Posterior Mediastinal Mullerian Cyst (Hattori Cyst) in a Perimenopausal Woman: A Case Report and Review of the Literature

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ABSTRACT

Several types of congenital cysts can be found in the mediastinum accounting for 12-30% of all mediastinal masses. Hattori was the first to describe a Mullerian cyst in the mediastinum in 2005. Mullerian cyst is a rare type of congenital cyst that can rarely arise in this location. Till now, 57 cases of Mullerian cysts arising in the mediastinum have been reported with the majority affecting women in perimenopausal age. Herein, we present a case of a posterior mediastinal Mullerian cyst in a middle-aged woman who was treated successfully by surgical resection.

Key Words: *Posterior mediastinum, Mullerian cyst, Perimenopausal.*

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INTRODUCTION

Congenital mediastinal cysts account for 12-30% of all mediastinal masses. Guided by the clinical picture and radiological imaging, the Mullerian cyst should be included as one of the differential diagnoses of mediastinal lesions.[1] A Mullerian cyst is a rare type of congenital cyst that can arise in many locations, other than genitourinary organs or pelvis, like the mediastinum. Here, it can be found in the posterior and middle mediastinum.[2] However, Mullerian cysts are more frequent than what is assumed and reported in the literature.[2,3] In 2005, Hattori was the first to describe and establish a distinct entity of Mullerian cysts in the mediastinum,[2,3] which is important as they are often misdiagnosed preoperatively, and they also seem not to be so rare.[3] Since then, to the best of our knowledge, only 57 cases of Mullerian cysts arising in the mediastinum have been reported.

CASE REPORT

We report a case of a 57-year woman with a significant gynaecological history (chronic abnormal uterine bleeding and salpingectomy for ectopic pregnancy), who had an incidental discovery five years ago, of a left paravertebral, T5-6, round lesion located in the posterior mediastinum measuring 2.5 × 3 cm on a chest CT scan, with radiological findings suggesting a cystic lesion (Figure 1).

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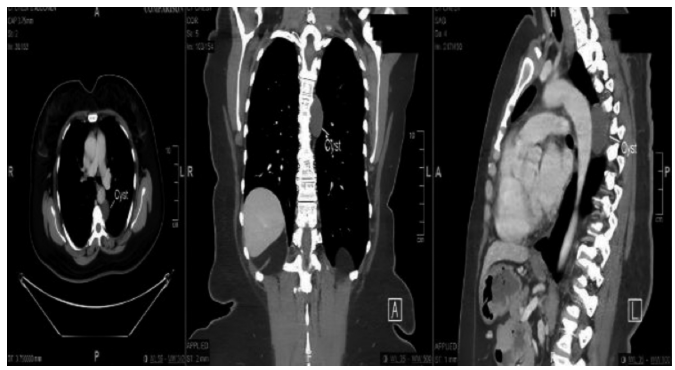

Figure 1: Axial, coronal and sagittal views from CT scan (from left to right) showing a posterior mediastinal paravertebral cyst.

Thoracic MRI showed a diffuse T2 signal signifying fluid and there was no communication with the spinal canal (Figure 2). The lesion remained stable radiologically over 5 years of follow-up till the patient agreed to surgical management. The patient underwent surgical resection of this cyst by uniport video-assisted thoracoscopy surgery (uVATS).

Histopathological examination of formalin-fixed, paraffin-embedded, and Haematoxylin and Eosin (H&E)-stained tissue sections revealed a thin cystic wall supported by fibrous stroma and lined by ciliated columnar epithelium (Figure 3 A, B).

Immunohistochemistry (IHC) showed diffuse nuclear staining for Estrogen Receptor (ER) and Wilmes' Tumour Gene 1 (WT-1), (Figure 3 C) and cytoplasmic positivity for CK7 and Pan-CK (AE1/AE3), while Calretinin, TTF-1, S-100, CDX-2 and P53 were negative, Ki-67 was low to intermediate. The cyst was diagnosed pathologically as a mediastinal Müllerian cyst. The patient had an uneventful postoperative recovery.

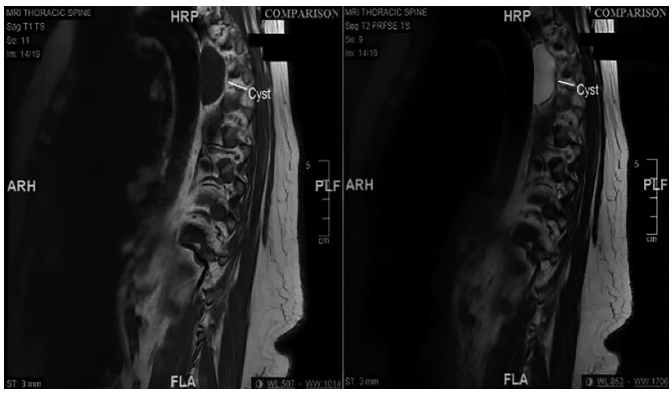

Figure 2: Hypointense appearance of the parasternal cyst in T1-weighted MRI (left) and hyperintensity in T2-weighted MRI (right) indicative of cystic lesion.

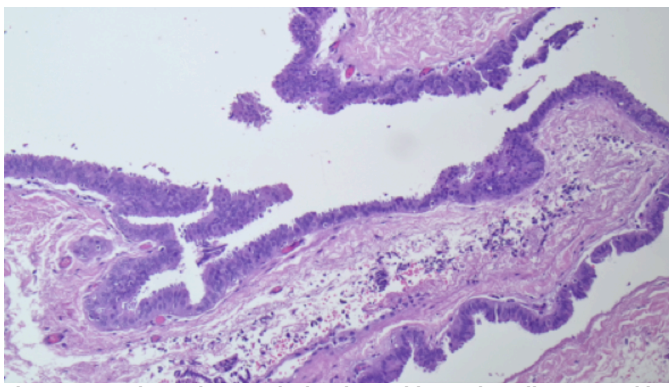

Figure 3A: A photomicrograph showing a thin cystic wall supported by fibrous stroma and lined by cuboidal epithelium (H&E, ×100).

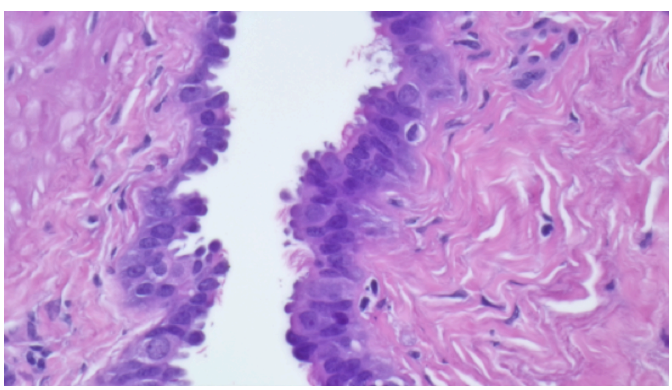

Figure 3B: A photomicrograph demonstrating the cyst lining composed of the ciliated epithelium (H&E, ×200).

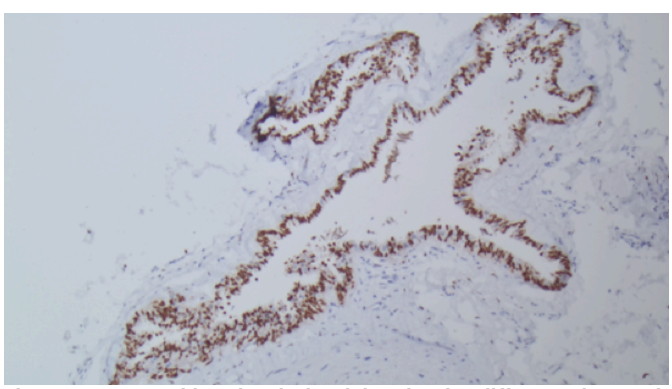

Figure 3C: Immunohistochemical staining showing diffuse nuclear positivity for ER. (ER, ×100). WT-1 showed similar reactivity.

DISCUSSION

Mullerian cysts are more common in perimenopausal women (40-60 years old), with a few exceptions in the younger age group.[2] Also, a relationship between a Mullerian cyst, obesity, and the presence of a significant gynaecological history such as menstrual irregularity or hysterectomy, was reported by Thomas and Dulmet in a retrospective study on 9 cysts with a Mullerian differentiation.[4]

Mullerian cysts are known to express ERs and progesterone receptors (PRs) on IHC, which is a helpful test to rule out more common differential diagnoses such as thymic cysts, bronchogenic cysts, oesophageal cysts, and celomic cysts.[3] All these cysts share similar histological findings of thin cyst walls supported by fibrous stroma lined mainly by ciliated columnar epithelium.[3]

These cysts are often misdiagnosed as bronchogenic cysts due to the presence of ciliated epithelium. However, the absence of cartilage and the positive ER and PR staining will differentiate between the two.[4]

Mullerian cyst aetiology remains unclear and is a subject of interest. Batt *et al*. described Hattori cyst pathogenesis by mullerianosis theory, which is described as an endosalpingeal, endometrial, or endocervical tissue that was misplaced during organogenesis forming a Mullerian choristoma.[5] The confidence of diagnosing Mullerian choristomas increases when the three Mullerian tissues (endosalpingeal, endometrial, or endocervical tissues) are present. However, a choristoma, where only one Mullerian tissue type is present, can be diagnosed with a high degree of probability, when three criteria are met: i) no evidence of endometriosis; ii) no direct communication with the endocervix, endometrium, or endosalpinx; and iii) no history of reproductive organs surgery.[5]

Despite apparently benign behaviour, and no reports of recurrence of Mullerian cysts, a case was reported by Skancke *et al*. of unusual, multiple, three large mediastinal Mullerian cysts, presenting with a history of respiratory distress due to compression of lungs and major airways, which were treated successfully by thoracoscopic aspiration of these cysts followed by surgical resection.[6]

The definitive treatment of Mullerian cyst is surgical resection, which can be done by minimally invasive methods including robot-assisted thoracoscopic surgery (RATS)[7,8] or by uVATS,[9] as demonstrated in the present case.

Future studies with regular follow-ups to assess the clinical outcomes, in comparison to other cysts found in the mediastinum, are warranted. In addition, we recommend considering Mullerian cyst as one of the differential diagnoses whenever encountering a posterior mediastinal cystic lesion, particularly in perimenopausal women.

DISCLOSURE:
This manuscript was presented at the Saudi Thoracic Society held on 9th November 2023.

PATIENT'S CONSENT:
Informed consent for patient information to be published in this article was not obtained because no personal or identifiable information/images are published in this article.

COMPETING INTEREST:
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AUTHORS' CONTRIBUTION:
SKK, FAS, WMH: Drafting and revising manuscript.
SWH: Literature review and manuscript writing.
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