



Case Report

Copy Right@ Yassine El Ouai

Diffuse Pulmonary Lymphangiomatosis : A Case Report and Review of the Literature

Yassine El Ouai^{1*}, V Litique², K Oualla¹, Z benbrahim¹, L Amaadour¹, S Arifi¹, JM Limacher² and N Mellas¹

¹Medical Oncology department, Hassan II University Hospital Center, Morocco

²Onco-Hematology department, Louis Pasteur Hospital Center, France

***Corresponding author:** Yassine El Ouai, Medical Oncology department, Hassan II University Hospital Center, Fez, Morocco.

To Cite This Article: Yassine El Ouai, V Litique, K Oualla, Z benbrahim, L Amaadour, et al, Diffuse Pulmonary Lymphangiomatosis : A Case Report and Review of the Literature. Am J Biomed Sci & Res. 2021 - 12(5). AJBSR.MS.ID.001799. DOI: [10.34297/AJBSR.2021.12.001799](https://doi.org/10.34297/AJBSR.2021.12.001799).

Received: April 23, 2021; **Published:** May 06, 2021

Abstract

Cystic lymphangiomatosis is a benign malformative tumor of the lymphatic system most commonly affecting the crano-facial, cervical or thoracic region, and usually found in childhood. Its abdominal and especially retro-peritoneal location is rarer. The clinical presentation is very polymorphic. The diagnosis is suggested by imaging, but it requires histological confirmation. Its treatment is surgical and not always easy.

We report the case of a 44-year-old patient with a history of curative breast cancer, who was hospitalized for evaluation and management of a right pleural effusion and a pericardial effusion. A chest CT scan revealed diffuse pulmonary lymphangiomatosis.

The patient benefits from a puncture of these effusions, systemic corticosteroid therapy, then, following an early recurrence of these, colchicine treatment, with a favorable outcome at fourteen months from the diagnosis. The case of this patient illustrates that the course of diffuse pulmonary lymphangiomatosis in adults appears to be characterized by a less aggressive course and by a more favorable prognosis than that of children.

Introduction

Diffuse pulmonary lymphangiomatosis is a rare congenital disease characterized by overgrowth and dilation of intrathoracic lymphatic vessels. It appears between birth and adolescence in most cases but can manifest itself at any age with nonspecific respiratory symptoms. The prognosis is typically described as bleak, especially since the onset of the disease is early. We report the case of a 44-year-old woman diagnosed with pulmonary lymphangiomatosis, and review the literature to discuss the presentation, diagnosis and known treatments for this condition.

Case Report

We report the case of a 44-year-old patient who was followed at the Pasteur Hospital in Colmar for breast cancer, treated as curative by surgery, chemo and radiotherapy 8 years ago, then remained under good control.

The patient underwent to the ER for NYHA stage VI dyspnea. A chest CT showed right pleural and pericardial effusion. The diagnosis of diffuse pulmonary lymphangiomatosis was retained.

Discussion

The term lymphangiomatosis describes the presence of multiple lymphangiomas within one or more tissues. Any organ can be involved, but bone, spleen, liver, mediastinum, lung and neck are most commonly involved [1-3]. The central nervous system is never affected due to its lack of lymphatics. Although bone lymphangiomatosis is found in up to 75% of patients, the vast majority of cases involve the head and neck [1,4]. The smooth muscle surrounding the lymphatic vessels is intact, unlike that found in lymphangioleiomyomatosis, one of the differential diagnoses to be evoked. Depending on the tissue affected, clinical manifestations such as thoracic and pericardial effusions, often chylous, chylous ascites and lymphedema are noted.

Diffuse pulmonary lymphangiomatosis results from an abnormal embryogenesis of the lymphatic vessels contained in the chest cavity. Since the entire lymphatic system, from the mediastinum to the pleura, passing through the heart, lungs, pericardium, ribs and vertebrae may be involved [5,6], the pathology is also known



by the more apt term of Diffuse thoracic lymphangiomatosis. It has been described in both neonates and 80-year-old patients [1,7], but the initial presentation occurs during childhood or adolescence in 70% of cases [7]. Epidemiological data in the literature concerning the prevalence of diffuse pulmonary lymphangiomatosis are very limited. According to Alqahtani et al, lymphangiomas represent approximately 6% of benign pediatric tumors [4]. In their retrospective review of 186 patients with lymphangiomas, only nineteen (or 10%) had thoracic or visceral involvement.

Among the diagnostic methods useful in the evaluation of pulmonary lymphangiomatosis, X-ray imaging by CT or MRI is essential. Radiologic features suggestive of diffuse pulmonary lymphangiomatosis are diffuse thickening of the interlobular septa with sometimes a nodular appearance, thickening of interstitial tissue, pleural thickening and mediastinal infiltration [8,9]. Note that several of these elements are described on the patient's thoracic CT report. Although some authors consider MRI superior to CT in assessing the extent of disease, both radiological modalities are useful in diagnosis and their performance depends on local habits and the expertise of the radiologist. Lymphoscintigraphy is a minimally invasive examination that can also aid in the diagnosis, showing a build-up of contrast material in the lung or in the mediastinum [12]. The definitive diagnosis, however, is based on the histopathological analysis of the affected tissue ; distinguishing between lymphangiomatosis and lymphangiectasia can be difficult.

Treatment for diffuse pulmonary lymphangiomatosis is currently poorly defined and depends on the extent of the lesions present at the time of diagnosis. Localized forms may benefit from surgical resection, either by thoracoscopy or by thoracotomy. However, for diffuse forms, the usefulness of surgery is limited because it may be impossible to differentiate aberrant lymphatic vessels from healthy tissue, or because it is impossible to excise these same lymphatics without damage nearby structures. If the excision is incomplete, recurrence is certain. Pleurodesis or thoracic duct ligation are supportive treatments that may be effective for recurrent pleural effusions.

Drug treatment in pulmonary lymphangiomatosis would appear to be limited to a palliative goal. Trials of systemic corticosteroid therapy (modality used in the case described), vincristine, somatostatin, tamoxifen, interferon alpha, and pegylated interferon alpha-2b have been documented [5,11,12] with contradictory results. Some authors also describe the use of radiotherapy at doses varying between 18 and 20 Gy delivered over ten to twelve sessions with good results. The postulated mechanism of action in this case is cell proliferation and endothelial edema which results in the lumen of the capillaries being blocked and involved [5].

Despite tissue infiltration which is insidious and slow, and although histological analysis systematically reveals the presence of well-differentiated and therefore pathologically benign cells, diffuse pulmonary lymphangiomatosis is most often progressive over time, resulting in pleural and pericardial effusions. Recurrence or compression of mediastinal and / or parenchymal structures. The prognosis is variable depending on the age of presentation of the pathology. According to a review of the literature by Alvarez et al. of 48 patients with pulmonary lymphangiomatosis, twelve of the 31 children (newborns to sixteen years) included, or 39%, died of the disease. No deaths were noted among the seventeen remaining adults. These observations suggest that a late presentation of the disease in adulthood may be associated with a better prognosis for mortality. In this case, the first clinical manifestations, in the form of pleural effusion, were apparent in our patient at the age of 21 years. The lack of progress at the age of 39 would place the patient in this category. However, precise data in terms of morbidity and mortality in the literature remain limited. Spontaneous remissions have been described but are believed to be exceptional [5].

Conclusion

Diffuse pulmonary lymphangiomatosis is a rare pathology that remains poorly understood. It results from an abnormality in the embryogenesis of the lymphatic vessels contained in the chest cavity, resulting in the formation of lymph-filled cysts in their path. Unlike pulmonary lymphangiomyomatosis, which is less rare, and which is one of the main differential diagnoses to be made, there is no aberrant peribronchovascular proliferation of smooth muscle. Although diffuse pulmonary lymphangiomatosis occurs in pediatric patients in the majority of cases, it has also been described in adults of all ages.

Symptoms and clinical manifestations are nonspecific and include dyspnea, cough, diffuse chest pain, and effusions which are often chylous. Chest CT or MRI imaging is essential to guide investigations, although histological analysis of the affected tissue is necessary for a definite diagnosis. Treatment for the limited forms is surgical, while for the more extensive forms, systemic corticosteroid therapy is the most common treatment. The doses used and the duration of treatment are poorly defined and vary considerably between studies. This corticosteroid therapy can be combined with other molecules such as vincristine, somatostatin, tamoxifen, or alpha interferon; however, the results of these latter treatments are inconsistent. Finally, some authors also describe the use of radiotherapy in the treatment of the disease.

The prognosis for diffuse pulmonary lymphangiomatosis remains generally reserved, especially since the disease occurs

early. However, as in the present case, clinical stability, even without background therapy, can sometimes be observed when the disease first manifests in adulthood. More documentation and more research is needed to find a treatment that can affect the outcome of affected patients.

References

1. Oliver G (2004) Lymphatic vasculature development. *Nat Rev Immunol* 4(1): 35-45.
2. Bermejo EJ, Mongil Poce R, Arrabal Sánchez R, A Fernández de Rota Avecilla, A Benítez Doménech, et al. (2004) Linfangiomatosis torácica difusa : diagnóstico y tratamiento. *Arch Bronconeumol* 40(12): 599-601.
3. Ozeki M, Michinori Funato, Kaori Kanda, Masahumi Ito, Takahide Teramoto, et al. (2007) Clinical improvement of diffuse lymphangiomatosis with pegylated interferon alfa-2B therapy : Case report and review of the literature. *Pediatr Hematol Oncol* 24(7): 513-524.
4. Alqahtani A, Nguyen LT, Flageole H, Shaw K, Laberge JM (1999) 25 years' experience with lymphangiomatosis in children. *J Pediatr Surg* 34(7): 1164-1168.
5. Rostom AY (2000) Treatment of thoracic lymphangiomatosis. *Arch Dis Child* 83: 138-139.
6. Yekeler E, Dursun M, Yidlirim A, Tunaci M (2005) Diffuse pulmonary lymphangiomatosis : Image findings. *Diagn Interv Radiol* 11(1): 31-34.
7. Tamay Z, Saribeyoglu E, Ones U, Anak Sema, Güler Nermin, et al. (2005) Diffuse thoracic lymphangiomatosis with disseminated intravascular coagulation in a child. *J Pediatr Hematol Oncol* 27(12): 685-687.
8. Swensen SJ, Hartman TE, Mayo JR, Colby TV, Tazelaar HD, et al. (1995) Diffuse pulmonary lymphangiomatosis : CT findings. *J Comput Assist Tomogr* 19(3): 348-352.
9. Bellini C, Boccardo F, Campisi C, Bonioli E (2006) Congenital pulmonary lymphangiectasia. *Orphanet J Rare Dis* 1: 43.
10. Ozeki M, Funato M, Kanda K, Masahumi Ito, Takahide Teramoto, et al. (2007) Clinical improvement of diffuse lymphangiomatosis with pegylated interferon Alfa 2B therapy : Case report and review of the literature. *Pediatr Hematol Oncol* 24(7): 513-524.
11. De Lima AS, Martynychen MG, Florêncio RT, Lêda Maria Rabello, João Adriano de Barros, et al. (2007) Pulmonary lymphangiomatosis : A report of two cases. *J Bras Pneumol* 33(2): 229-233.
12. Kandil A, Rostom AY, Mourad WA, Y Khafaga, A R Gershuny, et al. (1997) Successful control of extensive thoracic lymphangiomatosis by irradiation. *Clin Oncol* 9(6): 407-411.