CASE REPORTS

Extra Pelvic Endometriosis and Catamenial Pneumothorax

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Summary

Extra pelvic endometriosis is rare and its presentation is varied. A case of pulmonary and umbilical endometriosis which presented as catamenial pneumothorax is presented. Due to poor response to medical treatment, a total abdominal hysterectomy and bilateral salpingo-oophorectomy was done to relieve the patient of her recurrent symptoms.

Key Words: Pneumothorax, Endometriosis, Catamenial pneumothorax, Umbilical endometriois

Introduction

Non-gynaecologic or extra pelvic endometriosis is fascinating due to the diversity of reported sites and the rather unusual symptoms and signs patients present with. We present here a rare case of pulmonary and umbilical endometriosis which presented as spontaneous pneumothorax in a medical unit. The etiology, pathophysiology and treatment of extrapelvic endometriosis is discussed.

Case Report

AA, a 45-year-old Para 2 was first seen on 16th January 1989 presenting with right pleuritic chest pain. She gave a two-year history of cyclical haemoptysis and slight bleeding from the umbilicus which occurred during her menses. She also had dysmenorrhoea throughout her menses which lasted for seven days. Her last childbirth was in 1982.

On admission, chest X-ray showed a small to moderate right pneumothorax and this was managed conservatively. Cytological examination of the sputum did not reveal any endometrial cells; sputum culture and sensitivity was also negative for any organisms. The umbilicus also appeared normal. A computerised axial tomography (CT) scan of the chest, abdomen and pelvis showed abnormal soft tissue at the cardiophrenic angle on the right side. The pelvic organs appeared normal. Fine needle aspiration biopsy of the soft tissue under CT control did not reveal any abnormal cells. A provisional diagnosis of pulmonary and umbilical endometriosis was made and she was commenced on oral Danazol 200 mg twice daily.

She however, was re-admitted four months later with similar problems. Chest X-ray done this time showed a moderately large right sided pneumothorax. A chest tube was immediately inserted. Following treatment for two weeks the residual pneumothorax was still noted. After discussion with the cardiothoracic team, a right thoracotomy, decortication, stapling of bullae and talc pleurodesis were undertaken. Histopathological examination of the excised nodules was compatible with endometriosis. Danazol therapy was continued for another two months. She subsequently defaulted follow up treatment. In May 1993, she was seen again with severe haemoptysis and bleeding from the umbilicus. The problem was discussed in detail with the patient and in view of her age and she having completed her family and poor response of extrapelvic endometriosis

to medical therapy, total abdominal hysterectomy and bilateral salpingo-oophorectomy was offered. She underwent surgery on 25th May 1993 along with excision of the umbilical nodule. The post-operative recovery was uneventful. Histopathological examination of the operative specimen showed the presence of endometriosis on the right ovary and the umbilical nodule with adenomyosis of the myometrium. She was seen again at follow-up a year later and was noted to be well.

Discussion

The symptoms and complaints associated with extra pelvic endometriosis are quite diverse and usually relate to the physiological function of the ectopic site. Symptomatology is usually related to the menstrual cycle as seen in this patient.

Pulmonary endometriosis has been described to manifest as either asymptomatic pulmonary nodule or as pneumothorax, haemothorax or cyclical haemoptysis as seen in our patient. Of 84 cases of pulmonary endometriosis reviewed in the world literature by Karpek *et al*[1], 63 (75%) presented as catamenial pneumothorax, 9 (10.7%) as catamenial haemothorax, 7 (8.3%) as cyclical haemoptysis and five (6%) as asymptomatic pulmonary nodules. Of the cases with catamenial pneumothorax, 95% occurred in the right chest and 22% had concurrent pelvic endometriosis.

Fenestration or defects of the diaphragm have been well recognised and are located more commonly in the right diaphragm. A theory has been proposed of a continuous current of peritoneal fluid circulating from the pelvis to the right upper quadrant of the abdomen which if true, would favour the right diaphragmatic area over other areas coming into contact with floating endometriotic particles during retrograde menstruation[2]. Another theory to explain the occurrence of endometriosis in such distant sites as the lung is the vascular-lymphatic dissemination theory.

Various diagnostic procedures performed to produce tissue diagnoses include pleuroscopy, bronchoscopy and thoracotomy. Laparoscopy with pneumoperitoneum if followed by pneumothorax, would suggest a diaphragmatic communication.

Needle biopsy of a pulmonary or diaphragmatic nodule (as undertaken in this patient) occasionally has been done to confirm the benign nature of the nodule (especially if asymptomatic). The immediate management (as carried out in this patient) is lung reexpansion and whenever necessary chest-tube drainage. Following that, hormonal suppression of ovulation with either testosterone, depo-provera, oral contraceptive pills, danazol or gonadotrophin releasing hormone have been tried with varying results. They generally have not been successful for extra pelvic endometriosis[3]. Recurrent or progressive disease however, is best treated by total hysterectomy and bilateral oophorectomy. Low dose oestrogen replacement does not seem to result in the recurrence of endometriotic symptoms.

Endometriosis of the umbilicus is rare and the incidence quoted is around 0.5-1.0 per cent of all extrapelvic endometriosis[3]. Diagnosis is from clinical (menstruating tumour) and histopathological examination. Treatment is by total excision of the nodule. In cases that recur or when complete surgical excision is not possible, or when reproduction is no longer desired bilateral oophorectomy with hysterectomy may be the appropriate treatment of choice.

References

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