar infiltration. Pleural thickening and /or effusion were seen in seven of eight patients. Despite the typical appearance they stressed the importance of biopsy in a definite diagnosis [1,6].

Despite our intensive search and our best knowledge till date, no case of diffuse lymphangiomatosis has been reported with cannon ball appearance on chest radiography and simultaneous involvement of lung and liver without any clinical symptoms but presented with unrelated febrile convulsions.

CASE PRESENTATION

This 4 year male child came with complaints of high grade pyrexia of 3 days, intermittent, without chills and rigors and1st episode of generalised involuntary movement with uprolling of eyeballs, frothing from mouth, clenching of teeth lasting 5 minutes followed by unconsciousness for 1 hour. He was first son of 26yr mother and 29yr old father with h/o first degree consanguinity present. Past, family and personal history was non-significant.

The child was conscious, oriented. Weight, height, occipito-frontal circumference and MUAC were appropriate for age. The neurological examination was normal. Abdominal examination showed visible dilated veins with slight distension, palpable liver of size 5 cm, sharp borders, nodular surface (Figure -1; 3 large nodules palpable), liver span 13cm and spleen was not palpable. Cardiovascular and respiratory system examination was normal.

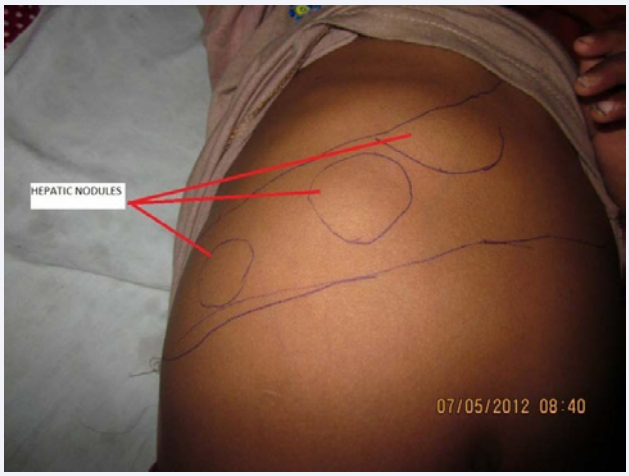

Figure 1 Massive Hepatomegaly with nodules.

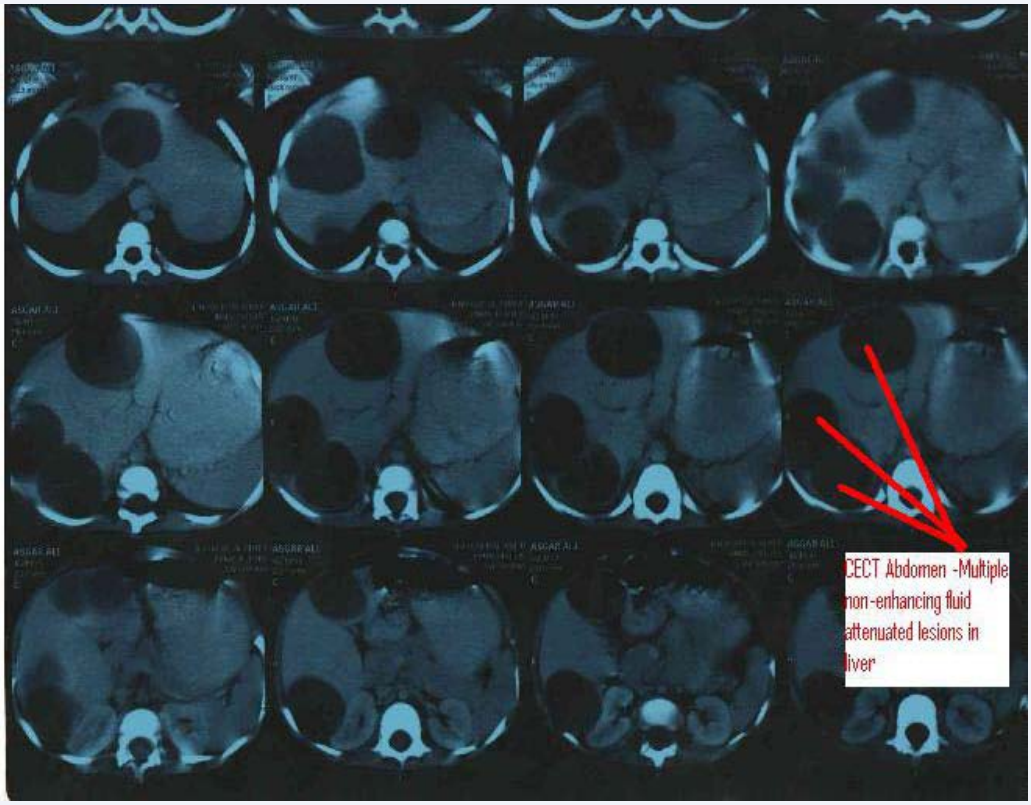

Figure 2 CT Abdomen: showing hepatic lesions.

Blood and other investigation are as shown in Table 1. CECT Abdomen and thorax-Multiple non-enhancing fluid attenuated lesions seen distributed in both lobes of liver [Figure-2] which are sub-capsular in position (largest 43x49mm). Similar lesions are seen in right lung field [Figure-3] corresponding to chest radiograph [Figure-4] lesion and anterior basal segment of right lower lobe adjacent to pericardium (largest 43x47mm). Few enlarged pre-tracheal and sub-carinal nodes are seen (Largest size of 8mm). CT scan showed Liver size-14cm and spleen size-9.3cm. Multiple enlarged lymph nodes were seen in mesenteric region pre and para-aortic region (largest 18x12mm). Generalized increased bone density was present. Microscopic examination of biopsy specimens revealed an increase in both the size and number of thin walled lymphatic channels along with lymphatic spaces that are interconnecting and dilated, lined by a single attenuated layer of endothelial cells.

DISCUSSION

Generalised lymphangiomatosis is a rare congenital condition caused by maldevelopment of the lymphatic systems during the

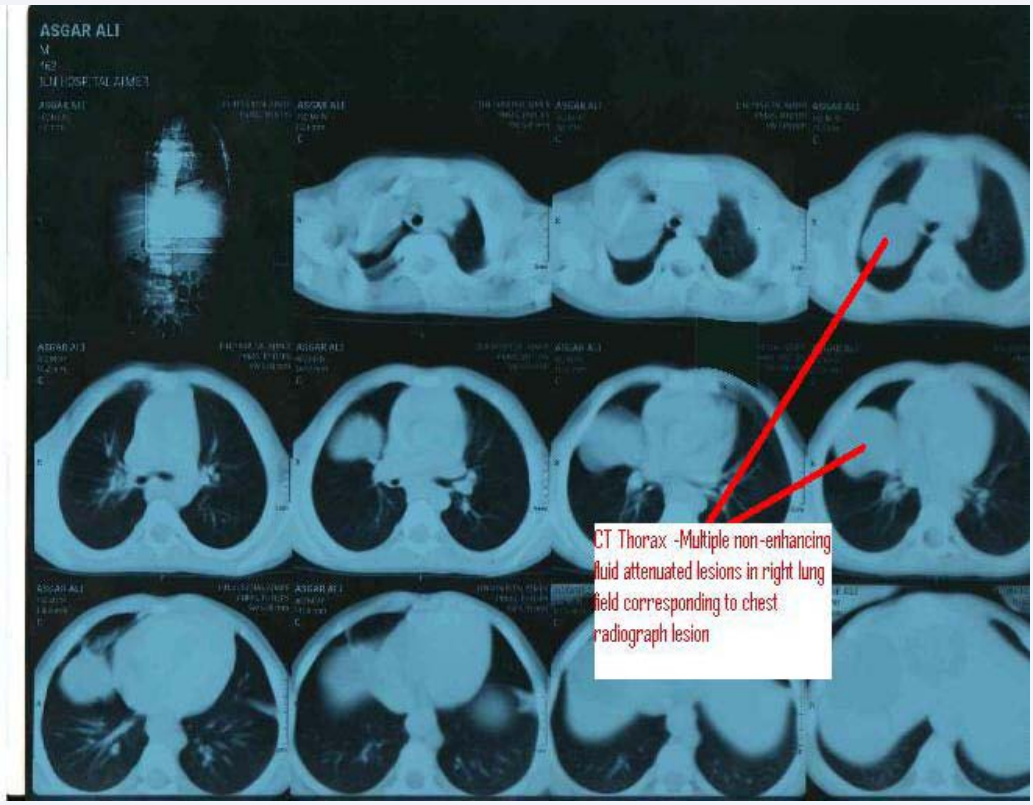

Figure 3 CT Thorax showing lung lesions.

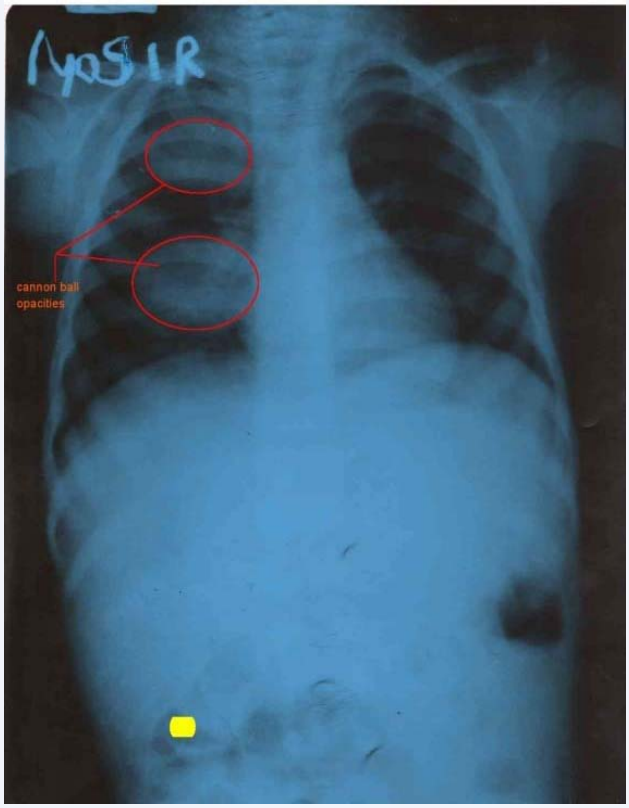

Figure 4 (A) – X-ray chest with cannon ball-appearance.

intrauterine period. It was first described by Rodenber in 1828; 65% of lesions are found in pediatric patients and it does not have any gender predilection.

Diagnosing the disease histologically can be difficult because its morphology overlaps with that of other disorders such as generalized fibromatosis and diffuse hemangiomatosis. Therefore, the use of cross-sectional imaging methods, such as CT scan and MRI scan is an important means of differentiating between these pathologies [4].

This disorder covers a spectrum of abnormalities and affects multiple organ systems. An isolated presentation usually carries a better prognosis than does multi-organ involvement. Hepatic involvement is exceptional presentation of diffuse lymphangiomatosis [7].

Similar to radiological finding mentioned by A Y Rostom, our patient's thoraco-abdominal CT scan also shows multiple non-enhancing fluid attenuated lesions (cystic) seen distributed in both lobes of liver which are sub-capsular in position (largest 43x49mm) [Figure 2]. Similar lesions are seen in right lung field [Figure 3] corresponding to chest radiograph lesion and anterior basal segment of right lower lobe adjacent to pericardium (largest 43x47mm). Chest Radiograph shows 2 cannon ball opacities in right mid and lower zones [Figure 4].

From above discussion it is apparent that simultaneous involvement of lung and liver in children is not reported and in x-ray typical cannon ball appearance and presentation of febrile seizures that is unrelated to disease is never reported before this case.

DETAILS OF CONTRIBUTION OF EACH AUTHOR

JP Narayan was involved in review of literature and in manuscript writing. B S Karnawat supervised the management and drafted the manuscript. Avadhesh Joshi was involved in taking details of patient, patient management and review of literature. JP Narayan shall act as a guarantor.

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