

CURRENT TOPIC

Treatment of thoracic lymphangiomas

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Lymphangiomas are rare benign neoplasms believed to be the result of abnormal development of the lymphatic system. They grow very slowly usually localised to one organ, but occasionally involve several organs in one part of the body. In the chest, involvement of the mediastinum, lung, heart, pleura, pericardium, ribs, and vertebrae may be seen in the same patient. Histologically they are made up of spaces lined with endothelium, varying from capillary size to several centimetres in diameter. They contain clear straw coloured fluid occasionally mixed with blood. The spaces are supported by a stroma of variable thickness containing lymphoid tissue. The aetiology is unknown but believed to be a result of congenital malformation of the lymph vessels. Radiation induced cutaneous lymphangioma has been described in a patient with breast cancer following mastectomy and radiotherapy.¹

Clinically, lymphangiomas are classified into three types:

- (1) Simplex, which is made up of capillary sized thin, walled lymphatic channels. This type usually affects the skin (lymphangioma circumscriptum)
- (2) Cystic lymphangioma (or cystic hygroma): this may range in size from a few millimetres to several centimetres, seen in a young age, commonly in the neck or the axilla
- (3) Cavernosum: this type is made up of dilated lymphatic channels, often with fibrous adventitial coats. This is the type which usually affects organs in the thorax, abdomen, and bones.

Bill and Sumner² pointed out that the three types may coexist in the same patient. Reports of single organ involvement have been published relating to nearly all organs of the body except for the CNS, which is devoid of lymphatic channels.

In one large series, lymphangioma of the head and neck accounted for nearly 50% of cases in children, while 10% of cases had visceral disease including the thorax.³ Thoracic lymphangioma has been described in all age groups including infants, neonates, and in the stillborn.⁴ Presenting symptoms are related to pulmonary and/or mediastinal mass, and signs and symptoms of pleural and/or pericardial effusion. Rib or thoracic vertebral bone involvement may present as chronic pain or acute pain in cases of pathological fracture. Bone

involvement in the young may result in shortening or deformity of the affected bone.

The plain chest radiograph will reveal either a localised lung lesion, with the solitary type, or diffuse pulmonary involvement with or without thoracic lymphadenopathy. Pleural or pericardial effusion is also common at presentation. Rib or vertebral involvement will be seen as rarefaction of bone, earlier in the course of the disease, or pathological fracture later on.

Parenchymal lung involvement with lymphangiomas has a characteristic computed tomography (CT) scan appearance. This can be seen as lobular or a cystic mass of heterogeneous soft tissue density in the localised type or diffuse involvement, seen as patchy areas of ground glass appearance.

Using conventional and high resolution CT scan (HRCTS), 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 the mediastinal fat and associated perihilar infiltration.⁵ Pleural thickening and/or effusion were seen in seven of the eight patients. Despite the typical appearance, they stressed the importance of biopsy for a definitive diagnosis. In one study comparing chest *x* ray with HRCTS of the chest in children with diffuse lung disease, Lynch *et al* confirmed the value of HRCTS scan over other conventional radiological methods.⁶

Any part of the mediastinum can be affected— anterior, middle, posterior, and superior, perhaps with slight prevalence in the anterior part. This will be seen as a well defined multilocular water density mass with or without compression or displacement of nearby structures.⁷ Shaffer *et al* pointed out that the magnetic resonance imaging (MRI) findings are not dissimilar from those of a CT scan and may include a cystic component isointense to muscles on T1 weighted images and hyperintense to fat on T2 weighted images.⁸ Comparing CT, ultrasound, and MRI findings, Pui *et al* concluded that MRI was more accurate in delineating extent of disease.⁹ This is important for preoperative surgical planning, and for these cases, coronal and sagittal views will be of great help to the surgeon

Treatment

Understanding the natural history of the disease aids the choice of treatment option.

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Accepted 27 April 2000

While spontaneous regression has been reported in two cases, and stabilisation of disease following surgery in five of 11 patients described in one series, this is very rare.⁷ The majority of cases will have slowly progressing disease; eventually this may lead to serious morbidity and even mortality in some cases.^{10–11}

For localised mediastinal or solitary lung lesions, thoracotomy and resection or thoracoscopic resection¹² is recommended. However, with the diffuse type, especially when there is multiorgan involvement, for example, mediastinum, lungs, heart, pleura, pericardium, ribs, and thoracic vertebrae, the role of surgery is limited apart from a biopsy for tissue diagnosis or draining of pericardial effusions and aspiration with pleurodesis for patients with recurrent pleural effusion. In one patient with recurrent chylothorax, ligation of the thoracic duct resulted in permanent cure.^{13–14}

Systemic chemotherapy¹⁵ and interferon alfa¹⁶ have been tried for patients with extensive inoperable lesions with limited success. Radiotherapy has been used for patients with extensive cutaneous involvement^{17–18} and for patients with abdominal involvement with excellent results.¹⁹ Similar results were reported for patients with extensive thoracic lymphangiomatosis treated by irradiation.^{20–21} Johnson *et al* treated one 13 year old patient with recurrent chylothorax with prompt and permanent resolution.²⁰ In a similar case, a 36 year old patient with chylothorax and chylopericardium and a mediastinal mass refractory to surgery and repeated drainage of pericardial and pleural effusions, was also treated by irradiation with complete regression of disease lasting for 12 months at the time of reporting.²¹ In the recent report by Kandil *et al*, three patients with extensive thoracic lymphangiomatosis were treated primarily by irradiation; this resulted in complete and permanent control of the disease with a follow up of 20 months, 32 months, and eight years in the three patients.²²

There is no agreement as to the best radiation doses to treat this disease. Aristizabal and Runyon used 40 Gy for disease in the pelvis and 26 Gy to the abdomen¹⁹; such dose if given to the lungs will result in radiation pneumonitis and lung fibrosis later on. It is generally agreed that radiation pneumonitis will seldom occur with a fractionated total dose of 20 Gy or less.²³ The dose used by Johnson and colleagues²⁰ and Dajee and Whitehouse²¹ was 20 Gy in 10 fractions. In the report by Kandil *et al* a dose of 18 Gy in 12 fractions over 18 days (uncorrected for lung transmission) was given with no acute or chronic lung toxicity, and was effective in controlling this disease.²² This dose has also been used in one of the three patients to treat cervical vertebra and skull base with similar good result. The radiation mechanism is not fully understood, but may be a result of radiation induced lymphatic endothelial oedema and proliferation leading to obstruction

of these channels, not unlike that seen in arterioles and microvasculature following radiotherapy.¹⁹

In summary, thoracic lymphangiomatosis is a benign congenital condition that may involve any or all organs within the chest. This can be seen in the neonate, infants, children, and adults. Although the radiological appearance may be typical, a tissue diagnosis should be obtained in all suspected cases. Observation of small asymptomatic lesions may be justified. However, with more extensive localised lesions excision is recommended. Recurrent pleural effusion should be treated by drainage and pleurodesis. For more extensive and complicated lesions involving more than one organ in the chest, interferon alfa or a fractionated dose of 18–20 Gy is an effective and safe treatment.

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