Müllerian cyst in posterior mediastinum: A report of a case

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A B S T R A C T

INTRODUCTION: A Müllerian cyst is a recently defined rare benign tumor of the posterior mediastinum. It is necessary to distinguish it from neurogenic tumor or bronchogenic cyst arising in the posterior mediastinum. Herein, we have reported and reviewed a case of Müllerian cyst in the light of the existing literature.

PRESENTATION OF A CASE: A 40-year-old woman was referred to our department for abnormal mediastinal tumor on computed tomography (CT). Chest CT revealed a 2-cm nodule in the left posterior mediastinum, while magnetic resonance imaging (MRI) T2-weighted scan revealed an elliptical, homogenous, and high-intensity neoplasm, and bronchogenic cyst or neurogenic tumor was suspected. She did not present with any symptoms. A thoroscopic surgery was performed for the cyst removal. Histopathological examination revealed that the cyst wall was covered with a layer of columnar epithelium. Immunohistochemical staining revealed the presence of estrogen receptor (ER) and progesterone receptor (PgR). Therefore a diagnosis of mediastinal Müllerian cyst was made.

DISCUSSION AND CONCLUSION: It is important to differentiate Müllerian cyst in the posterior mediastinum from other mediastinal cystic tumor for optimal decision-making in treatment.

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1. Introduction

Cystic tumors are quite common in the mediastinum, some of which manifest in the posterior mediastinum as neurogenic tumors, bronchogenic cysts, mesothelial cysts, and other types. Hattori [1] was the first to postulate that cystic tumors may be caused by Müllerian duct occurring in the posterior mediastinum in 2005. Since then, several studies have reported mediastinal Müllerian cysts. Herein, we have reported a patient with a Müllerian cyst in the posterior mediastinum and have reviewed her case in the light of the existing literature.

The presented work has been reported in line with the SCARE criteria [2].

2. Presentation of case

A 40-year-old woman visited our hospital for abnormal mediastinal tumor on computed tomography (CT). She had never smoked and did not present with any physical symptom or family history of malignancies. Her chest radiograph did not show any abnormal shadow; however, her CT scan revealed a mass of 10 × 12-mm diameter with a smooth margin at the left posterior mediastinum. Her chest magnetic resonance imaging (MRI) demonstrated a solitary lesion, and T2-weighted scans revealed a homogeneous hyperintense lesion, indicating a cystic tumor. Surgical resection was thoracoscopically performed, consistent with the diagnosis of a neuroenteric cyst. The cystic tumor was filled with a yellowish fluid, which was drained during the surgery. The cyst wall was incompletely resected because the tumor was in close proximity to the vertebral body. Pathological examination revealed that the cyst was covered with cuboidal or ciliated columnar epithelium, supported by an underlying fibrous stroma. Immunohistochemical staining demonstrated that the epithelial cells were positive for estrogen receptor (ER), progesterone receptor (PgR), and paired box gene 8 (PAX-8). The cyst was diagnosed pathologically as a mediastinal Müllerian cyst. The postoperative course was uneventful, and the patient was doing well at 12-months follow-up after surgery (Figs. 1–4).

3. Discussion

A preoperative diagnosis for mediastinum cyst is especially difficult for the existence of an aorta, heart and vertebra. Furthermore, the development of the tumor is unclear because of unknown etiology. In invasion or expansion to the surrounded organ, it might cause catastrophic events. In this report, we described very rare case of a Müllerian cyst in posterior mediastinum.

Müllerian cysts commonly develop in the male pelvis, where the Müllerian duct is left behind, although female Müllerian cysts in the retroperitoneum and mediastinum have also been reported [3]. After the first report by Hattori in 2005 [1], until date, 25 cases

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of Müllerian cysts have been reported, including the present case [1,4–15]. The 25 reported cases of Müllerian cysts were reviewed for the site, size, and symptoms of the tumor.

Occurrence in the left thoracic cavity was noted in 14 cases (56%), which is slightly higher than that in the thoracic cavity (Table 1). In addition, except for the report of multiple cysts in one case, majority of the reported tumors occurred around the Th4 vertebra. The median size of Müllerian cyst was approximately 30 mm. The cysts in 11 cases (44%) were asymptomatic, while coughing was reported in 6 cases (24%). On immunohistochemical staining, ER was found to be positive in 22 patients (88%) and PgR was positive in 21 cases (84%). In all reports, the cysts were filled with serous liquid. No recurrence after the removal of the tumor was reported.

The etiology of Müllerian cysts in the mediastinum remains unidentified. Batt et al suggested that the cyst is the choristoma from the primary Müllerian apparatus, similar to the one in the postulated pathogenesis for the Mayer-Rokitansky-Kuster-Hauser syndrome reported by Ludwig [6]. In stage of 17 embryos, the anlage of the Müllerian tube proliferates parallel to the Wolff tube on the caudal side. The lesion with the epithelium of the Müllerian duct left behind Th3-5 was found to be the source of the Müllerian duct and is believed to have been increased due to the abnormal hormonal stimulation, resulting in the formation of a cystic lesion.

However, the etiology of Müllerian cyst located in the mediastinum should ideally be different from those in the retroperitoneum, considering that the latter often has endocervical differentiation, which is typically not seen in the mediastinal cysts [9].

Hattori [1] was the first to suggest that Müllerian cyst in the mediastinum is a new disease entity, which was initially misdi-
agnosed as bronchogenic cysts because of its ciliated epithelium. Thomas-de-Montepelli reported that, in 3 of the 9 cases of Müllerian cyst occurring in the mediastium, benign serous cysts were initially misdiagnosed as pleuropERICARDIAL cysts [4]. As compared with bronchogenic cysts, the epithelium lining of cystic structures with Müllerian differentiation do not have a cartilage structure and are composed of ER- and PR-positive cells.

4. Conclusion

The prognosis of this tumor is presumed to be good, with no reports of recurrence after surgical removal. However, a case of respiratory distress was reported due to multiple development and progression [13]. Most of the reported cases of a Müllerian cyst in the mediastinum required further diagnosis with resected tissue because preoperative diagnosis was difficult. A small cystic tumor can thus be differentiated from a bronchogenic cyst or a neurogenic tumor arising in the posterior mediastinum. For a definitive diagnosis, immunohistochemical examination of the resected specimen is warranted to avoid misdiagnosis.

Conflict of interest statement

All the authors have nothing to declare.

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Ethical approval

I certify that this kind of manuscript does not require ethical approval by the Ethical Committee of Federico II University.

Consent

Written informed consent for publication of his clinical details and clinical images was obtained from the patient. A copy of the consent form is available for review by the Editor of this journal on request.

Author's contribution

Atsushi Sekimura: design, conception of the article, drafting of the article; Katsuo Usuda and Nozomu Motono: revisions, interpretation of the data; Shun Iwai and Aika Funasaki acquisition of the data and other reports; Hidetaka Uramoto: critical revisions and final approval.

Registration of research studies

N/A.

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