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Case Report

CASE REPORT

A case of a Müllerian cyst arising in the posterior mediastinum

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Abstract

A mediastinal Müllerian cyst is composed of heterotopic cystic Müllerian tissue resembling structures of the fallopian tube. We herein report a case of a mediastinal Müllerian cyst discovered during a medical check-up of a 41-year-old woman. She had no symptoms but had been diagnosed with hyperprolactinemia at the age of 24 years, for which she received hormonal therapy for 3 years. Chest computed tomography demonstrated a 3-cm-diameter cystic tumor in front of the Th10 vertebra. Thoracic surgery was performed to remove the tumor. The tumor was a monolocular cyst covered with a thin capsule, and pathological examination showed a thin-walled cyst lined by ciliated or non-ciliated columnar cells resembling tubal epithelium. Immunohistochemical analysis showed positive expression for paired box gene 8 and estrogen and progesterone receptors. The pathological diagnosis was a Müllerian cyst. A Müllerian cyst should be always be considered in patients with a mediastinal cyst.

INTRODUCTION

Mediastinal cysts have a broad range of etiologies, such as bronchogenic, thymic, neuroenteric cysts and Müllerian cysts. A Müllerian cyst is a type of Müllerianosis in which developmentally misplaced Müllerian tissues [1]. Müllerian cysts are usually found around the genitourinary organs or pelvis [1]. The incidence of Müllerian cyst among all mediastinal cysts ranges from 5.5% (9/163) to 15.8% (3/19) [2–4]. Making a correct preoperative diagnosis of mediastinal Müllerian cysts is clinically difficult. Diagnosis can be made by histological finding, ciliated epithelium with Müllerian differentiation. Mediastinal Müllerian cysts follow a benign course with no reported recurrence so that surgical excision is the treatment of choice. Mediastinal Müllerian cysts are usually located at the Th3-5 paravertebral level [3–5]. We herein present the first known Müllerian cyst located at Th10, the lowest reported level, in the posterior mediastinum.

CASE REPORT

A 41-year-old woman was referred to our hospital because of an abnormal shadow on a chest roentgenogram during a medical check-up. She had no complaints. She was a non-smoker and not obese (42 kg, 158 cm tall). Although she had not

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undergone abdominal or thoracic surgery, she had a history of hyperprolactinemia at the age of 24 years, for which she received hormonal therapy with dopamine agonist for 3 years. Physical findings and laboratory tests were normal except for the prolactin concentration (42.3 ng/mL; reference range: 6.1–30.5 ng/mL). Chest computed tomography showed a 3-cm-diameter cystic tumor in front of the Th10 vertebra (Fig. 1A and B). Chest T2-weighted magnetic resonance images showed a tumor with homogenous high-intensity signals equal to water, and T1-weighted images showed a hypointense lesion (Fig. 2). The patient underwent two-port thoracoscopic tumor resection. The cyst wall was thin and lucent. There was no adhesion or direct communication between the tumor and the spinal cord, tracheobronchial system, esophagus or surrounding tissue.

Histologic examination of the specimen revealed a thin-walled cyst lined by ciliated or non-ciliated columnar epithelium with scant connective tissue and bundles of smooth muscle fibers in the wall. The lesion resembled a paraovarian/tubal cyst.

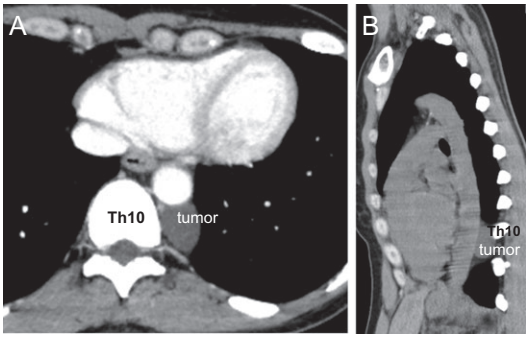

Figure 1: (**A** and **B**) Chest computed tomography. A paravertebral cystic tumor was located at Th10 level

There was no evidence of malignancy (Fig. 3A and B). Immunohistochemical staining revealed a positive reaction of the lining cells to paired box gene 8 (PAX8), estrogen receptor (ER), progesterone receptor (PgR) and CK7, and a negative reaction thyroid transcription factor 1 (TTF-1), CK20, calretinin and D2-40. Lining fibers under the cyst wall were positive for α-smooth muscle actin (αSMA) but negative for CD10 (Fig. 4). The pathological diagnosis was a mediastinal Müllerian cyst. The postsurgical course was uneventful, and the patient was discharged from the hospital on postoperative Day 4.

DISCUSSION

A mediastinal Müllerian cyst was initially reported by Hattori [2] in 2005. Since then, ~20 case reports have been published [3–5]. Most of mediastinal Müllerian cysts develop during the perimenopausal period and are reportedly associated with obesity and various gynecologic histories, such as hormonal therapy, hysterectomy, artificial abortion and oophorectomy [4]. Therefore, hormonal abnormalities may be suspected in relation to the development of a mediastinal Müllerian cyst. Our patient had a gynecologic history of hormonal therapy.

In this case, the tumor was located at the Th10 level. Mediastinal Müllerian cysts are usually located at the Th3-5 paravertebral level [3–5]. Although the origin of mediastinal Müllerian cysts is unclear, several theories have been proposed. Batt *et al.* [5] explained by Ludwig's theory. Ludwig [6] reported that 'in stage 16 embryos, a thickening of the coelomic epithelium develops on the cranial end of the plica mesonephridica 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 suggest not only a likely pathogenesis of a cystic structure lined by fallopian tubal epithelium but also a likely explanation for its presence in the thorax at the Th3-5 paravertebral level. The cyst in the present case was located at

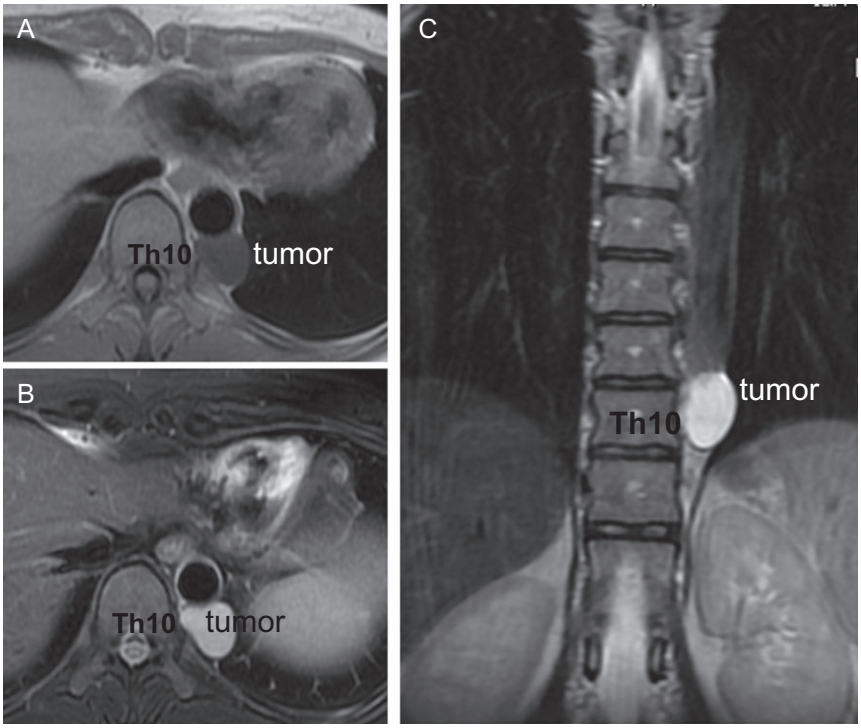

Figure 2: (**A–C**) Chest magnetic resonance imaging. A T2-weighted image showed a homogenous tumor with a high-intensity signal equal to water density

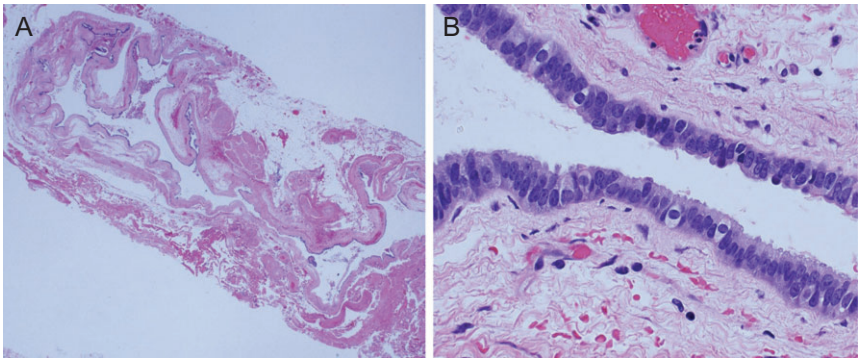

Figure 3: Histological findings. (**A** and **B**) Microscopic view showing that ciliated or non-ciliated columnar epithelium resembling fallopian tube, and scant connective with a bundle of smooth muscle fibers in the cyst wall. (Hematoxylin and eosin: **A**; ×4, **B**; ×200)

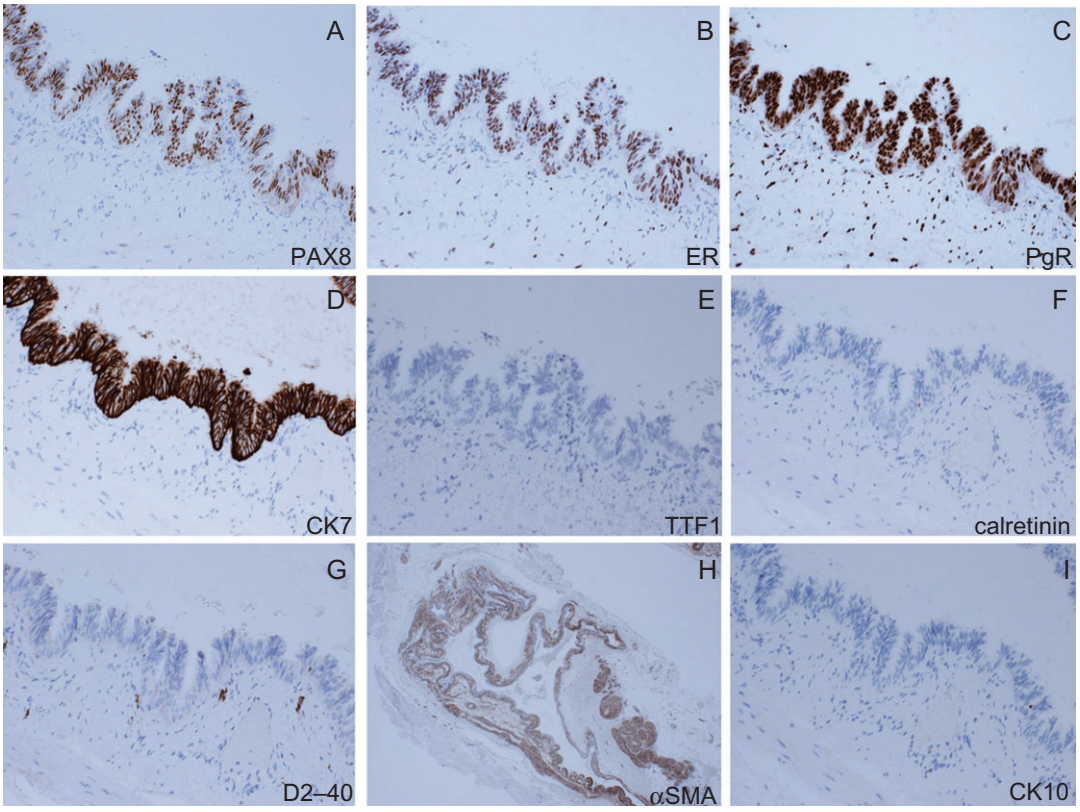

Figure 4: Immunohistochemical findings. Nuclei of epithelial cells stained positively for (**A**) paired box gene 8 (PAX8), (**B**) estrogen receptor (ER), (**C**) progesterone receptor (PgR), and (**D**) CK7 negatively for (**E**) thyroid transcription factor 1 (TTF-1), (**F**) calretinin and (**G**) D2-40. The lining fibers under the cyst wall stained positively for (**H**) α-smooth muscle actin (αSMA) and negatively for (**I**) CD10. The immunoprofile supported a diagnosis of Müllerian cyst. [**A**-**I**: immunohistochemical staining, ×40 (expect for **H**: ×4.)]

the Th10 paravertebral level, which is the lowest level ever reported.

Alternative hypotheses for the origin of mediastinal Müllerian cysts may include origination from ectopic ovarian tissue, coelomic metaplasia, endometriosis or surgical implantation [7, 8]. Hattori [2] suggested that the cyst could present misplaced mesothelium and mesenchyme with Müllerian characteristics. In fact, extrapelvic Müllerian cysts have been reported at unusual sites, such as the knee joint, as well as the posterior mediastinum [9, 10]. These presence of cysts at distant sites may be explained by vascular or lymphatic dissemination [9]. Lauchlan [8] described Müllerian tissue implantation due to metaplasia of ectopic peritoneal cells into secondary Müllerian epithelium. He theorized that the Müllerian tissue was distributed in a centrifugal manner from the ovaries and was not limited to the pelvis. We prefer this theory to Ludwig's theory because of the location of the cyst in the present case.

Mediastinal Müllerian cysts are found in the paravertebral area and are often treated based on the clinical diagnosis of a bronchogenic cyst or neurogenic tumor, leading to pathological misdiagnosis of bronchogenic cysts due to ciliated epithelia [4]. In this case, however, no cartilage or tracheal glands were present under the cyst. The lining epithelium closely resembled tubal epithelium or endometrial glands. Additionally, the

immunohistochemical study revealed positive staining for ER, PgR and PAX8, which are certainly the best markers of Müllerian cysts [3, 4]. The possibility of an endometriotic cyst was excluded because the lining stroma or fibers under the wall were positive for αSMA but completely negative for CD10. There were no clinical differences between this case and previous studies of mediastinal Müllerian cyst except for the tumor being located Th10 level. But this clinical significance might be key to finding solution to the origin of mediastinal Müllerian cyst.

In conclusion, although rare, the possibility of a Müllerian cyst should be considered in woman with a cystic lesion in the posterior mediastinum because of its distinct Müllerian histogenesis.

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None.

CONFLICT OF INTEREST STATEMENT

No conflicts of interest.

FUNDING STATEMENT

Not applicable.

ETHICAL APPROVAL

None required.

CONSENT

We obtained written consent from the patient for the publication of this case report and any accompanying images.

GUARANTOR

Motoaki Yasukawa.

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