

# Solitary cystic mediastinal lymphangioma

*To the Editor:*

Solitary cystic mediastinal lymphangioma (CML) is a very uncommon benign vascular tumour developed from lymphatic vessels. Cystic lymphangioma is a cyst caused by a congenital malformation of the lymphatic vessels and can affect any site in the body, but <1% of lymphangiomas are mediastinal [1] and pulmonary lesions are even less common. They are most often located in the anterior mediastinum. We report a case of anterior CML in a 16-yr-old male, with a short review of the literature.

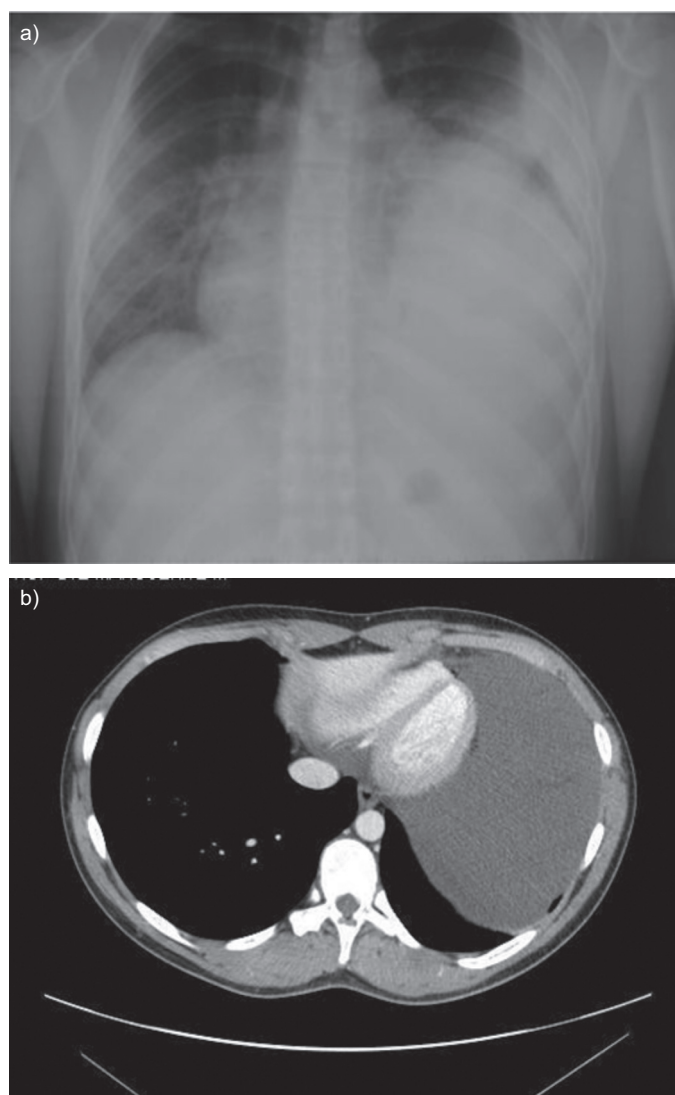
A 16-yr-old male was admitted to the Thoracic Oncology unit (Hôpital Nord, Marseilles, France) for recurrent left pleural effusion. The patient reported a 6-month history of chest pain and dyspnoea with hyperthermia. An initial diagnosis of left empyema was suspected. It was managed by iterative thoracentesis with saline pleural lavages, antibiotics and physiotherapy. On admission, physical examination was unremarkable except for left-sided dullness on percussion. The patient was afebrile. Chest radiography revealed a large basal opacity of the left lung silhouetting out the heart and diaphragm. Initially, this opacity was thought to represent a recurrent pleural effusion (fig. 1a). Computed tomography scans revealed a cystic lesion in the left basithoracic and anterolateral pleura with smooth borders measuring 63 × 90 mm (fig. 1b). No other intra- or extrapulmonary cystic lesions were found. With a presumptive diagnosis of a pleuropericardial cyst, video-assisted thoracic surgery was performed. Cytology and bacteriology were performed. The cyst contained serous fluid and had no connection or adhesion to adjacent organs, and was vascularised by small vessels from the mediastinal parietal pleura. Resection of the mass was performed. Histologically, the cyst wall was fibrous and lined by smooth muscle cells and by a single layer of flattened endothelial cells (fig. 2a). The margins of the resection were free from tumour. The lack of red blood cells in the cyst contents excluded haemangioma (fig. 2b and c). The final histological diagnosis was cystic lymphangioma. The post-operative clinical course was satisfactory, without complications and without recurrence, as documented by repeat computed tomography after 43 months of follow-up.

We hereby report the incidental finding of a solitary pleural-based pulmonary lymphangioma of the anterior mediastinum. CML is a rare benign vascular tumour that represents 0.7–4.5% of mediastinal tumours [1, 2]. More than 90% of CML is found in the first 2 yrs of life, 60% before the age of 5 yrs and only exceptionally in adulthood. To our knowledge, only few cases have been reported [1–4]. It has been described in cases of chromosomal and Mendelian abnormalities such as Turner syndrome, Down syndrome, and trisomy 13 and 18. CMLs are haemodynamically inactive, mature tumours with a low potential for carcinogenesis [1, 5].

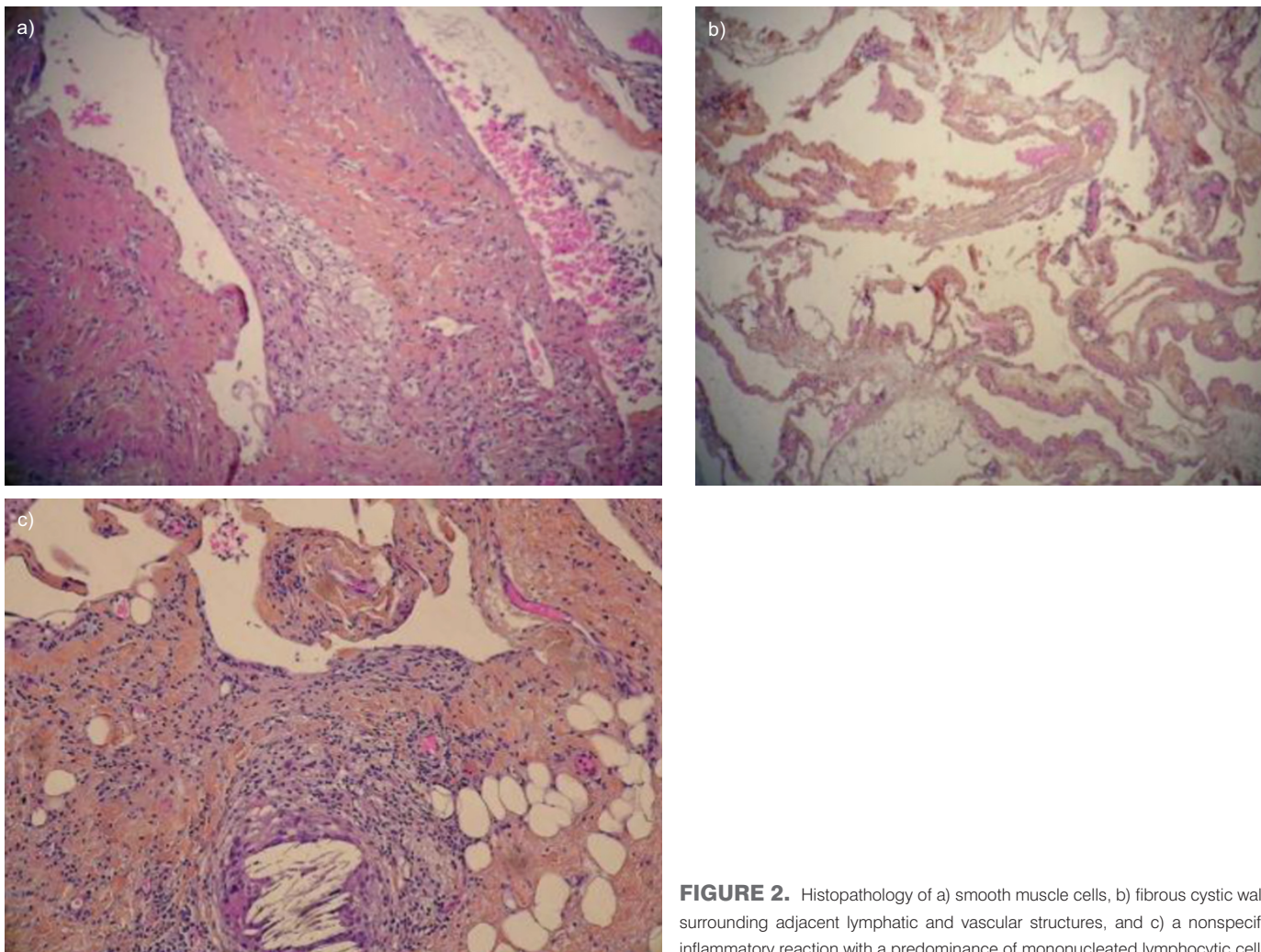
Cystic lymphangiomas were first described by Wernher in 1843 [6]. The first case of chest wall lymphangioma was reported in 1973 [6]. Cystic lymphangioma is a congenital malformation that

probably results from sequestration of lymphatic tissue that fails to communicate with the rest of the lymphatic system [1, 7]. The symptoms are based on the size and location, and vary widely, ranging from an acute mass syndrome to chest pain or local complication (rupture, infection, cystic haemorrhage or superior vena cava syndrome) [8]. Others have implicated specific factors such as infection, tumour or trauma [9].

According to the classification by Landing and Farber [10], lymphangiomas are classified into three types including: simple or capillary lymphangioma, cystic lymphangioma, and cavernous lymphangioma. Capillary lymphangiomas



**FIGURE 1.** a) Chest radiography showing a large basal opacity of the left lung silhouetting out the heart border and left hemidiaphragm, causing mediastinal shift. b) Chest computed tomography showing an anterolateral fluid collection located in the base of the left lung with smooth borders measuring 63 × 90 mm and causing regional atelectasis.



**FIGURE 2.** Histopathology of a) smooth muscle cells, b) fibrous cystic walls surrounding adjacent lymphatic and vascular structures, and c) a nonspecific inflammatory reaction with a predominance of mononucleated lymphocytic cells.

consist of dilatation of capillary-sized lymphatic vessels, which are connected to a normal lymphatic network. The cavernous variant contains dilated lymphatic sinuses in an actively growing lymphoid stroma, which are also connected to normal lymphatics. Cystic lymphangioma is characterised by multiple large cyst-like spaces lined by flat endothelial cells that may be empty or filled with clear proteinaceous or chylous fluid containing lymphocytes and, sometimes, red blood cells [3, 11].

A retrospective, multicentre study showed that in childhood, adolescence and young adults, lymphangiomas occupy cervical or mediastinal sites in phrenic chains and are often entangled with venous elements. This location along the pre-vascular phrenic lymph nodes makes surgical dissection difficult. Entanglement with vascular elements suggests a congenital origin [4].

In the past, lymphangiomas were distinguished from haemangiomas by the lack of intraluminal blood. However, core biopsies of lesion tissue using immunohistochemical and elastin staining are more useful in identifying lymphangiomas from other types of lesions [9].

The lymphatic nature of these lesions is sometimes difficult to demonstrate histopathologically, as it requires immunohistochemistry. A new monoclonal antibody, D2-40, which reacts

specifically with the lymphatic endothelium, can now be used. In this case, the endothelial-like lining cells of the cysts were positive for D2-40, establishing the lymphatic nature of these lesions. To our knowledge, there are few reports of cases of lung solitary lymphangioma with immunohistochemical confirmation using the monoclonal antibody D2-40 [11, 12].

The anterior mediastinal seats of our case tend toward confusion with other tumours, such as a specifically local congenital thymic cyst, a cyst or a bronchogenic teratoma. Similarly, a low anterior location would suggest a pleuropericardial cyst. The presence in the cyst walls of smooth muscle cells and lymphoid nodules can rectify the diagnosis.

The treatment of choice for cystic pulmonary lymphangioma is surgical resection [1, 11, 13]. The chances of recurrence are low if resection is complete and recurrence is common otherwise (35% *versus* 6% recurrence in case of complete resection) [4].

Complete resection may occasionally be difficult because of close proximity to vital structures [14, 15]. Other treatment modalities have been reported, such as marsupialisation, injection of sclerosing agents, steroids, diathermy and radiotherapy in unresectable cases, but they are generally ineffective and may lead to haemorrhage and infection [13].



In conclusion, primary pulmonary lymphatic disorders or neoplasms are rare and are often difficult to diagnose and classify [2]. Lymphatic abnormalities frequently mimic other pathologic processes, particularly neoplastic processes. These lesions are benign (as malignant change has not been documented) but tend to infiltrate surrounding tissues [8]. Provided that the tumour is completely resected, recurrences are rare. We believe this case adds a significant contribution to the literature due to its rarity and unusual location.

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**Statement of Interest:** P. Chanez has provided consultancy services for Almirall, Boehringer Ingelheim, Centocor, GlaxoSmithKline, Merck Sharp & Dohme, AstraZeneca, Novartis, Teva, Chiesi and Schering Plough; has served on advisory boards for Almirall, Boehringer Ingelheim, Centocor, GlaxoSmithKline, AstraZeneca, Novartis, Teva, Chiesi, Schering Plough and Merck Sharp & Dohme; has received lecture fees from Almirall, Boehringer Ingelheim, Centocor, GlaxoSmithKline, AstraZeneca, Novartis, Teva, Chiesi, Schering Plough and Merck Sharp & Dohme; and has received industry-sponsored grants from Almirall, Boehringer Ingelheim, Centocor, GlaxoSmithKline, AstraZeneca, Novartis, Teva, Chiesi and Schering Plough.

**Provenance:** Submitted article, peer reviewed.

**Acknowledgments:** The authors would like to acknowledge M-J. Payan (Hôpital Nord, Marseilles, France) for her expert interpretation of the histopathology specimens.

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DOI: 10.1183/09059180.00002212

# Pulmonary toxicity after long-term treatment with lenalidomide in two myeloma patients

*To the Editor:*

A white male was diagnosed with immunoglobulin G  $\kappa$ -chain multiple myeloma at age 63 yrs. He initially received a combination of thalidomide, doxorubicin and dexamethasone with concomitant radiotherapy (30 Gy) to the dorsal vertebrae. The first course of this combination was complicated by

profound pancytopenia and lobar pneumonia with severe sepsis in the setting of neutropenia, leading to the discontinuation of thalidomide because of concerns regarding haematological toxicity. The pneumonia was not microbiologically documented but considered as being probably of bacterial origin given the clinicoradiological presentation and a satisfying