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RESPIRATORY CASE REPORTS

Cystic Lymphangioma of the Mediastinum

Mediastenin Kistik Lenfanjioması

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Abstract

Lymphangiomas are benign tumors originating from the lymphatic system, and it is very rare to be seen in adults and located in the mediastinum. In our study, we presented our case of mediastinal cystic lymphangioma, which we detected in a 60-year-old male patient.

Keywords: *Mediastinum, cyst, lymphangioma.*

Öz

Lenfanjiomlar lenfatik sistem kaynaklı benign tümörler olup yetişkin yaşta ve mediasten yerleşimli olarak görülmeleri oldukça nadirdir. Bizde çalışmamızda 60 yaşında erkek olguda saptadığımız mediastinal kistik lenfanjiom olgumuzu sunduk.

Anahtar Kelimeler: *Mediasten, kist, lenfanjiom.*

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Lymphangiomas are rare benign lesions originating from the lymphatic system that most commonly occur in children, and typically in the cervical region. The cystic form of lymphangioma, known also as cystic hygroma, is the most well-known type, while mediastinal lymphangiomas are much rarer, accounting for only 0.7–4.5% of all mediastinal tumors in the adult population (1-4).

We present here the case of an adult with mediastinal cystic lymphangioma, discussed in the context of relevant literature.

CASE

We describe here the case of a 61-year-old male who presented with hoarseness whose X-ray revealed no pathology, while a contrast-enhanced chest CT scan detected a lesion measuring 20x12 mm in the upper mediastinum adjacent to the aortic arch that raised suspicion for mediastinal lymphadenopathy, as well as a 7 mm nodule in the left lower lobe. A positron emission tomography (PET) scan showed no metabolic activity in the lesions, while an area of increased uptake with SUVmax: 5.3 was noted at the anterior aspect of the right 2nd rib, corresponding to a previous traumatic rib fracture (Figure 1).

The patient underwent a left anterior mediastinotomy for diagnosis and treatment during which the mediastinal lesion was excised. A histopathological examination showed positive immunohistochemical staining for D2-40, CD34 and CD31, while no reactivity was observed for CK, leading to a diagnosis of Cystic Lymphangioma (Figure 2).

Follow-up examinations in the first postoperative year reveal no recurrence or pathological findings (Figure 3).

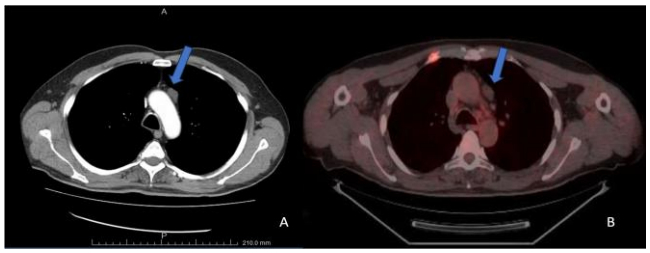

Figure 1: *Lesion identifiable adjacent to the aorta on thorax CT* ***(A)****, PET scan revealing the absence of metabolic activity in the lesion* ***(B)***

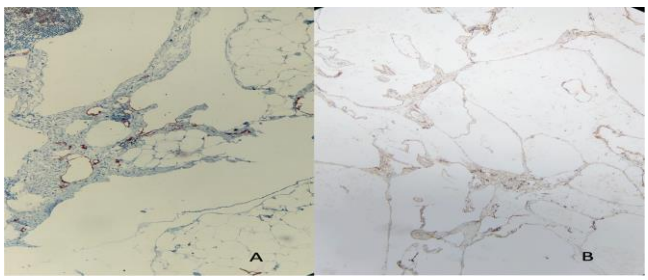

Figure 2: *Immunohistochemical examination showing lymphatic vessel structures stained with D2-40 (x5)* ***(A)****, Immunohistochemical examination showing lymphatic vessel structures stained with CD31 (x10)* ***(B)***

DISCUSSION

Lymphangiomas are rare benign lesions originating from the lymphatic system that fall under three categories: lymphangioma simplex, cavernous lymphangioma, and cystic lymphangioma (cystic hygroma). The most commonly observed cystic form is defined as a malformation resulting from a connection failure between the lymphatic and venous systems, the cause of which remains unclear. Most lymphangiomas are located in the neck and axilla, while less than 1% are found in the mediastinum (4,5).

Approximately 90% of lymphangiomas are diagnosed within the first two years of life, and around 50% are identified at birth. Such lesions are rare in adulthood and are typically asymptomatic, presenting as slow-growing masses. Mediastinal lymphangiomas are even rarer, and are usually asymptomatic and discovered incidentally (2,3,6). The presented case is a rare example of a mediastinal cystic lymphangioma.

Around 75% of cystic lymphangiomas develop in the neck, and approximately 25% in the axilla, and a very small proportion of these are located in the mediastinum, where they are often asymptomatic. Symptomatic lesions occur due to compression and can present with symptoms such as dysphagia, dyspnea, cough or chest pain (4,5). In Oshikiri et al.'s (2) study of five cases of mediastinal lymphangiomas, four of the cases were asymptomatic, while the other presented with superior vena cava syndrome resulting from compression. Similarly, the cases described by Saleiro et al. (3), Suehisa et al. (5) and Zhou et al. (7) all had asymptomatic mediastinal cystic lymphangiomas. In contrast, dyspnea and wheezing were observed in the case presented by Rali et al. (8). In our case, the patient presented with hoarseness, which is a relatively uncommon symptom in mediastinal lymphangiomas.

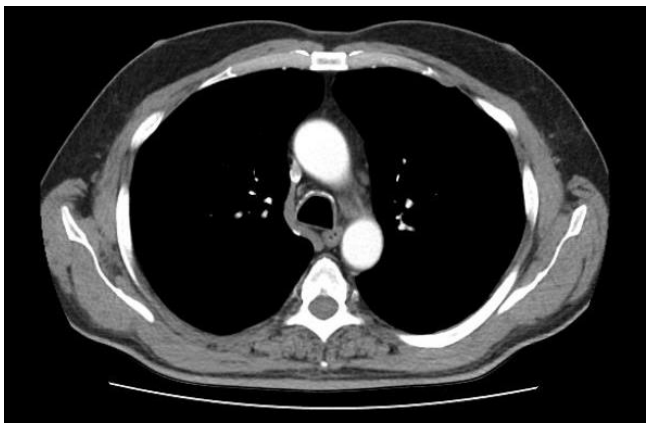

Figure 3: *Thorax CT appearance at 1-year postoperative follow-up*

Mediastinal cystic lymphangiomas are often discovered incidentally through radiological imaging. Chest X-rays

typically do not reveal specific lesions, although larger lesions may be seen as well-defined, round-shaped mediastinal enlargements. Both Thorax Computed Tomography (CT) and Magnetic Resonance Imaging (MRI) are effective in detecting mediastinal lymphangiomas. While chest CT is more commonly used and can help determine the size, density and location of a lesion, it is unreliable for definitive diagnosis. Furthermore, while chest MRI can reveal the heterogeneity of cystic components and the intense vascularization of lesions, it is also not conclusive for diagnosis. The optimum approach to definitive diagnosis is surgical excision followed by histopathological examination (3–5). In our case, the chest CT revealed a well-defined, homogeneous lesion in the mediastinum. Although unprecedented in the literature, we also performed a positron emission tomography (PET) scan, which revealed no metabolic activity in the lesion. As a definitive diagnosis could not be made radiologically, we proceeded with surgical resection for diagnosis and treatment.

Histopathological examinations of mediastinal cystic lymphangiomas typically show immunohistochemical staining for CD31, and D2-40 positivity in the lymphatic vessels within the lesion (5). In our case, the excised mass showed positive staining for both CD31 and D2-40, based on which the diagnosis was confirmed.

Surgical excision is considered the optimum treatment choice for lymphangiomas, and complete excision of the lesion is recommended to prevent recurrence. Recent studies have shown video-assisted thoracoscopic surgery (VATS) to be successful in mediastinal lymphangiomas (4,5,7). Although VATS has emerged as the first-choice surgical method in recent years, especially for mediastinal masses, we opted for an anterior mediastinotomy approach in our case as we felt more comfortable with the procedure.

In conclusion, although cystic lymphangiomas are frequently congenital and present in the neck region in childhood, the possibility of mediastinal lymphangiomas should also be considered in adults. Once diagnosed, lesions should be excised surgically to avoid complications and recurrence.

CONFLICTS OF INTEREST

None declared.

AUTHOR CONTRIBUTIONS

Concept - M.K., İ.A., A.Ç.U.; Planning and Design - M.K., İ.A., A.Ç.U.; Supervision - M.K., İ.A., A.Ç.U.; Funding - M.K., İ.A., A.Ç.U.; Materials - M.K., A.Ç.U.; Data Collection and/or Processing - İ.A.; Analysis and/or Interpretation - M.K.; Literature Review - M.K.; Writing - M.K., A.Ç.U.; Critical Review - M.K.

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