

# Clinical and radiological features of generalised lymphangiomas

CS Wong 黃鎮昇  
Tiffany YC Chu 朱曉青

We report a paediatric patient who presented with fever, shortness of breath, and vague abdominal discomfort. Lesions removed surgically proved to be generalised lymphangiomas and were treated conservatively. The spectrum of abnormalities and radiological features are discussed.

## Introduction

Generalised lymphangiomas is a rare congenital disorder mainly affecting children. It is believed that the disorder is due to maldevelopment of lymphatic systems during the intrauterine period. Pathologically, it consists of dilated chyle-filled spaces along the lymphatic systems. It can affect a wide range of organ systems including the pleural cavity, peritoneum, spleen, and the musculoskeletal system. Reports concerning the spectrum of imaging findings seen in this disorder are sparse and local reports of this disorder are even more rare. The aim of this report was to illustrate the clinical and radiological features of this rare congenital disorder.

## Case report

A 6-year-old girl presented to our institute 10 years ago with low-grade fever, shortness of breath, and vague abdominal discomfort. A physical examination showed cervical lymphadenopathy and signs of bilateral pleural effusions which were confirmed by a plain chest X-ray. Her spleen was also enlarged and her erythrocyte sedimentation rate and C reactive protein were raised.

A computed tomographic (CT) scan of her thorax and abdomen showed a bilateral pleural effusion, cystic masses inside the peritoneum, and numerous splenic cysts. Plain radiographs of the right humerus and both femurs showed multiple ill-defined lytic lesions (Fig 1). The provisional diagnosis was malignancy, such as lymphoma, with multiple metastases.

The management plan was discussed in the inter-departmental X-ray meeting and it was decided that tissue diagnosis was warranted. The patient then underwent a laparotomy. No solid tumour was found but multiple cystic masses were found inside the abdomen. Most were removed except those in the spleen and taenia coli. The tissue removed was sent for pathological analysis and the diagnosis of lymphangiomas was made. The pleural fluid was drained aseptically. A smear and culture for tuberculosis were negative but chyle was found in the fluid.

A diagnosis of generalised lymphangiomas was finally established. The patient was then treated conservatively, her symptoms subsided, and she has remained asymptomatic for more than 10 years. Recent follow-up CT scans (Fig 2) demonstrated a cystic mass in the peritoneum, bilateral loculated pleural effusions, and fluid collections in the mediastinal compartment. Her spleen was enlarged with multiple cystic lesions. Multiple soft-tissue

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Department of Diagnostic Radiology,  
Princess Margaret Hospital, Laichikok,  
Hong Kong  
CS Wong, MB, ChB, FRCR  
TYC Chu, MB, ChB, FRCR

Correspondence to: Dr CS Wong  
E-mail: singsingboy@gmail.com



FIG 1. Plain radiographs during the initial presentation showing multiple lytic bony lesions (arrows) and bilateral pleural effusions

nodules were noted in the subcutaneous region of the anterior abdominal wall. Lytic bone lesions, diffusely affecting the thoracolumbar vertebrae, bilateral iliac bones and femurs were found.

## Discussion

Generalised lymphangiomatosis is a rare congenital disorder caused by maldevelopment of the lymphatic systems during the intrauterine period. It was first described by Rodenber in 1828<sup>1</sup>; 65% of lesions are found in paediatric patients and it does not have any gender predilection.<sup>2</sup>

Diagnosing this disease histologically can be difficult because its morphology overlaps with that of other disorders such as generalised fibromatosis and diffuse haemangiomas. Therefore, the use of cross-sectional imaging methods, such as CT and magnetic resonance imaging, is an important means of differentiating between these pathologies. There are some isolated reports of an association with other mesenchymal dysplasias, such as Maffucci syndrome and diaphyseal aclasis.

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. The prognosis of the disease also depends on its extent. Skeletal involvement is well described in the literature and the pattern varies.<sup>3</sup> The most typical sites of involvement are the tibia, humerus, ilium, skull, mandible, vertebrae, and small bones of the hands. The bony lesions are usually osteolytic with multiple septae. They can lead to pathological fracture, joint deformity, and cause severe pain.

Another widely reported feature is extensive mediastinal, pleural, and peritoneal involvement with cystic fluid collections. Cytologic examination of these collections reveals lymph and chyle. When these features occur together with lytic bony lesions, the prognosis is generally believed to be worse.

Splenic involvement is another important feature. It has been reported that splenic lymphangiomatosis is often an incidental finding with characteristic imaging features. Computed tomographic scans show an enlarged spleen containing multiple well-circumscribed cystic lesions which usually show no contrast enhancement.<sup>4</sup> Although rare, subcutaneous nodules, as seen in our case, have occasionally been described in the literature.<sup>1</sup>

Treatment is usually conservative. Surgical intervention is indicated when complications, such as pathological fractures and symptomatic deformity, are encountered. Recently, bisphosphonate medication has been investigated as a means of preventing bone loss in skeletal lymphangiomatosis but the optimal

## 泛發性淋巴管瘤病的臨床及影像學分析

本文報告一名小兒患者，應診時有發燒、呼吸短促及腹部不適。病人進行手術病灶切除，證實有泛發性淋巴管瘤病，其後接受保守治療。本文並討論此病多種異常的徵狀及影像學特徵。

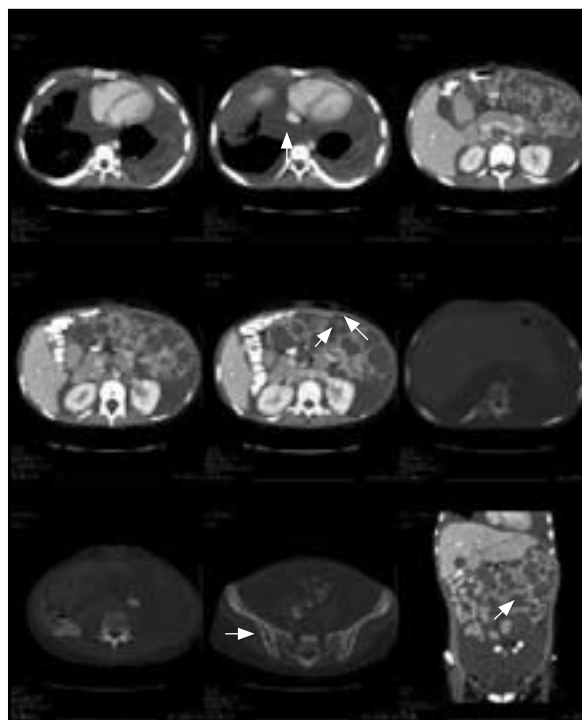


FIG 2. Selected follow-up computed tomographic images showing loculated fluid collections in the mediastinum (↑), pleural cavity, and peritoneum. The spleen was markedly enlarged with numerous cystic lesions inside (↗). Multiple well-defined lytic lesions (→) were noted in the bone window settings. Moreover, a few subcutaneous nodules (↖) were seen in the middle image of the second row

dosage for oral bisphosphonate has not been well established in paediatric patients.

One drawback of this report is that images taken during the follow-up period could not be retrieved, rendering direct serial comparison difficult. Nevertheless, based on the descriptions in the radiology reports and discharge notes, we believe that the disease has remained static clinically and radiologically.

## Conclusion

Generalised lymphangiomatosis is a rare congenital disorder caused by maldevelopment of lymphatic systems which manifest as a wide spectrum of abnormalities. Diagnosing it histologically can

sometimes be difficult due to its features overlapping with other generalised vascular disorders. Cross-sectional imaging substantially broadens visualisation of the spectrum of abnormalities and may thus contribute to management of the disease by preventing an initial misdiagnosis.

## References

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