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Case Report

A Case of Mullerian Cyst Arising in Posterior Mediastinum

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A mediastinal Mullerian cyst was initially reported as a new category of congenital cyst by Hattori, et al. in 2005. We treated a 53-year-old female referred to us with a posterior mediastinal tumor found at the Th5 prevertebral level by chest-computed tomography during a medical check-up. She had a history of mediastinal teratoma, which was removed at the age of 35. Chest magnetic resonance imaging revealed homogenous, high-intensity signals in T2-weighted images. The lesion was resected using a thoracoscopic procedure, and histologic and immunohistochemical staining revealed a ciliated cyst of Mullerian origin. The newly established mediastinal Mullerian cyst should be included in the differential diagnosis of posterior mediastinal cysts.

Keywords: Mullerian cyst, mediastinal cyst, posterior mediastinal tumor, Hattori's cyst

Introduction

We treated a patient with a mediastinal Mullerian cyst, initially reported by Hattori et al. in 2005, which is a newly categorized congenital cyst that should be differentially diagnosed from bronchogenic and pericardial cysts.[1)]

Case Report

A 53-year-old female was referred to our hospital due to discovery of a posterior mediastinal tumor in chest-computed tomography (CT) findings obtained during a medical check-up. She was menopausal and had a history of a left anterior mediastinal teratoma, which was removed via a left thoracotomy at the age of 35 years. Physical findings and laboratory tests were normal. Chest CT demonstrated a cystic tumor 2 cm in diameter in front of the 5th vertebra (**Fig. 1a**). Chest magnetic resonance imaging (MRI) showed a tumor with homogenous high intensity signals equal to water density in T2-weighted images (**Fig. 1b**). The patient underwent tumor resection with a thoracoscopic procedure conducted in a two-port manner, in which each port was made at the right 5th intercostal space. The cyst wall was thin and lucent. After careful dissection, absence of adhesion or communication between the tumor and surrounding tissue was confirmed. The cystic content was composed of transparent fluid.

A histologic examination of the specimen revealed a thin-walled cyst lined by ciliated or flattened epithelium, with scant connective tissue and a bundle of smooth muscle fibers in the wall (**Fig. 2a**). The lesion resembled a para-ovarian cyst, rather than a pericardial, thymic, or bronchogenic counterpart. There was no evidence of endometriosis. Immunohistochemical staining revealed a positive reaction of the lining cells to estrogen and progesterone receptors (**Fig. 2b**). Calretinin was negative.

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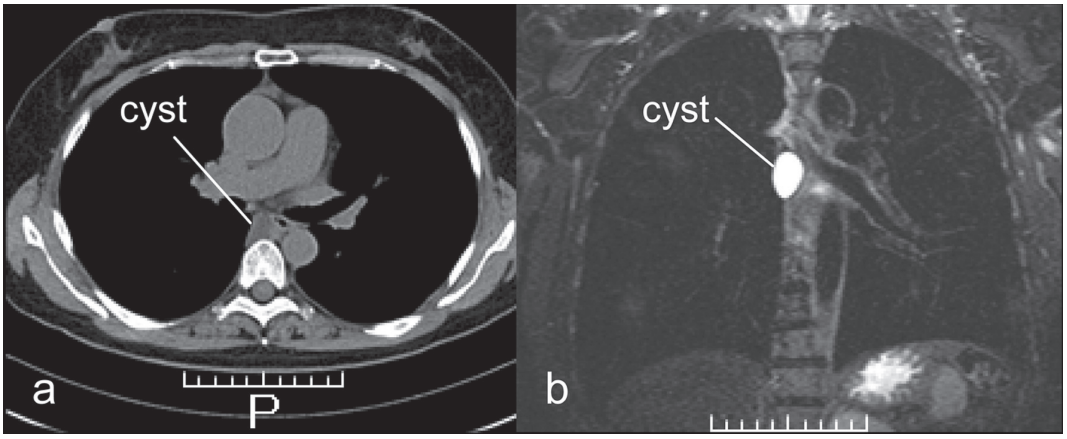

Fig. 1 **a**: Chest CT showing a prevertebral cystic tumor. **b**: Chest MRI T2-weighted image showing a homogenous tumor with a high intensity signal equal to water density.

The pathological diagnosis was mediastinal Mullerian cyst (Hattori's cyst). Her post-surgical course was uneventful, and the patient was discharged on postoperative day 6.

Discussion

Mullerian cysts are usually found around genitourinary organs or in the pelvis and rarely in the retro-peritoneum apart from the female genital tract.[2] A mediastinal Mullerian cyst, initially reported by Hottori, et al. in 2005, is a newly established entity, with only 14 known cases reported (**Table 1**).[1,3–6] However, the incidence of mediastinal Mullerian cyst may be higher, as the rate of incidence among all mediastinal cysts was reported to be 15.8% (3/19) by Hattori,[1] and 5.5% (9/163) by Thomas-de-Montpreville and colleague.[4]

Most mediastinal Mullerian cysts develop during the peri-menopausal period, and have also been reported to be associated with obesity and various gynecologic histories, such as hormone replacement therapy, hysterectomy, artificial abortion, and oophorectomy.[4] Thus, for retroperitoneal Mullerian cysts occurring in patients who have received treatment for menstrual irregularities or are overweight,[7] hormonal abnormalities may be suspected in relation to the development of a mediastinal Mullerian cyst. The present patient was obese and menopausal, though there was no notable gynecologic history.

The origin of a Mullerian cyst in the mediastinum is unclear, though Batt et al.[6] favored Ludwig's theory[8] for a thoracic location of developmentally misplaced endosalpingeal or endo- salpingeal epithelium. In meticulous embryologic studies, Ludwig reported that "in stage 16 embryos, a thickening of the coelomic epithelium develops on the cranial end of the plica mesonephrica at the level of the third to fifth thoracic vertebral blastema, and forms the anlage of the funnel area (of the fallopian tube)." Ludwig's studies of fetuses suggest not only a likely pathogenesis of a cystic structure lined by fallopian tubal epithelium, but also a likely explanation for persistence in the thorax at the T4-6 paravertebral level.

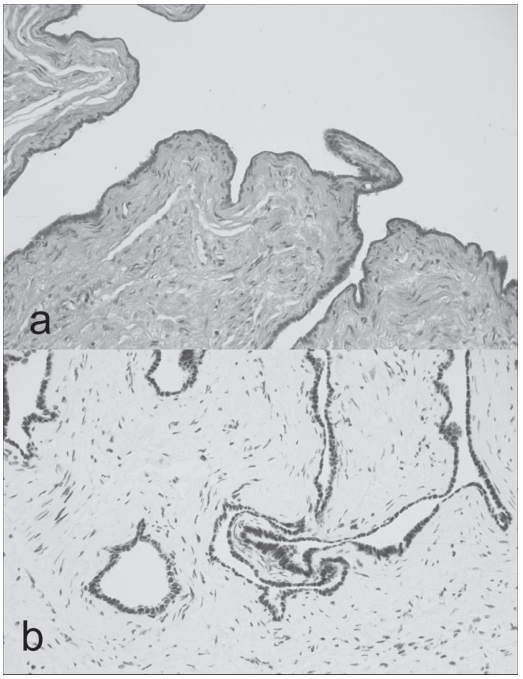

Fig. 2 A: Ciliated or flattened epithelium, and scant connective tissue with a bundle of smooth muscle fibers found in the cyst wall (H&E; × 100). B: Immunohistochemical staining showing a positive reaction of lining cells for estrogen receptor (diaminobenzene; × 200).

Table 1 Characteristics of Mullerian cyst in literatures

	Age/sex	Symptoms	Paravertebral level	Diameter	ER/PR
Hattori[3]	52/F	Persistent cough	Right Th6	2.5 cm	+/+
	18/F	Asymptomatic	Right Th5	2.0 cm	+/+
	49/F	Cough	Left Th4	2.0 cm	+/+
Thomas-de-Montpreville[4]	40/F	Chest pain	Left Th4	1.5 cm	+/+
	46/F	Cough	Left Th4	3.3 cm	+/+
	47/F	Cough	Right Th4/5	5.0 cm	+/+
	48/F	Asymptomatic	Left Th5	3.0 cm	+/+
	50/F	Chest pain	Right Th3/4	3.2 cm	+/+
	51/F	Asymptomatic	Left Th3/4	3.0 cm	+/+
	56/F	Asymptomatic	Left Th8	1.3 cm	–/+
	58/F	Cough	Prevertebral Th5	4.5 cm	–/–
	59/F	Chest pain	Right Th2-4	2.5 cm	–/–
Businger[5]	54/F	Asymptomatic	Left Th4-6	4.5 cm	+/+
Batt[6]	41/F	Chest pain	Left Th6	2.1 cm	+/+
Present case	53/F	Asymptomatic	Right Th5	2.0 cm	+/+

ER: estrogen receptor; PR: progesterone receptor

Mediastinal Mullerian cysts are found in the paravertebral area of the posterior mediastinum and often treated under the diagnosis of bronchogenic cyst or neurogenic tumor, with pathologic misdiagnosis of bronchogenic cyst due to ciliated epithelia.[4] Immunohistochemistry may be useful for the diagnosis in difficult cases, such as bronchogenic cysts without cartilage or mesothelial cysts with hyperplastic lining. Cytokeratin 5/6 is normally expressed in squamous epithelia, myoepithelial cells, basal cells of glandular epithelia, and mesothelial cells, whereas Mullerian cysts are negative or occasionally weakly positive for that marker.[4] Expressions of epithelial membrane antigen (EMA) and calretinin, and absence of an actin-positive subepithelial smooth muscle layer may help in the diagnosis of a pleuropericardial cyst. However, estrogen and progesterone receptors are certainly the best markers of Mullerian cysts.[2–4] In the present case, we considered that the lesion might be a bronchogenic cyst in our preoperative evaluation, though positive staining for estrogen and progesterone receptors in the lining epithelium led to our final diagnosis of mediastinal Mullerian cyst.

Conclusion

A mediastinal Mullerian cyst is a newly established entity that should be included in the differential diagnosis of posterior mediastinal cysts.

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