Progressive Enlarging Symptomatic Mediastinal Mullerian Cyst in a Female Patient with High Estradiol Level

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ABSTRACT

Mullerian cyst is a very rare disease. We reported the case of a 48-year-old woman with a gynecologic abnormality and high estradiol having a progressively enlarging posterior mediastinal cystic mass that caused intermittent chest tightness. The computed tomography and magnetic resonance imaging showed a well-defined cystic mass with homogenous fluid content located in right paravertebral area. The mass was surgically removed and was proven to be Mullerian cyst pathologically.

CASE REPORT

A 48 year old woman visited our hospital for help due to intermittent chest tightness for more than one year. There was no aggravating or relieving factor for the chest tightness. Her medical history showed that she had bilateral corpus luteum cyst, metabolic syndrome, end stage renal disease and had instrumental fixation after unknown neck injury.
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cells with fibrous stromal tissue (Fig. 4). Smooth muscle layer can be seen in the wall focally. The lining epithelial cells are immunoreactive for cytokeratin AE1/AE3, calretinin, estrogen receptor (ER) and progesterone receptor (PR), while non-reactive for thyroid transcription factor (TTF) -1 and cluster of differentiation (CD) 31 stains. Neither cartilage nor sero-mucinous glands are seen in the cystic wall. The final diagnosis is posterior mediastinal Mullerian cyst. The chest tightness symptom was relieved after surgery and the patient was discharged from our hospital uneventfully. Neither recurrence of the chest tightness, nor the recurrence of the mass is noted in the follow up.

Figure 1. Chest radiographs reveal a progressive enlarging right perihilar soft tissue density (arrow) a. 8 years before admission b. 3 years before admission c. One year before admission and d. At admission.
Figure 2. Chest computed tomography showed a well-defined cystic mass in mediastinum. a. Axial and b. Coro-nary view demonstrated the mass (arrow) being homog-enous low density, measuring 5.1 cm in maximal diameter, located in right costovertebral area of posterior medi-astinum and being associated with right main bronchus indentation (arrowhead).

Figure 3. Chest MRI demonstrated a paravertebral mass at right posterior mediastinum (arrow). a. Axial T1-weighted image showed homogeneous hypointensity. b. Axial T2-weighted image with fat suppression showed homogeneous hyper-intensity. c. Sagital balanced steady-state free precession image showed the close relation of the mass and the vertebras.
DiSCUSSION

Mediastinal cysts comprise 12 to 30% of all primary mediastinal masses [2, 3]. The spectrum of mediastinal cysts was classified by their etiology, including bronchogenic, esophageal duplication cysts of foregut origin, mesothelial cysts, thymic cysts, and other miscellaneous cysts, such as Mullerian cyst. The primary symptoms of mediastinal cysts are chest pain, dyspnea, and cough. About 36.8% patients are symptomatic [1].

The Mullerian cyst maybe defined as a ciliated columnar epithelium surrounded by a thin smooth muscular layer with immunoreactive for ER and PR expression. Most of Mullerian cyst occurred in ovary. Extra-genitourinary Mullerian cyst is rare entity, those developed in mediastinum, is even rarer. The previous reported Mullerian cysts of mediastinum (15 cases) all occurred in women and located in paravertebral regions.

Several factors accompanying Mullerian cyst had been reported, amongst hormone replacement therapy, obesity, previous gynecologic diseases are the mostly reported [4, 5]. Our patient is overweight and had history of bilateral corpus luteum cysts being resected, and moreover, an abnormal hormonal profile with high serum estradiol level. The clinical presentation are included cough (5/13), chest pain (3/13), and asymptomatic (5/13). The association between lesion size, location, and symptomatic presentation is not well correlation due to limited reported cases. The
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lesion size ranges from 1.5 to 5.0 cm at diagnostic time and the lesion size of symptomatic patient is also variable. The smallest symptomatic lesion size is 1.5 cm [5-7]. However, no previously report has clearly shown that the Mullerian cyst can grow overtime. In our case, the images have shown the Mullerian cyst enlarges significantly over time.

In conclusion, for patient with mediastinal cysts, patient characteristics, such as female gender, overweight, previous gynecological history, and abnormal hormonal profile are clues for diagnosis of mediastinal Mullerian cyst.

REFERENCES