A Case of Mediastinal Paravertebral Mullerian Cyst Resected Using Robot-Assisted Thoracoscopic Surgery

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Case Report

A Case of Mediastinal Paravertebral Müllerian Cyst Resected Using Robot-Assisted Thoracoscopic Surgery

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Although various tumors develop in the mediastinum, mediastinal Müllerian cysts, a new category of mediastinal tumors, are rare. We report the case of a 42-year-old woman with an incidentally discovered paravertebral mass at the level of the left 5th thoracic vertebra. Chest computed tomography revealed a 14-mm paravertebral mass, and chest magnetic resonance imaging revealed a cyst. The posterior mediastinal cyst was resected using robot-assisted thoracoscopic surgery. Hematoxylin and eosin staining revealed the presence of ciliated columnar epithelium inside the cyst wall, without cytologic atypia. Immunohistochemical staining showed the presence of estrogen receptor, progesterone receptor, and paired box gene 8. The final histopathological examination revealed that the cystic mass was a Müllerian cyst.

Keywords: Müllerian cyst, mediastinal tumor, robot-assisted thoracoscopic surgery

Introduction

Posterior mediastinal tumors are commonly known as bronchial cysts, neurological tumors, lymphomas, or esophageal cysts. However, Müllerian cyst (MC), which is the remnant of the Müllerian duct, is rare.[1-3] Mediastinal MC, categorized as a new mediastinal tumor, was first reported by Hattori in 2005.[1] Herein, we reported a case of posterior mediastinal MC resected by robot-assisted thoracoscopic surgery (RATS).

Case Presentation

A 42-year-old female presented a posterior mediastinal tumor on chest computed tomography (CT) during a medical check-up. No symptoms were observed and laboratory data did not reveal any abnormal findings. Plain chest CT revealed a well-defined paravertebral mass of 14 × 7-mm in size at the level of the left 5th thoracic vertebra in the posterior mediastinum (**Figure 1A**). Chest magnetic resonance imaging revealed a cystic mass with low signal intensity in T1-weighted images and high signal intensity in T2-weighted images (**Figure 1B and C**). Although the differential diagnosis

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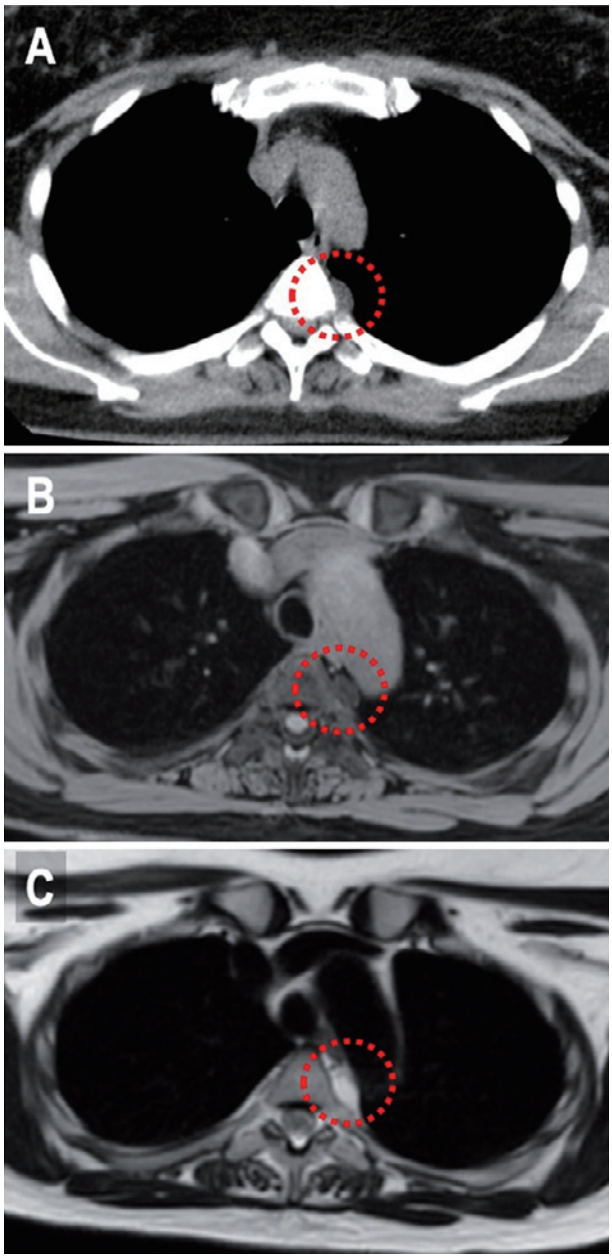

Figure 1. Preoperative images. (**A**) Plain chest computed tomography revealed a well-defined 14 × 7 mm paravertebral mass at the level of the left 5th thoracic vertebra in the posterior mediastinum. (**B**) Chest magnetic resonance imaging (MRI) shows a cystic mass with low signal intensity on T1-weighted images. (**C**) The cystic mass shows high signal intensity on T2-weighted MRI images.

showed that the mass was a schwannoma, a preoperative diagnosis of the bronchial cyst was made; informed consent was obtained from the patient and surgery was performed.

Under general anesthesia, the patient was intubated with a double-lumen endotracheal tube and placed in the right lateral decubitus position. The RATS excision of the posterior mediastinal tumor was performed using a three-port incision. As the second arm, an 8-mm camera port was placed in the 8th intercostal space (ICS) in the mid-axillary line. Two 8-mm assisted ports were inserted in the 6th ICS anteriorly in the anterior-axillary line as the first arm and the 8th ICS in the posterior axillary line as the third arm (**Figure 2A**). After positioning the da Vinci® Xi (Intuitive Surgical, Sunnyvale, CA, USA), the first and third arms were mounted in two assisted ports with bipolar fenestrated grasping forceps and Maryland bipolar forceps, respectively. A rigid 30° oblique viewing endoscope was used. Carbon dioxide (CO_2) was insufflated at a pressure of 5 mmHg. Adjacent to the distal aortic arch, a cystic mass was visible in the 5th ICS through the parietal pleura (**Figure 2B**). First, the cystic mass was dissected from the parietal pleura by grabbing and pulling the lower edge of the cystic mass (**Figure 2C**). Since the cystic mass was discontinuous with the intercostal nerves and vessels, a complete dissection was easily performed from the posterior mediastinum (**Figure 2D**). The resected mediastinal cystic mass was placed in a bag and removed through the assisted port of the third arm, followed by the insertion of a 21-Fr silicon tube through the camera port. Macroscopic observation revealed that the resected cystic mass measured 12 × 7 × 4 mm and was filled with fluid. The operative time was 39 min and the estimated blood loss was only 1 g.

Histopathologically, hematoxylin and eosin staining revealed ciliated columnar epithelium inside the cyst wall without cytologic atypia, and immunohistochemical staining revealed the presence of estrogen receptor (ER), progesterone receptor (PgR), and paired box gene 8 (PAX8) (**Figure 3**). Based on these findings, the final histopathological examination diagnosed the cystic mass as MC. The patient's postoperative course was uncomplicated.

Discussion

Various tumors develop in the mediastinum. Posterior medial tumors are commonly known as bronchial cysts, neurological tumors, lymphomas, and esophageal cysts. Of the mediastinal tumors, the mediastinal MC, the remnant of the Müllerian duct, is a newly established entity and is rare.[1-3] MC appears anywhere along the path of

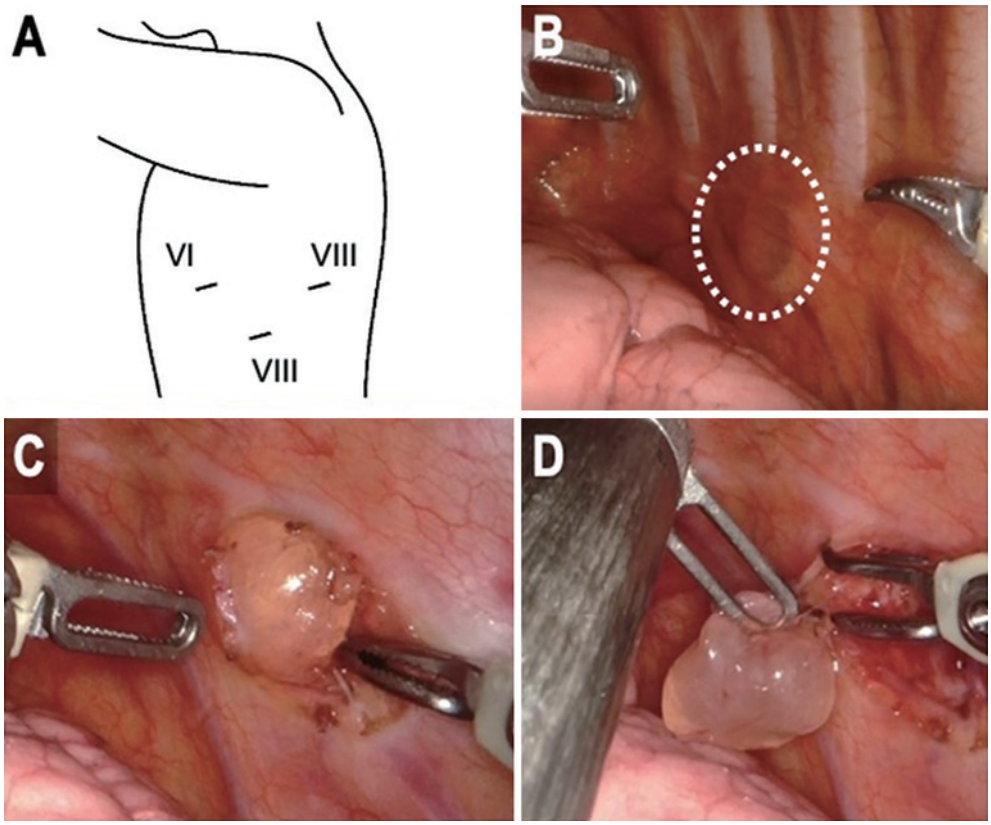

Figure 2. Intraoperative photographs. Configuration of port incisions in robot-assisted thoracoscopic surgery (RATS) and intraoperative photographs. (**A**) In the illustration, the lines marked with VI and VIII indicate the 6th and 8th intercostal spaces (ICSs), respectively. An 8-mm camera port is placed at the 8th ICS in the mid-axillary line as the second arm. Two 8-mm assisted ports are inserted at the 6th ICS anteriorly in the anterior-axillary line as the first arm and the 8th ICS on the posterior axillary line as the third arm. (**B**) When the inside of the thoracic cavity is observed with a rigid 30° oblique viewing endoscope, a tumor visible through the parietal pleura is found at the level of the left 5th thoracic vertebra. The tumor is indicated by the white dotted circle. (**C**) The lower end of the tumor is gripped and pulled with robotic instruments, and the tumor moves from the parietal pleura. (**D**) Since the cystic mass is discontinuous with the intercostal nerves and vessels, a complete incision is easily made from the posterior mediastinum. All three photographs (**B**-**D**) are extracted from digital video data.

Müller duct regression.[4,5] MC is commonly found in the pelvis in males but has also been reported to appear in the retroperitoneum and mediastinum in females.[3,6] The cause of MC in the mediastinum remains unknown. Sekimura et al. investigated and reported 25 cases of MCs in detail. Mediastinal MCs originate more on the left side of the thoracic cavity, with 14 cases (56%) around the Th4 vertebra, with a median tumor size of approximately 30 mm. Asymptomatic cases were found in 11 patients (44%), and cough was the most common symptom in six patients (24%). The cyst was filled with serous fluid. There were no postoperative recurrences of mediastinal MCs.[6] Immunohistochemistry may be useful for diagnosis in difficult cases. ER and PgR are the best markers of MCs. Of the 25 reported cases, 22 (88%) were positive for ER and 21 (84%) were positive for PgR. In addition, PAX8 is a member of the paired box gene family that plays an important role in organogenesis in the Müller system and is useful in the diagnosis of MCs.[7] Meanwhile, these patients are negative or occasionally weakly positive for cytokeratin 5/6.[2,8-10] In this case, ER, PgR, and PAX8 were positively stained.

Batt et al. suggested that MC is a choristoma of the primary Müller duct, and the lesion in which the epithelium of the Müller duct remained in the 3rd to 5th thoracic vertebrae was found to be the source of the Müller duct. The remaining Müller ducts are believed to expand and form cystic lesions due to abnormal hormonal stimulation.[3,11-13] In this case, the MC had appeared at the 5th thoracic vertebral level.

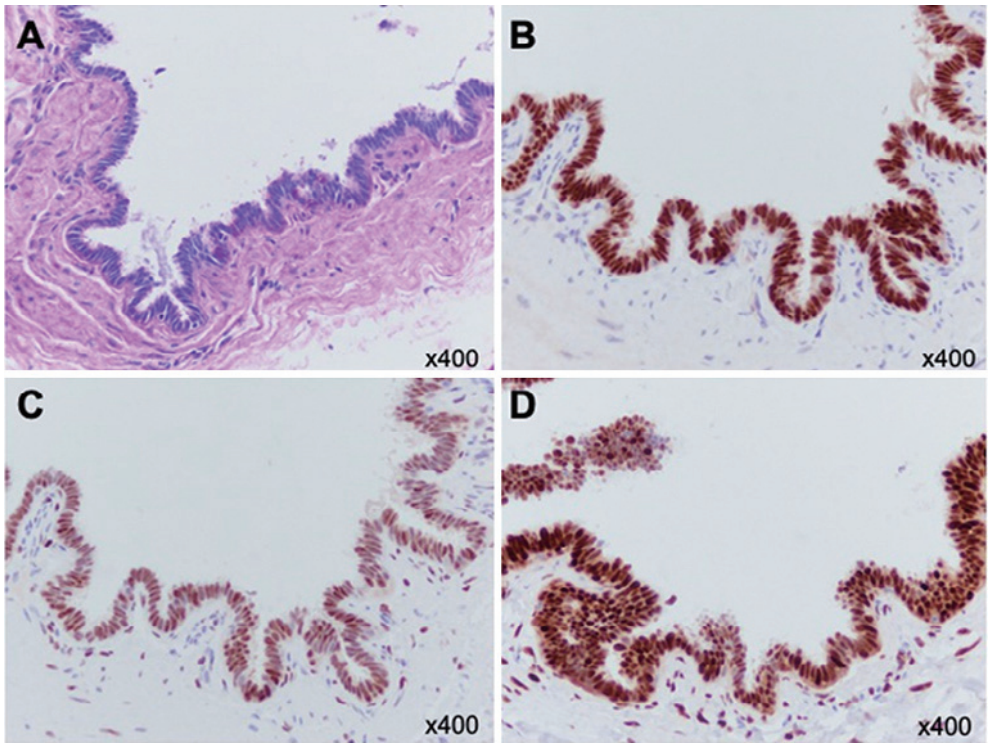

Figure 3. Histopathological findings of the resected cystic mass. (**A**) Hematoxylin and eosin staining revealed the presence of ciliated columnar epithelium inside the cyst wall, without cytologic atypia. (**B**) Immunohistochemical staining showed that the lining cells expressed estrogen receptors strongly and uniformly. (**C**) The lining cells strongly express the progesterone receptor. (**D**) The lining cells strongly and uniformly express the paired box gene 8.

MCs are commonly developed in male pelvises. Regarding sex differences in mediastinal MCs, all posterior mediastinal MCs appear in premenopausal and postmenopausal females in the first report.[1-3, 5-7, 9-11, 14] Since cells in the MC wall are positive for ER and PgR, the involvement of female hormones is suspected. MCs are also considered associated with obesity and gynecological history, such as hormone replacement therapy, hysterectomy, ovariectomy, and artificial abortion.

In this case, RATS mediastinal tumor resection was performed. Many MCs appear more in the posterior mediastinum, thoracic cavity on the cranial side, and left side of the thoracic cavity. Because the aorta is on the left side of the thoracic cavity, in surgery for middle or posterior mediastinal tumors near the aorta, the aorta can interfere with the surgical procedure. One of the limitations of RATS is that it must be performed in a two-dimensional field of view, with a special long and stiff endoscopic instrument in the thoracic cavity. Being surrounded by the heart, descending aorta, and vertebral body, the posterior mediastinum has a narrow space, and the robotic surgery system, which allows surgeons to perform complicated operations with delicate instruments, is useful for resecting posterior mediastinal tumors, including MC, and provides greater instrumental maneuverability and a wider range of motion for surgeons with a three-dimensional view.[15-17]

Conclusion

We report a rare case of a posterior paravertebral MC resected using RATS. A new category of mediastinal tumors, mediastinal MC, is required for the differential diagnosis of posterior mediastinal tumors.

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Conflicts of Interest: Masato Kanzaki received honoraria from Intuitive Surgical, Inc., Japan. The other authors have no conflicts of interest to declare.

Author Contributions: Akira Ogihara and Masato

Kanzaki conceived the study design. Yoji Nagashima provided the pathological findings. All authors conducted this study and collated the manuscript as a group effort. Akira Ogihara wrote the first draft of this manuscript. Masato Kanzaki assisted Akira Ogihara in preparing this manuscript. All authors have read and approved the final manuscript.

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