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Thoracic Endometriosis Detected During Medical Thoracoscopy in a Tertiary Care Setting - A Retrospective Observational Study

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ABSTRACT

Background: Thoracic endometriosis is a very rare clinical situation mostly presenting with pleural diseases, that is often misdiagnosed or missed due to the lack of specific clinical features. This report aims to explore the clinical characteristics, thoracoscopic findings, histopathology, treatment, and prognosis of women with thoracic endometriosis, providing a reference for the diagnosis and management of thoracic endometriosis.

Design/Methods: Clinical data of 3 patients diagnosed with pleural endometriosis from among 50 consecutive thoracoscopic procedures conducted in a tertiary care hospital in North Kerala were analysed from electronic hospital records. The clinical features, thoracoscopic findings, histopathological features, treatment process, and prognosis were summarized.

Results: Among the 3 female patients with pleural endometriosis, there were 2 cases of catamenial pneumothorax and one case of catamenial haemothorax. The age ranged from 43 years to 44 years. All the 3 cases were diagnosed by thoracoscopic pleural biopsy.

Conclusions: Thoracic endometriosis syndrome is extremely rare, and clinical diagnosis is always missed or delayed due to nonspecific symptoms. Diagnosis mainly relies on pathological findings and immunohistochemical detection. Most cases were managed conservatively with hormonal therapy or by inducing surgical menopause.

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Introduction

Endometriosis is a common gynaecological disorder, affecting 10–15% of women of reproductive age. It is considered to be due to extrauterine growth of endometrial tissue, including endometrial glands and stroma. The ectopic tissue is typically located in the peritoneal cavity, most often in the pelvis, but endometriosis has been reported in nearly all body compartments [1]. Although rare, thoracic involvement is the most frequent extra-abdominopelvic site of endometriosis [2]. Up to 80% of women with thoracic endometriosis present with concomitant pelvic endometriosis. Thoracic endometriosis syndrome (TES) is the term used to denote various clinical manifestations resulting from the presence of functional endometrial tissue in a thoracic structure. Endometrial tissue is seeded in visceral or parietal pleura, lung parenchyma, airways, or diaphragm [2]. Clinical manifestations are more likely to occur during menses, because of the hormonal responsiveness of ectopic endometrial tissue [2]. TES includes five well-recognized clinical entities grouped into two forms, namely the pleural form with catamenial pneumothorax (CP), non-catamenial endometriosis-related pneumothorax (NCP), catamenial haemothorax (CHt); and the pulmonary form with catamenial haemoptysis (CH), and lung nodules [2,3]. Approximately 90% of patients with TES experience catamenial thoracic pain, and different entities may be associated [2]. The right hemithorax is involved in more than 90% of all forms, with the exception of nodules (70%) [2]. Video thoracoscopy was the preferred surgical technique (84%; 95% CI, 66%-96%). Intraoperative evaluation revealed the presence of diaphragmatic anomalies in 84% of cases (95% CI, 73%-93%) [1].

Objectives

1. To evaluate the clinical profile, diagnosis, thoracoscopic findings, histopathology, and management of thoracic endometriosis.
2. To provide a clinical pathway for the diagnosis and management of thoracic endometriosis.

Study Design and Methods

This is a retrospective electronic data-based study of 50 consecutive medical thoracoscopy done from 1st January 2022, using a semi-rigid thoracoscope under conscious sedation in a tertiary care hospital in North Kerala.

The study period was from January 2022 to March 2023

Inclusion Criteria: All completed medical thoracoscopy in which an adequate biopsy specimen was obtained for a definite histopathological diagnosis.

Exclusion Criteria: When the procedure was terminated before complete inspection of pleura or a histopathological specimen was not obtained.

Data Analysis: Demographic clinical, thoracoscopic, histopathological, and treatment data were retrieved and entered into an Excel sheet. Mean and percentage were calculated.

The study was done after getting permission from the institutional ethics committee.

Results

Out of the 50 consecutive cases of medical thoracoscopy, 3 cases were diagnosed as thoracic endometriosis (6%). All of them were in their reproductive age group ranging from 43 years to 44 years (Mean- 43.3 Years). One patient completed her family, one was unmarried and the third patient had primary infertility (Table 1). Of the 3, two cases were catamenial pneumothorax (Figure 1) (66.6%) and the third case was catamenial haemothorax (33.3%). Medical thoracoscopic findings included right-sided haemothorax (Figure 2) in case- 3 with diaphragmatic pleura showing chocolate-coloured cysts and charry black indurated lesions classically described as "Gunshot" lesions (Figure 3). Biopsy from these sites showed endometrial tissue confirming the diagnosis. The other 2 cases were recurrent pneumothorax treated multiple times with intercostal drainage. Medical thoracoscopy was performed during the 4th recurrence in one case and during the first recurrence in the other. Thoracoscopy showed chocolate-coloured nodules and classical charry black lesions on the diaphragmatic pleura (Figure 4) and histopathology confirmed the diagnosis of pleural endometriosis (Figure 5). Both cases of catamenial pneumothorax were treated with talc pleurodesis without any further recurrence. Catamenial haemothorax was initially treated with hormone therapy to suppress menstruation. Catamenial haemothorax and one case of catamenial pneumothorax had concomitant pelvic endometriosis and were subjected to hysterectomy and bilateral salpingo-oophorectomy (Table 2). The other case of catamenial pneumothorax was put on initial hormonal treatment for 6 months and then discontinued. She did not report any further recurrences.

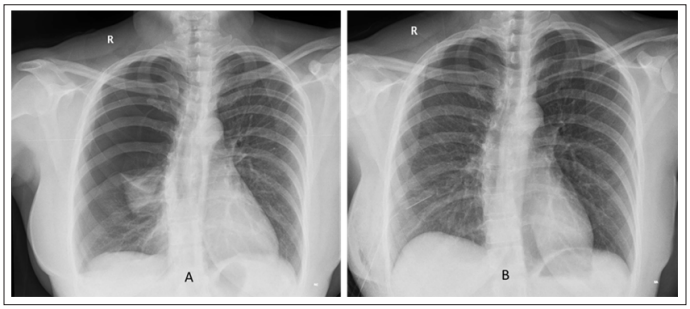

Figure 1: Catamenial Pneumothorax- 4th Recurrence (A), Chest X Ray after Medical Thoracoscopy and Pleurodesis(B).

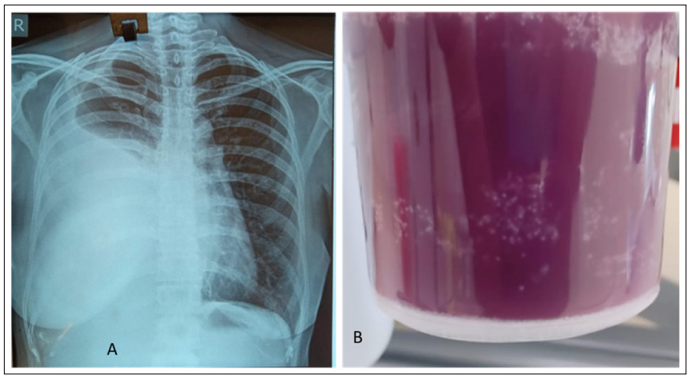

Figure 2: Right-Sided Moderate Pleural Effusion (A) and Aspirated Blood from Pleural Space (B)

Table 1: Demographic and Clinical Data of Patients with Thoracic Endometriosis

Case no.	Age	Family status	Diagnosis	Presentation	Pelvic endometriosis
1	43	Unmarried (Nun Sister)	Catamenial Pneumothorax	Right-sided Recurrent pneumothorax, first recurrence	Present
2	44	Primary infertility	Catamenial Pneumothorax	Right-sided Recurrent pneumothorax, 4th recurrence	Absent
3	43	Completed family	Catamenial haemothorax	Right-sided moderate haemothorax	Present

Table 2: Thoracoscopic Findings, Histopathology and Treatment offered to the Patients

Case no.	Thoracoscopic findings	Histopathology	Treatment	Follow up
1	Multiple charry black nodular lesion (Gunshot lesion) in the diaphragmatic pleura	Endometrial glands and stroma	Talc Pleurodesis with a hysterectomy and bilateral salpingo-oophorectomy	No recurrence
2	Multiple charry black nodular lesion (Gunshot lesion) in the mediastinal pleura	Endometrial glands	Talc Pleurodesis with hormonal therapy.	No recurrence
3	Multiple chocolate coloured cysts and gunshot lesions in the diaphragmatic pleura	Endometrial glands and stroma	Initial hormonal therapy and later hysterectomy and bilateral salpingo-oophorectomy	No recurrence

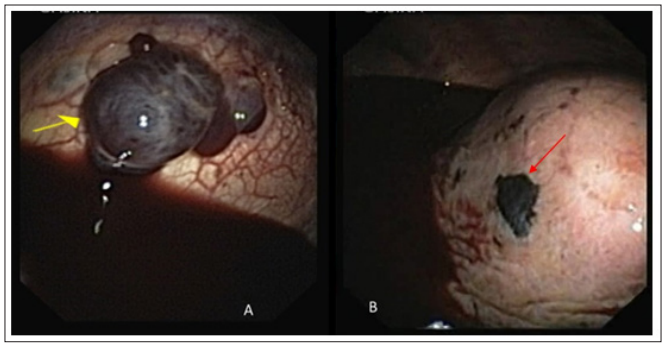

Figure 3: Multiple Chocolate-Coloured Cysts (Yellow arrow) and Gunshot Lesions in the Diaphragmatic Pleura (Red Arrow)

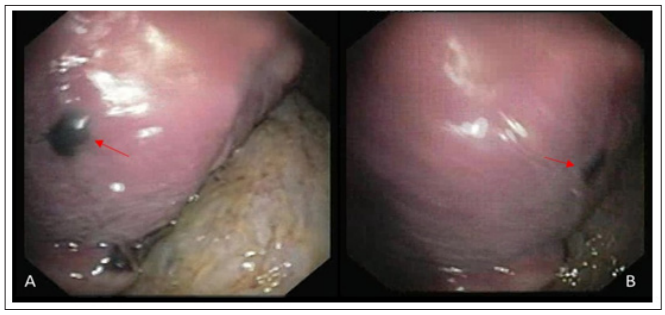

Figure 4: Multiple Charry Black Nodular Lesions (Gunshot Lesion) in the Diaphragmatic Pleura (Red Arrow)

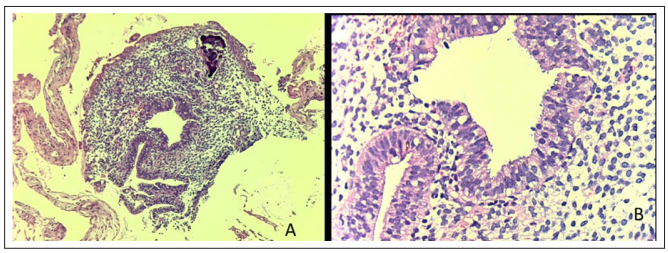

Figure 5: Histopathological Examination of the Biopsy Specimen Showed Endometrial Glands and Stroma (A- Low Power, B- High power)

Discussion

TES is considered rare, but may be underdiagnosed [2,4]. There were only 38 pathologically confirmed cases of TES in the literature till the year 2000 [5]. A study on 110 patients showed that the mean age at presentation was 35 ± 0.6 years, with a range of 15 to 54 years. Interestingly, the peak incidence for pelvic endometriosis is between 24 and 29 years, whereas the peak incidence for TES is approximately 5 years later [6]. All the 3 cases in this series were in their early forties.

Pneumothorax is the most common manifestation of thoracic endometriosis (73%). Catamenial haemothorax (CHt) is less common (14%), followed by CH (7%) and lung nodules (6%) [2]. Approximately 20% of women of reproductive age who experience spontaneous recurrent pneumothorax have thoracic endometriosis [3,7]. In our series 66.6% cases were pneumothorax. This is consistent with the available literature.

Catamenial haemothorax represents the second-most common manifestation of TES, occurring in 14 % of known cases, and affects the right side in about 80 % of the time. Wilkins et al. reported 15 cases of TES presenting with haemothorax and all of his cases were in the right hemithorax [8]. Concomitant pelvic endometriosis was found in 100 % of cases [6]. All the 3 cases in this series had right sided lesions and 2/3rd cases had concomitant pelvic endometriosis.

Diagnosis is often delayed until several episodes have occurred as the patient fails to associate symptoms with menstruation. One patient in this series had four recurrences of pneumothorax before a confirmative diagnosis was made. Pleural fluid cytology is usually not helpful. Level of cancer antigen 125 (CA-125) may be elevated in the serum and body cavity fluid of patients with endometriosis [9]. The concentration of CA-125 correlates with both the severity and the clinical course of the disease [5]. Diagnosis of TES is usually based on clinical grounds, after excluding other pulmonary diseases [2]. Symptoms have a catamenial pattern, occurring between 48 hours before and 72 hours after the onset of menses, and typically recurring during each cycle [3,7,10]. CP is defined by at least two episodes of pneumothorax occurring during this time interval. The right-side predominance of symptoms represents a diagnostic clue.

Chest X-rays in cases of pleural endometriosis usually reveal a pneumothorax or occasionally a pleural effusion [5]. Spiral CT may show pleural or diaphragmatic thickening in involved areas [11]. In the present case, CT revealed a right-sided moderate pleural effusion without any pleural thickening or diaphragmatic lesions. Although the CT aspect of TES is poorly specific, CT remains the first-line imaging method, as it can rule out other diagnoses and map the lesions for surgery if necessary [4,12-15].

Medical Thoracoscopy is the investigation of choice as it enables visualization of typical lesions and biopsy from these lesions (Figure 4). Pleura reveals multiple, dark red or blue nodules (Raspberry nodules), and cysts on the diaphragmatic pleura. In this case series, multiple dark brown cysts were seen, the largest measuring 2x1.5 cm. Most of them were bleeding on touch. Apart from that, a few charry black nodules were also seen, which are the characteristically described “gunshot” lesions diagnostic of pleural endometriosis.

Microscopically, typical endometriosis consists of both endometriotic glands and stroma. The glands usually have an endometrioid appearance ranging from inactive, and proliferative to hyperplastic. The endometriotic stroma characteristically resembles eutopic inactive or proliferative endometrial stroma [16]. Immunohistochemically, in the series reported by Flieder and associates, most glands showed cytoplasmic positivity with broad spectrum cytokeratin, cytokeratin 7, and BER-EP4, and strong nuclear staining for estrogen and progesterone receptors [9]. Estrogen and progesterone receptors were present in endometriotic glands and stroma in a lower concentration than in eutopic endometrium [17,18].

Therapy for TES includes the suppression of endometrial tissue and the prevention of further pelvic seeding. Medical therapy should be considered as the first line of treatment [19]. Ghio et al., reported a case of catamenial pneumothorax with chest pain and used medroxyprogesterone acetate as therapy [20]. According to Light, hormonal therapy (progestational agents, danazol, and leuprolide acetate) fails in at least 50 % of cases [21]. Medical treatment for endometriosis symptoms (with or without surgery) is generally needed for longer periods of time because of the chronic and recurrent nature of the disease. Progestins may be an appropriate alternative for the medical management of endometriosis, as these agents are relatively well tolerated, and have a more limited metabolic impact than other agents [22]. Treatment with GnRH

analogs, such as leuprolide, is limited to only 6 months because these agents induce a hypoestrogenic state that substantially decreases bone mineral density. Poor tolerability represents the major drawback of danazol as a treatment for endometriosis. This agent has both androgenic and anabolic properties. Pleurodesis may be considered as a means of preventing the pleural recurrence [23]. Patients with concomitant pelvic endometriosis may be subjected to surgery (Bilateral salpingo-oophorectomy), if they have completed their family.

Conclusion

Thoracic endometriosis is a rare, unusual lesion that may mimic various conditions. Diagnosis of TES is challenging, as these women's symptoms may not immediately be attributed to endometriosis, and as some radiological abnormalities (especially pneumothorax and haemothorax) are non-specific. The key features are the temporal relation with menses, cyclical changes in radiographic abnormalities, right-sided predominance, and the distinctive posterosuperior location of diaphragmatic lesions. Diagnosis of thoracic endometriosis syndrome may be better established on the basis of clinical grounds such as catamenial manifestations, proven pelvic endometriosis, and susceptibility of thoracic symptoms to hormonal manipulations. Medical thoracoscopy provides better visualization of endometrial lesions in the pleura and enables biopsy to confirm pleural endometriosis.

Limitations

Thoracic endometriosis is a rare condition and hence, the number of subjects included in this study was only three. Generalization and conclusions from this small group remain a drawback of this observational study.

Conflicts of Interest: Nil

Funding: None received

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