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CLINICAL CASES

Well-differentiated primary pleural liposarcoma

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Abstract

Primary pleural liposarcoma is a rare, slow-growing tumor, representing < 10% of pleural tumors, originating from residual remains of primitive mesenchymal tissue. We present the case of a 61-year-old male patient with chest pain and rapidly progressive dyspnea, observing in the imaging studies a hypodense tumor in the left hemithorax with Hounsfield units of –30 to –130 fatty tissue range with dimensions of 29.5 × 16.1 × 21.9 cm. Excision surgery together with chemotherapy and radiotherapy is the therapeutic alternative that has reported the greatest benefit in these patients.

Keywords: *Pleural liposarcoma. Mesenchymal tissue. Hypodense tumor.*

Introduction

Primary pleural liposarcoma (PPL) is an extremely rare malignant tumor, originating from mesenchymal cell remnants and very rarely from a pre-existing lipoma. A total of 43 cases have been reported in the literature worldwide, with the first case report being published by Ackerman and Wheeler in 1942. In the documented cases, it predominates in the male gender, being more frequent in the 5th decade of life (Table 1). The diagnosis should be suspected on images containing fat density with or without calcifications by chest computed tomography (CT) and confirmed by histopathological study. Surgical resection of the tumor is the therapeutic method of choice, although with risks of recurrence at 5 years[1].

Clinical case

A 61-year-old male presented with a history of systemic arterial hypertension of 25 years. Her condition began 2 months before hospitalization with asthenia, adynamia, non-productive cough, distal cyanosis, and rapidly progressive dyspnea. Physical examination included condensation syndrome in the left hemithorax. Laboratory studies Hb: 19, platelets: 118, leukocytes: 5.9, Dimer D: 4561. CT scan of the chest with image of hypodense tumor in the left hemithorax with Hounsfield units of –30 to 130 fatty tissue range and dimensions 29.5 × 16.1 × 21.9 cm, observing ovoid lesions with hyperdense walls with the presence of calcifications and hypodense centers near the posterior aspect of the hemithorax. Transthoracic biopsy was taken with an Abrams needle, obtaining samples of pearly tissue, histological sections identify neoplastic lesion composed of adipose tissue, with a solid growth pattern. Immunolabeling for MDM2 was performed, which was negative. During his hospital stay, the patient presented respiratory and hemodynamic deterioration and was admitted to the intensive care unit, where he later died due to complications secondary to the underlying diagnosis.

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In the autopsy study, significant congestion was observed in the cephalic and cervical segments, jugular engorgement, and subclavian veins, with data of a superior vena cava syndrome. At the opening of the thoracic cavity, a tumor measuring 30 × 20 × 15 cm, well defined, with a lobed surface and adipose appearance, was observed in an anterior situation and occupying the entire left hemithorax (Fig. 1A). With collapse of the ipsilateral lung, displacement of the heart and mediastinal structures toward the right hemithorax (Fig. 1B). The appearance of the tumor was homogeneous with an adipose appearance, light yellow in color, with foci of necrosis and calcifications. Histologically, the tumor was composed of neoplastic adipocytes of mature appearance, with variation in their shape and size, accompanied by a fine vascular network with a characteristic chicken wire appearance (Fig. 1C). Nuclear atypia was demonstrated in both neoplastic adipocytes and stromal cells (Fig. 1D). Foci of sclerosis were observed, with dystrophic calcification, no evidence of dedifferentiation or heterologous elements was found. With the above, the diagnosis of well-differentiated liposarcoma, adipocytic subtype, was concluded. Immunohistochemical reactions were performed as a diagnostic adjunct, demonstrating diffuse cytoplasmic and nuclear positivity for S100 (Fig. 1E), and focal nuclear positivity for p16 in neoplastic adipocytes (Fig. 1F), as well as positivity for CD34 in blood vessels. Immunoreactions were performed for MDM2 and CDK4, which were negative; although it is expected to find positive these last markers since these oncogenes are molecularly amplified, their negativity does not rule out the diagnosis. In addition to what has already been mentioned, two previously unidentified lesions were found in the thyroid gland. A classic variant papillary thyroid carcinoma located in the left thyroid lobe, which measured 1.2 × 1.0 × 1.0 cm, and a clear cell follicular adenoma, located in the upper portion of the isthmus, which measured 1.5 × 1.3 × 1.2 cm.

Discussion

PPL is a rare tumor, accounting for 10% of tumors at the pleural level, other pleural lesions include benign lipomas, hemangioendotheliomas, hemangiopericytomas, angiomas, localized fibrous tumor of the pleura, and non-neoplastic cysts. PPL is usually slow-growing with an expansive rather than an infiltrative behavior, with non-specific clinical manifestations, leading to a delay in its diagnosis[2-4].

The World Health Organization classified this tumor into well-differentiated liposarcomas (40-50%), myxoid (20-30%), dedifferentiated subtype (15-20%), and pleomorphic (5-10%), depending on the histological components of the tumor. It occurs in adults in the fourth or fifth decade of life without gender predominance. Myxoid liposarcoma shows a specific and reciprocal chromosomal translocation that occurs in more than 95% of t(12; 16), (q13:p1), fusing the CHOP and FUS genes, causing a dysregulation in adipocyte differentiation and allowing the proliferation of immature forms. The round cell variant is characteristic and known for its worse prognosis[5]. Liposarcoma is thought to be derived from residual remnants of primitive mesenchymal tissue that may undergo a conformational change to malignant tissue, which later develops as a pleural liposarcoma[6,7].

In imaging studies it is described as a well-defined extrapulmonary mass that may have the presence of calcifications that can cause displacement of thoracic structures[8]. Color Doppler ultrasound can be useful in depicting the solid nature of these tumors and assessing the internal vascularity of the tumor. MRI is the study of choice for defining soft tissue tumors, delineating their location and planning the surgical procedure[9]. Atypical lipomatous tumor/well-differentiated liposarcoma, myxoid/round cell liposarcoma are S-100 protein positive and variably MDM2 positive. Pleomorphic liposarcoma is variably positive for both S-100 protein and MDM2, and well-differentiated liposarcoma is generally only positive for vimentin[5,10].

There are studies that suggest that p16 has greater sensitivity and specificity for the detection of well-differentiated liposarcoma and dedifferentiated liposarcoma; however, its diagnostic yield is higher in combination with MDM2 and CDK4. Although molecular confirmation of this tumor is desirable by demonstrating the amplification of MDM2 and/or CDK4, the morphological study and the clinical context of the tumor make the diagnosis. The treatment of choice is wide and complete resection, provided that adjacent structures allow it. Surgery is complemented by adjuvant radiation therapy, which has been shown to improve local control and survival rate, and sometimes with chemotherapy with doxorubicin, dacarbazine, and ifosfamide. Tumor recurrence is predominantly in the affected hemithorax; however, it can occur in the contralateral hemithorax and is in 70-90% of cases, with a 5-year survival of 71%[9,11,12].

Table 1. Global amount of pleural liposarcoma found since 1942, with a total of 43 cases reported until 2023

Author (year)	Age/sex	Pleura site	Tumor size (cm)	Histology	Therapy	Follow-up (months)
Ackerman and Wheeler (1942)	50/F	Left	Nt	Nt	Autopsy	FE/12 M
Gupta and Paolini (19767)	51/M	Right	21	POR	Autopsy	DN
Ambrosio (1974)	52/M	Left	Nt	Nt	C-Res	VSEE/66
Wouters et al. (1983)	19/F	Left	3.5	MYX	C-Res + Rad	Alive/55
Evans (1985)	61/M	Left	Nt	MYX	Autopsy	FE
Evans et al. (1987)	45/M	Left	Nt	MYX	Autopsy	MOE/0.07
Gregor et al. (1987)	54/M	Right	2-2.25	LPBD	C-Res	MCD/108
Munk and Muller (1988)	27/F	Left	Nt	Nt	Nt	DN
Carroll et al. (1992)	23/F	Left	29-21	MIXED	C-Res + RT	VSEE/16
Wong et al. (1994)	38/M	Right	Nt	MY	C-Res + RT	VSEE/5
Batouk (1995)	55/F	Right	10 × 7 and 1.5 × 0.5	LPBD	C-Res	VSEE/16
Okby and Travis (2000)	45/F	DN	16	MYX/RO	P/I-Res + QT	FE/7
Okby and Travis (2000)	73/M	Right	Nt	MYX	P/I-Res	FE/9
Okby and Travis (2000)	67/M	Right	18.5	LPBD	Nt	MCD/16
Okby and Travis (2000)	80/M	Right	20	MYX	C-or/I-Res	DN
Tosson and Krismann (2000)	51/M	2 Right/ 1 recurrent: left	18 × 12 × 7	MYX	C-Res/RAD	VSEE/5Y
Minniti et al. (2005)	50/M	Left	13	WDL	C-Res + Rad	VSEE/12
Takanami and Imamura (2005)	59/M	Right	12-5.3	LPD	C-Res	VSEE/6
Goldsmith and Papaguiamnopoulos (2007)	42/M	Left	Nt	MYX	C-Res + Rad	Alive/12
Papaguiamnopoulos and Goldsmith (2007)	80/F	Left	Nt	MYX	P/I-Res	FE/8
Benchetritt et al. (2007)	76/F	Left	18-11	LPD	C- o P/I-Res	MOE/0.1
Peng et al. (2007)	56/F	Left	Nt	LPBD	C-Res	VSEE/18
Dagli (2008)	56/F	Left	Nt	MYX	ND	DN
Alloubi et al. (2008)	58/M	Left	Nt	MYX	C-Res + Rad	VSEE/10
Elsayed (2010)	47/M	Right	18 × 12 × 10	LPBD	C-Res	VSEE
Chen et al. (2014)	19/M	DN	Nt	LPBD	C-Res	Alive/56
Chen et al. (2014)	30/F	DN	Nt	LPBD	C-Res	VSEE/48
Chen et al. (2014)	60/M	DN	Nt	LPBD	C-Res	VSEE/43
Chen et al. (2014)	20/F	DN	Nt	MYX	C-Res	Alive/90
Chen et al. (2014)	54/M	DN	Nt	MYX	C-Res	VSEE/26
Chen et al. (2014)	41/M	DN	Nt	LPD	C-Res	Died/15
Chen et al. (2014)	53/M	DN	Nt	LPD	C-Res	Died/11
Chen et al. (2014)	61/M	DN	Nt	LPBD	P/I-Res	VSEE/18

(Continues)

Table 1. Global amount of pleural liposarcoma found since 1942, with a total of 43 cases reported until 2023 *(continued)*

Author (year)	Age/sex	Pleura site	Tumor size (cm)	Histology	Therapy	Follow-up (months)
Chen et al. (2014)	DN/F	DN	DN	DN	DN	DN
Carrillo (2014)	49/F	Left	19×10	PL	C-Res	DN
Albujar Ching (2016)	56/M	Right	2 kg	MYX	C-Res	DN
Wang et al. (2017)	43/F	Left	21	MYX	C-Res	VSEE/8
Matsukumu (2018)	45/M	Left	10	LPD	P/I-Res	FE/4.2
Prabhakar (2019)	32/M	Right	20.8 × 13.6	MYX	QT-RT	VSEE
Kang (2020)	66/M	Left	4.5 × 3.3 × 3	C-Res	C-Res	VSEE
Layek (2022)	22/M	Left	23 × 18	DN	DN	VSEE
Palacios (2022)	46/M	Right	20 × 25	MYX	I-Res + QT	Alive

F: female; M: male; VSEE: alive with no evidence of disease; EF: died of disease; EOM: died of another disease; MCD: died of unknown causes; QT: chemotherapy; C-Res: complete resection; RT: radiotherapy; Nt: not size; P/Res: palliative and incomplete resection; By: poorly differentiated; Myx: myxoid; LPBD: well-differentiated liposarcoma; MYX/RO: myxoid and rounded type; LPD: dedifferentiated liposarcoma; PL: pleomorphic; DN: data not available.

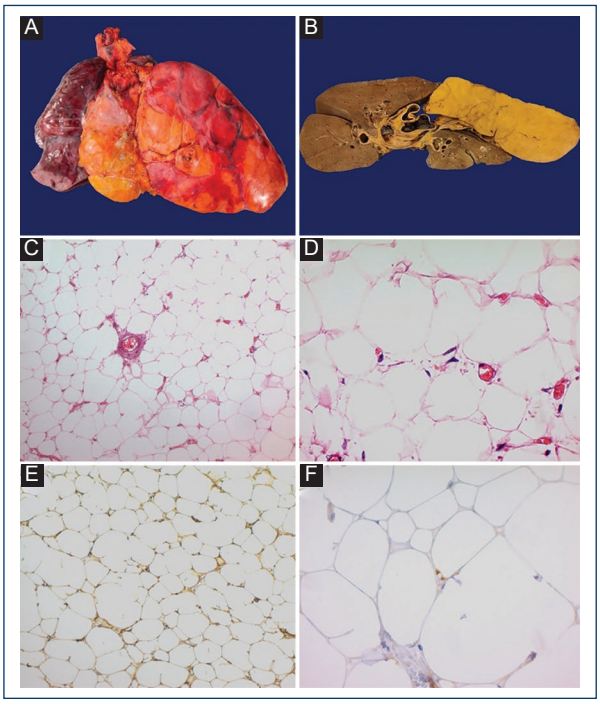

Figure 1. Liposarcoma. **A:** fresh cardiopulmonary block. **B:** tomographic section, piece fixed in formaldehyde. **C:** variation in cell size, vasculature in chicken wire, H&E, ×10. **D:** adipocytes with nuclear atypia, H&E, ×40. Immunoreactions: **E:** S100, positive in nucleus and cytoplasm of neoplastic adipocytes, ×10. **F:** p16 focal nuclear positive in neoplastic adipocytes, ×40.

Conclusion

Pleural liposarcoma is a rare tumor worldwide, the diagnosis is usually made late due to the presence of non-specific symptoms, being important to note that this entity should be suspected when a tumor with the presence of fat density in the thoracic cavity is found. However, there are no specific lesions, so the gold standard continues to be the histopathological study.

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Conflicts of interest

The authors declare no conflicts of interest.

Ethical considerations

Protection of humans and animals. The authors declare that no experiments involving humans or animals were conducted for this research.

Confidentiality, informed consent, and ethical approval. The authors have followed their institution's confidentiality protocols, obtained informed consent from patients, and received approval from the Ethics Committee. The SAGER guidelines were followed according to the nature of the study.

Declaration on the use of artificial intelligence. The authors declare that no generative artificial intelligence was used in the writing of this manuscript.

References

1. Prabhakar N, Vaiphei K, Vishwajeet V, Ramamoorthy E, Gorsi U, Dhooria S, et al. Primary pleural liposarcoma: a rare entity. Lung India. 2019;36:438-40.
2. Navarrete C, Arias MA, Peláez M. Primary pleural liposarcoma, pleomorphic variant. J Thorac Dis. 2014;6:E166.
3. Tosson R, Krismann M. Primary myxoid sarcoma of the pleura: 5 years follow-up. Thorac Cardiovasc Surg. 2000;48:238-40.
4. Goldsmith P, Papagiannopoulos K. Pleural myxoid liposarcoma: features of 2 cases and associated literature review. J Cardiothorac Surg. 2007;2:48.
5. Palacios RD, Castillo MD, Ruiz PJ, Rafael Silva F. Primary pleural myxoid liposarcoma: case report and literature review. Int J Res Med Sci. 2022;10:2024-7.
6. Layek A, Rajpoot A, Joshi P, Mishra M. Primary pleural liposarcoma: a rare differential for an opaque hemithorax. BMJ Case Rep. 2022;15: e246683.
7. Okby NT, Travis WD. Liposarcoma of the pleural cavity: clinical and pathologic features of 4 cases with a review of the literature. Arch Pathol Lab Med. 2000;124:699-703.
8. Minniti A, Montaundon M, Jougon J, Hourneau M, Begueret H, Laurent F, et al. Liposarcoma of the pleural cavity. An exceptional tumour. Monaldi Arch Chest Dis. 2005;63:170-2.
9. Albujar-Ching YT, Salazar-Loconi W, Hoyos-Arrascue J, Paredes-Ramírez V. Liposarcoma mixoide en pulmón. Rev Cuerpo Med Hosp Nac Almanzor Aguinaga Asenjo. 2016;9:257-60.
10. Chen H, Shen J, Choy E, Hornicek FJ, Shan A, Duan Z. Targeting DYRK1B suppresses the proliferation and migration of liposarcoma cells. Oncotarget. 2018;9:13154.
11. Thway K, Flora R, Shah C, Olmos D, Fisher C. Diagnostic utility of p16, CDK4, and MDM2 as an immunohistochemical panel in distinguishing well-differentiated and dedifferentiated liposarcomas from other adipocytic tumors. Am J Surg Pathol. 2012;36:462-9
12. Clay MR, Martinez AP, Weiss SW, Edgar MA. MDM2 and CDK4 immunohistochemistry: should it be used in problematic differentiated lipomatous tumors?: a new perspective. Am J Surg Pathol. 2016;40:1647-52.