
CASE REPORT

Lymphangiomas: a Wide Spectrum of Disease

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ABSTRACT

Lymphangiomas is a systemic disorder that can affect any organ system that contains lymphatics, including bones and soft tissues, resulting in a wide range of clinical and radiological manifestations. This report highlights lymphangiomas in a 29-year-old woman with both soft tissue and skeletal involvement. She gave a history of limb fractures but presented with left hypochondrial pain and imaging revealed multicystic lesions in the spleen. She had a splenectomy and its histology revealed lymphangiomas. Computed tomography scan revealed lucent lesions in her bones. As the presence of lucent bony lesions and visceral involvement can mimic a malignancy with bony metastases, lymphangiomas needs to be considered in the differential diagnosis, especially owing to the presence of lucent bone lesions with visceral involvement and chylous ascites.

Key Words: Angiomas; Bone neoplasms; Lymphangioma; Tomography, X-ray computed; Pleural effusion

中文摘要

淋巴管瘤病：多種不同表現的疾病

GC Gan、ML Wastie

淋巴管瘤病是一種系統性疾病，可發生在身體有淋巴管的任何器官，包括骨和軟組織，形成有多種不同臨床及影像表現。本文報告一名29歲淋巴管瘤病女性患者，病灶涉及其骨和軟組織。患者曾有骨折，病發時出現左季肋部疼痛，放射影像顯示其脾臟出現多囊性病變。患者接受脾切除，組織學檢查顯示淋巴管瘤病。電腦斷層攝影可見其病灶的骨透亮像。由於骨透亮病灶和涉及內臟的情況與有骨轉移的惡性腫瘤很相似，必須把淋巴管瘤病納入鑒別診斷中，尤其是當病灶出現骨透亮像，並涉及內臟和乳糜性腹水。

INTRODUCTION

Lymphangiomas is a benign malformation of the lymphatics characterised by abnormal proliferation of the lymphatic channels.¹ It is a congenital disorder with slow growth and multisystem involvement affecting the visceral and/or skeletal system, which results in a wide spectrum of clinical features. Histologically, there are three categories: capillary or simple, cavernous, and cystic.² When there is skeletal involvement, very often

internal organs (most commonly the spleen) are also involved.² This case report describes a young woman who had lymphangiomas involving her bones and spleen that simulated a malignancy with bony metastases.

CASE REPORT

In December 2002, a 29-year-old woman presented with upper abdominal pain in the left hypochondrium

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that she had been experiencing intermittently for two months. There were no other symptoms. Apart from being mentally challenged, she had a history of limb bone fractures. Computed tomography (CT) of the abdomen revealed multicystic lesions in the spleen and also a cystic lesion thought to arise from the pancreas.

When she was referred to our centre, an ultrasound revealed a grossly enlarged spleen, 18.5 cm in length with multiple cystic lesions. Another large lobulated cystic lesion was noted just anterior to the pancreas and was extending to the splenic hilum and across the midline to the right. At that time, apart from a mildly elevated serum total bilirubin (57.6 IU/ml), her other blood parameters were normal. Since malignancy affecting the spleen and the cystic lesion anterior to the pancreas could not be confidently excluded, the patient underwent splenectomy. Intra-operatively, the spleen had multiloculated cysts and there was also a

cyst anterior to the pancreas which was also resected. She recovered uneventfully and was discharged on the fourth postoperative day with penicillin prophylaxis. On follow-up two weeks later, she remained well.

The patient was lost to follow-up for the next three years, during which time she was well and had no complaints. Repeated CT in our hospital three years later showed multiple cystic lesions in the splenic bed (Figure 1). We postulated that the cystic lesions were recurrent disease in the residual splenic tissue as a result of incomplete splenectomy, or splenunculus / accessory splenic tissue. Also, there were cystic changes in her lower thoracic and lumbar spine, pelvic bones and in both proximal femurs. These lesions were not expansile, some had sclerotic margins but there was no periosteal reaction or cortical destruction, and no soft tissue components were evident. These cystic bone changes gave rise to a 'honeycombing' CT appearance. Measurement of the Hounsfield units of the cystic lesions in the bones was not helpful as these

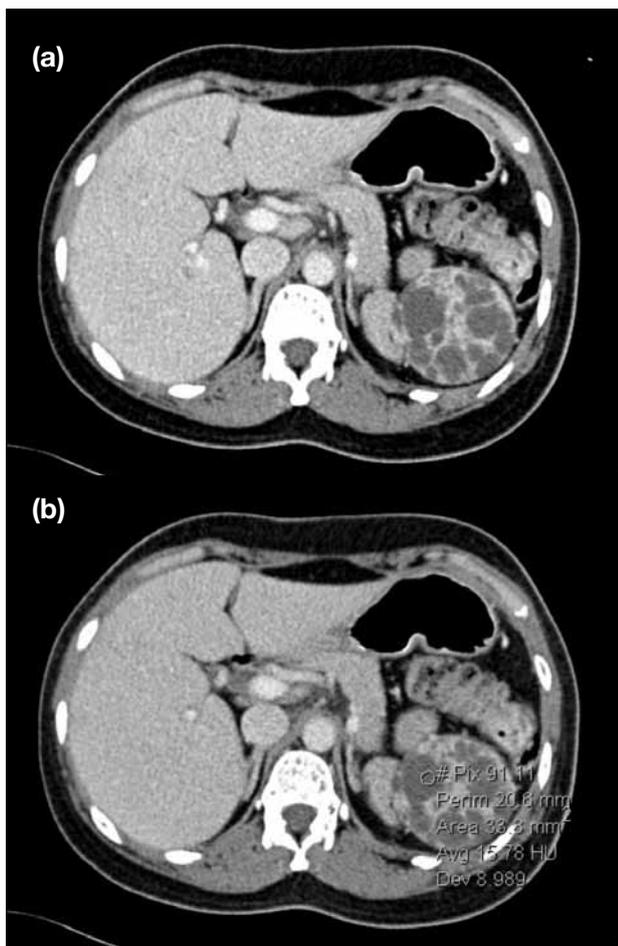


Figure 1. Axial computed tomographic scans of the abdomen three years after the splenectomy showing multiple cystic lesions in the residual splenic tissue in the splenic bed: (a) low-attenuation lesions in the residual splenic tissue, and (b) region of interest in the largest lesion showing cystic nature, HU +15.78.

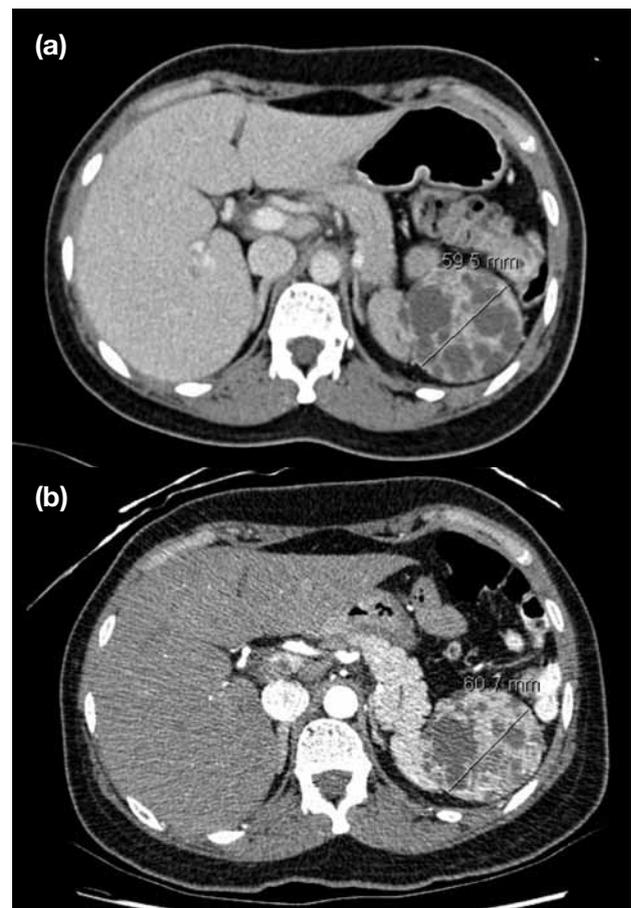


Figure 2. Axial computed tomographic scans of the abdomen on annual follow-up in (a) 2005 and (b) 2006 show no change in the multiple cystic lesions in the residual splenic tissue in the splenic bed.

had a wide range of values (-49 to +186). Subsequently, repeated annual CTs showed no changes in the size or appearance of the splenic or bony lesions (Figures 2 and 3). A radionuclide ^{99m}Tc -methylene diphosphonate (MDP) whole-body bone scan was normal, and did not show increased foci of uptake at sites corresponding to the cystic lesions noted on CT. Gross histological examination revealed an enlarged spleen with lobulated surface. There were multiple loculated cysts with scanty intervening splenic tissue. The contents of the cysts varied, being a straw-coloured fluid, haemorrhagic fluid and colloid-like material. Microscopically, the lymphoid tissue was replaced by cystic structures with dense fibrocollagenous walls that were inflamed or congested lined by low cuboidal or flattened epithelium, but there was no evidence of malignancy. These features were most consistent with lymphangiomatous cysts, and the cyst anterior to the pancreas was just a simple benign cyst.

DISCUSSION

Lymphangiomatosis is a rare clinical entity due to a congenital malformation of the lymphatic system, and has a wide range of clinical features and presentations. As it is sometimes difficult to differentiate this clinical entity from angiomas, it has been described by many terms, including cystic lymphangiectasis, lymphangiomatosis, diffuse skeletal angiomas, skeletal haemangiomas, multiple lymphangiectasis, hamartomatous haemolymphangiomas and haemolymphangiomas.² The spectrum of these diseases also includes Gorham's disease (vanishing bone disease) and Gorham-Stout disease (bony involvement with cystic changes). Lymphangiomatosis is a slow-growing disease and tends to present in children or young adults.

There are three types of lymphangiomatosis: capillary, cystic, and cavernous. The cystic is the most common type and comprises cysts of varying sizes (ranging from several millimetres to centimetres). Cystic hygroma is

an example. All three types may coexist in the same patient. On CT, cystic hygromas can appear uni- or multi-locular with fluid attenuation. The walls and septa are typically uniform in thickness and may enhance. Capillary lymphangiomas are rare. As they are usually found in the subcutaneous tissue and are small in size, they are rarely imaged.³ The cavernous type is most frequently reported in association with disease in the abdomen and thorax, with multiorgan and bone involvement.

With the exception of the central nervous system (CNS), there are lymphatics throughout the body, so any organ can be involved except for the CNS. The most common site of involvement is in the head and neck, where it manifests as cystic hygromas. In a review of eight patients who had diffuse lymphangiomatosis by Aviv et al,⁴ bony involvement and chylous pleural effusion were reported in seven out of eight patients, and the viscera and / or spleen were involved in half of the patients. In our patient, only splenic and bony involvement were detected and there were no cutaneous stigmata; nor did she have dyspnoea which would suggest a chylous pleural effusion. The most common sites of bony involvement are the pelvis, the vertebrae, the shoulder girdle, and the femora.² Typically, there are multiple sites of bony involvement that may or may not be contiguous.

The characteristic radiographic appearances of the bony lesions have been described as radiolucent areas with fairly narrow zones of transition. A faint, thin rim of sclerosis may be present. The bony lesions often start in the marrow but may be confined to the cortex. Osteopenia has also been described. There is neither a periosteal reaction nor adjacent soft tissue swelling. However, with soft tissue involvement, invariably there is adjacent bony involvement.^{4,5} On CT, Cohen et al⁵ reported the appearances of multiple sharply defined low-attenuation lesions with no cortical destruction, periosteal reactions, or soft tissue abnormalities. As

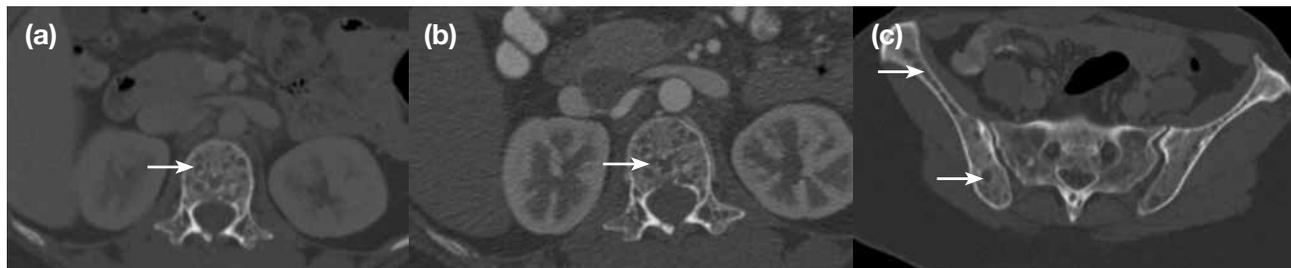


Figure 3. Axial computed tomography scans of the (a) thoracic spine at T9 and (b) lumbar spine at L5, and (c) right ilium in bone window on annual follow-up show relative unchanged appearance of the non-expansile cystic bony lesions with 'honeycomb' appearance (arrows).

in our patient, very wide range of Hounsfield units for the cystic lesions were obtained. Cohen et al⁵ also found that magnetic resonance imaging (MRI) could characterise the lesions, thus eliminating the need for biopsy. The lesions were described as having a very high signal on T2-weighted imaging and short T1 inversion recovery, consistent with the presence of fluid, with absence of any central enhancement or a faint rim of enhancement post gadolinium. The adjacent marrow and soft tissues did not show any abnormality.⁵

Scintigraphy has not been widely reported. Chu et al⁶ described irregular pooling of tracer in the soft tissue and bony lesions of lymphangiomatosis, which occurred rather quickly in the soft tissues, pleural and pericardial cavities, and lungs, but was delayed for up to 24 hours in the bones. In 99m Tc-MDP scans of the bones, they were able to detect areas of involvement as increased tracer activity before the appearance of changes on radiographs. However, others suggest that bone scintigraphy is inconsistent for the detection of bony lesions, as activity may be increased or decreased.⁴

The most common site of visceral involvement is the spleen, where it may be as part of a syndrome or the sole lesion. Splenic involvement can manifest as small focal subcapsular lesions, large cystic lesions (causing splenomegaly) and as lymphangiomatosis, in which the spleen is diffusely replaced by expanding lymphangiomas.⁷ This was the case in our patient, where there was only scanty remnant splenic tissue. These cystic splenic lesions can have Hounsfield units ranging from fluid/water attenuation (if the contents are serous), fat attenuation (if the contents are chylous), and even have higher attenuation due to haemorrhage discerned by CT.⁸ In this patient's CT scans, measured Hounsfield units of her splenic lesions ranged from +12.6 to +15.8, suggesting that the splenic cystic lesions had mostly serous contents. MRI has been used to characterise the contents of these cystic lesions.⁸

Lymphangiomatosis of the spleen can cause significant enlargement resulting in symptoms and increased morbidity due to complications (bleeding, rupture, hypersplenism, and consumptive coagulopathy).⁷ In our patient, despite a very large spleen, her blood counts and coagulation profile were normal. However, she presented with left hypochondrial pain probably due to a pressure effect or stretching of the splenic capsule. In lymphangiomatosis of the spleen, Barrier et al⁷ advocated total splenectomy with a search for any accessory spleens and their removal so as to avoid any recurrence. There is always a risk of recurrence from

any remnant splenic tissue. In our patient, as her spleen was completely removed according to the surgical / operative notes, we postulated that the mass in the splenic bed was most likely due to a splenunculus / accessory spleen that was not removed. Nevertheless, over a 6-year follow-up period, both her splenic and bony lesions remained stable.

Others have described various alternative treatment modalities for lymphangiomatosis with varying degrees of success. These include interferon alpha, on its own or together with steroids, radiation therapy as well as resection for intra-abdominal and skin lesions.^{4,8,9} The prognosis for this disorder also varies from death within a year of diagnosis to a fairly normal life span. It was noted that young patients, especially those with chylous pleural effusions, fared poorly.⁴ Our patient has been keeping well since her diagnosis 6 years ago.

CONCLUSION

With this report, it is our intent to highlight lymphangiomatosis that has a wide spectrum of manifestations, both clinically and radiologically. As the lucent bone lesions and visceral lesions of lymphangiomatosis can mimic malignant disease with bony metastases, the diagnosis of lymphangiomatosis must be considered especially in the presence of visceral involvement and chylous pleural effusions.

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