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Case Report

Cystic Lymphangioma: Study of a Rare Case with Double Localization in the Mediastinum

Btissam Benabderrazik[1]*, Asmae Rais[1], Nizar El Bouardi[1], Amal Akammar[2], Meryem Haloua[2], Youssef My Alaoui Lamrani[1], Meryem Boubbou[2], Mounia Serraj[3], Bouchra Amara[3], Ouadnouni Yassine[4], Smahi Mohamed[4], Mustapha Maaroufi[1] and Badreddine Alami[1]

[1]*Central Radiology Department, CHU Hassan II, Fes, Morocco*

[2]*Mother and Child Radiology Department, CHU Hassan II, Fes, Morocco*

[3]*Pneumology Department, CHU Hassan II, Fes, Morocco*

[4]*Thoracic Surgery Department, CHU Hassan II, University of Sidi Mohammed Benabdallah, Fès, Morocco*

***Corresponding Author:** Btissam Benabderrazik, Central Radiology Department, CHU Hassan II, Fes, Morocco.

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Abstract

Mediastinal cystic lymphangiomas are extremely uncommon vascular tumors that develop from the lymphatic vessels. Most patients are asymptomatic. Symptoms may include cervical swelling, dysphagia, dyspnea, chest pain, or cough. Here, we describe a case of mediastinal cystic lymphangiomas in a 19-year-old female patient, complaining of a soft, fluctuating right basi-cervical mass. The diagnosis is based on clinical signs and imaging (CT and MRI), which showed two mediastinal multilocular cystic masses and a percutaneous fine-needle biopsy verified the diagnosis of cystic lymphangioma. Surgical excision is curative, but our patient refused to undergo surgery since there was no significant impact on her quality of life.

Keywords: *Mediastinum; Lymphangioma; MRI; CT*

Introduction

Cystic lymphangioma is a rare benign congenital malformation arising from confined enlargement of lymphatic vessels and represents 0.7 - 4.5% of all mediastinal tumors [1].

The cervical and axillary regions are the most commonly affected [1].

Patients under the age of five account for the majority of cases. There are extremely few documented cases in adulthood [2].

Although patients with cystic lymphangioma of the neck and mediastinum are usually non-symptomatic, they may experience symptoms such as coughing, dysphagia, or vascular compression syndromes due to the growing size of the lesion [3].

Imaging, especially MRI, shows a mass containing multilocular septa with cystic parts within the lesion, which is an important finding.

We report a rare case of a 19-year-old female with mediastinal cystic lymphangiomas.

Case Presentation

A 19-year-old female patient presented with a 1-year history of painless swelling on the right inferior side of the neck and chest heaviness. There was no particular medical history-no fever, dyspnea, cough, or dysphagia.

Physical examination: Vital signs were stable. Heart sounds were normal, and there was no rale or decrease in thoracic sound.

The swelling in the neck was in the right supraclavicular region. It was soft, fluctuant, and transilluminant. The mass increased in size with the valsalva maneuver. The lower part of this mass could not be reached.

Spirometric tests and laboratory workups (complete blood count and C-reactive protein) were normal.

The patient underwent CT of the neck and chest, which showed two masses: one in the right upper and posterior mediastinum extending into the basicervical region, and a second one in the posterior mediastinum, measuring 5 x 4 cm and 6 x 5 cm, respectively. Those lesions had smooth margins, spontaneous low density and were slightly enhanced after administration of contrast, containing central cystic components

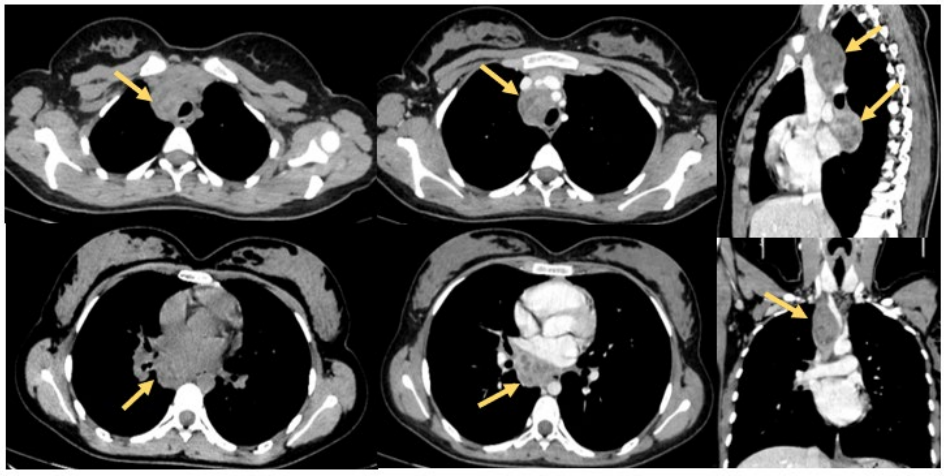

Figure 1: *Chest CT (axial, sagittal and coronal view) : Two lesions in the posterior medistinum (Arrow) showing spontaneous low density, slightly enhanced after administration of contrast with central cystic components.*

Magnetic resonance imaging revealed that the right upper mediastinal mass was extending to the basicervical region. The second lesion was in the posterior mediastinum without displacement of major mediastinal vessels or mass effect on the carina or stem bronchi.

The two masses have smooth margins with thick walls; they are isointense (to skeletal muscle) on T1-weighted images. T2-weighted images showed the multilocular cystic components with thick fibrous septa within the mass, which is a key feature in the diagnosis of cystic lymphangioma (Figure 2).

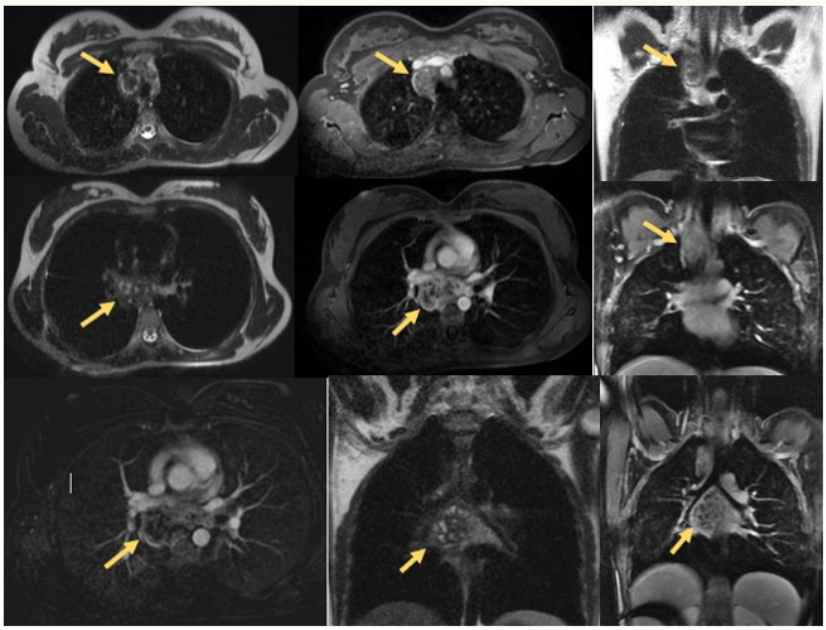

Figure 2: *Axial and coronal view T2W and T1 C+ FatSat MRI images: Two masses of the posterior mediastinum (Arrow) containing a multilocular cystic components with thick fibrous septa and late enhancement of the walls and septas after gadolinium administration.*

In our case, MRI gave us additional information about the morphology of the cystic-sepimented lesion, The patient underwent a fine-needle aspiration biopsy of the right cervico-mediastinal mass and the results of the cytologic exam and cyto-fluorimetry were concordant in terms of the diagnosis for cystic lymphangioma.

The patient refused to undergo surgery since there was no significant impact on her quality of life.

After 1 year of monitoring, the patient remained stable with no respiratory symptoms, and the right cervical fluctuant mass remained unchanged.

Discussion

Cystic mediastinal lymphangioma is a quite rare vascular tumor that arises from the lymphatic vessels [2].

It presumably results from the sequestration of lymphatic tissue and is categorized into three histological types: capillary, cystic, and cavernous lymphangiomas [4].

They are composed of cystic spaces, lined by endothelial cells and filled with clear proteinaceous or chylous fluid [5].

The majority of cases (75%) are located in the neck region and 3 - 10% of these lesions may extend into the superior mediastinum [9]. They can also occasionally occur in the axillary region (20%), the mediastinum, adrenal gland, kidney, bone, omentum, gastrointestinal track, retroperitoneum, spleen, liver and pancreas [6].

Only 1% of cystic lymphangiomas are localized in the mediastinum, usually in the right superior region, while the middle and posterior mediastinum localization is less common [3].

Clinically, they are seen in young children as the cavernous or cystic type that extend into the neck region or they manifest in adulthood as confined mediastinal cysts [7].

These tumors are usually found incidentally, but they may present cough, dyspnea, dysphagia, chest pain, or vascular compression syndromes [3].

CT shows a cystic lesion, homogenously low attenuating, with smooth edges that encase adjacent mediastinal structures [4].

On MR imaging, Cystic lymphangioma is isointense to muscle on T1-weighted images and hyperintense on T2-weighted images, with variability depending on the extent of cystic parts and proteinaceous content, with mild contrast enhancements of internal septa on the contrast-enhanced series [8].

MRI has the potential benefit of showing the cystic components and the multilocular septa within the mass, which is an important clue for the diagnosis [9].

In our case, the MRI provided more details about the nature of the cystic-sepimented lesions and the enhancement of the wall and septas of the masses after gadolinium administration. It also enabled us to analyze anatomic extensions with adjacent structures.

Other lesions that need to be taken into consideration in the differential diagnosis are hematomas, mediastinal abscess, cystic thymoma, teratoma and necrotic tumors [3].

The definitive diagnosis is histological after surgical resection; however, our patient declined surgery and underwent a fine-needle aspiration biopsy of the right cervicomediastinal mass along with a cytologic exam and cyto-fluorimetry. The results were consistent with the diagnosis of cystic lymphangioma.

Complications of lymphangiomas are rare but possible and include infection, cystic hemorrhage, superior vena cava syndrome, respiratory distress, chylothorax, and chylopericardium [10].

To our knowledge, there are no reports of spontaneous regression.

Surgical excision is usually curative but our patient refused to undergo surgery. Thoracotomy surgery offers the best exposure for a thorough dissection of all cyst, which is necessary to avoid recurrence, but it can be difficult depending on the volume of the mass and its extensions to mediastinal structures [11].

Sclerotherapy and radiation therapy are additional treatment modalities that have been documented in unresectable cases, but they are largely ineffective and can cause infection or haemorrhaging [2].

Sclerotherapy, using OK-432 agents, has been used in cervical cystic hygromas; however, it is not recommended for solitary mediastinal lesions [12].

The possible adverse effects of radiotherapy, including necrosis, radiation damage, delayed growth, malignant transformation to angiosarcoma, and other radiation-related tumors, have made it controversial at the moment [13].

Conclusion

Our case report illustrates a rare case of cystic lymphangioma with double localization in the posterior mediastinum.

Imaging, particularly MRI is helpful in the diagnosis of cystic lymphangioma, clearly demonstrating morphology of the mass and accurate anatomic extensions with adjacent structures before surgery, and allowing close monitoring when surgical treatment is refused or postponed.

Conflict of Interest

The authors have no conflicts of interest to declare.

Disclosure

All the co-authors have seen and agree with the contents of the manuscript and there is no financial interest to report. We certify that the submission is original work and is not under review at any other publication.

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