DIAGNOSTIC DILEMMA

Diaphragmatic endometriosis

Michael J W Cooper, Peter Russell and P James Gallagher

Endometriosis is a relatively common condition usually found in the pelvis. However, lesions do occur outside the pelvis and, more rarely, in the upper abdomen. In the case reported here, the patient presented with chronic right shoulder tip pain. The diagnosis of extrapelvic endometriosis is often not considered in such circumstances. This patient's symptoms were relieved by surgical excision of the diaphragmatic lesion. ***(MJA 1999; 171: 142-143)***

Clinical record

A 19-year-old nulliparous woman presented with a history of three years of increasing right shoulder tip pain associated with menstruation. The pain had progressed over recent months to be virtually constant and debilitating, to the extent that sleeping was impaired. There was no evidence of catamenial (menstrual) pneumothorax, a common presenting sign in patients with diaphragmatic endometriosis. Two laparoscopies and a laparotomy had been performed in the two preceding years for pelvic endometriosis, and, although shoulder tip pain had been present, a definitive diagnosis of diaphragmatic involvement was not made. Therapy with progestogen and danazol had not reduced the pain.

Diaphragmatic endometriosis was provisionally diagnosed. The patient was admitted to hospital for planned laparoscopic resection of the endometriosis. At laparoscopy a large diaphragmatic deposit was noted at the base of the liver over the diaphragm, extending up to the inferior vena cava. Several small satellite lesions were noted on the surface of the diaphragm. Further deposits were noted at the left uterovesical fold, both uterosacral ligaments and in the left pararectal area. Time constraints and the patient's symptoms necessitated a decision to excise only the diaphragmatic disease initially. The liver could not be adequately mobilised laparoscopically and the procedure was converted to laparotomy via a subcostal incision. The satellite lesions and a full-thickness 8 cm by 4 cm section of diaphragm were excised, opening into the pleural cavity. The defect was oversewn and a chest drain placed at the conclusion of the procedure.

The patient noted complete resolution of symptoms in the recovery room, which has persisted 15 months after the surgery. There were no postoperative complications and the patient was discharged five days after the procedure. The pathology confirmed multiple foci of endometriosis involving the diaphragm, with adjacent areas of scarring and residual chronic inflammation (Figure).

Six months after the procedure the patient had another laparoscopy, at which the residual pelvic disease was excised. At this procedure the diaphragm was visualised, but, because of the position of the liver, the previous operative site was unable to be seen. The patient has remained pain-free and has subsequently conceived, miscarried and is currently pregnant again.

Discussion

Endometriosis is an enigmatic non-neoplastic disorder that is not particularly well understood, characterised by the presence of normal-appearing endometrial tissue outside the uterine cavity. It is most common in women in the reproductive years, but may be seen in girls before menarche, in postmenopausal women and even in men who have been castrated and are receiving oestrogens. The common sites are on or beneath the pelvic peritoneum, where it may, for example, involve ovaries, uterine ligaments, bladder and bowel, obliterate the cul-de-sac or obstruct ureters. Less commonly, it may produce intra-abdominal lesions or involve hypochondrial structures (liver, diaphragm)[1] and right-sided thoracic viscera, and, rarely, the extremities. The main symptoms include pelvic pain, infertility and menstrual irregularity, most commonly premenstrual spotting. Diaphragmatic involvement often causes recurrent catamenial pneumothorax.

A number of theories have been proffered for the pathogenesis of endometriosis. The most plausible of these is metaplasia of the peritoneum and/or the subperitoneal mesenchyme under the influence of metaplasia-inducing substances such as oestrogens and as yet unidentified factors liberated from degenerating endometrium.[2] The main alternative theory is of retrograde menstruation.[2] However, retrograde menstruation is probably a normal event and yet only 5%–10% of women develop endometriosis. If this theory were correct, we would expect endometriosis to develop and spread throughout the pelvis over time, and this is not noted in clinical practice.

In most cases the lesions are reasonably superficial, but they can at times be deeply situated or even locally "inva-

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in young men may relate to cultural expectations that men should be tough and resilient[14] — a suggestion that might be equally relevant in Australia, New Zealand, the United States and Canada. It is possible that a better understanding of cultural influences and how to positively modify them might be relevant to suicide prevention.

Acknowledgements

The study was funded by Griffith University and Queensland Health through their co-funding of the Australian Institute for Suicide Research and Prevention. We thank the Australian Bureau of Statistics for providing information on suicide and population numbers.

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(Received 14 Jan, accepted 7 Jun, 1999)

This article has been peer-reviewed and published on the *e*MJA site.
See http://www.mja.com.au/public/issues/iprs2/cantor/cantorframe.html

Time Capsule

The obstetrician's dilemma

"Obstetricians, not unreasonably, feel themselves to be a beleaguered species. Constantly under attack for adopting a 'high tech' approach to what is arguably a normal physiological process, they also find themselves the target of very expensive negligence claims if they fail to intervene successfully when problems occur."...

To some extent, obstetricians are victims of the technological advances and the media publicity of such advances within the specialty. This, coupled with the sometimes unrealistic expectations of modern pregnancy, labour and delivery, and with the adversarial nature of the law of torts that pertains to Australia, has led to an increase in litigation in obstetrics and, correspondingly, a significant projected increase in the cost of medical defence. One response to these increasing pressures could be an exodus of currently-practising obstetricians from the specialty such as is occurring in the United States. While such an exodus might fulfil the needs of the individual obstetricians who are involved, it would be devastating for the specialty and for the nation's women and children. Should a recruitment problem also occur, then standards surely would fall and litigation in the area of obstetrics could be expected to rise as resources become more and more limited. Other solutions to the problem of litigation in obstetrics should be explored....

The area that remains for the greatest potential change is that of the law itself. Widespread criticism has been levelled at the shortcomings of the tort system. The procedure for pursuing a claim may be lengthy and expensive and, thus, often available only to the "rich" or to those who are able to obtain legal aid. By definition, the process is adversarial and frequently causes doctors to close ranks and not to offer an adequate explanation to a plaintiff. The emphasis is on the establishment of fault, and cause and effect in injury cases turn the tort system into a lottery. Compensation is not based on need but on the ability to prove fault. Finally, only a small proportion of individuals who suffer medical injuries are compensated through the tort system and this may mean that the losses that are incurred as a result of injury are compensated inadequately, although other sources of compensation may be available.

Other criticisms of the current system are that it may be difficult to obtain the services of a solicitor with relevant expertise and that there may be difficulty in obtaining the services of doctors who are willing to act as expert witnesses for a plaintiff....

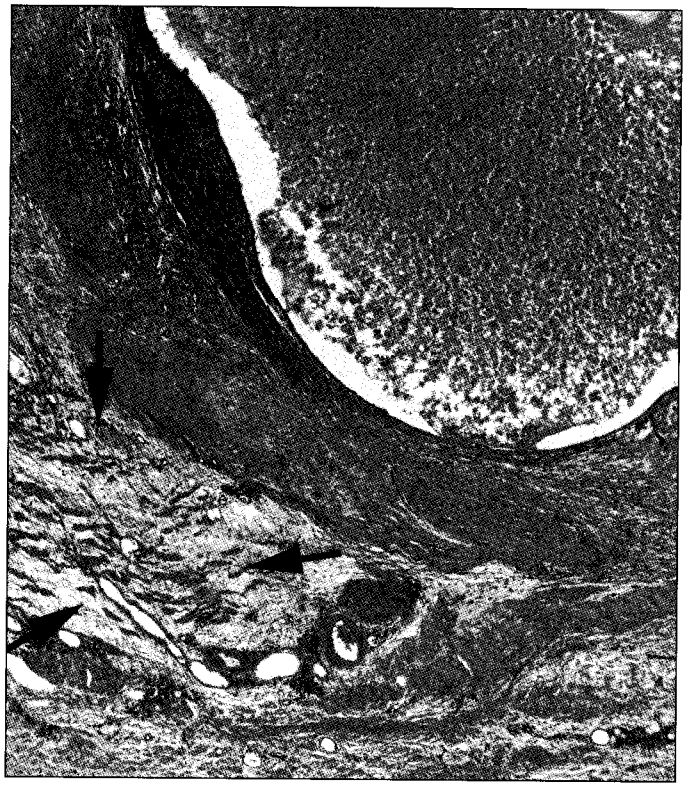
It is clear that obstetricians face a dilemma in practising their specialty in the face of increasing threats of medical litigation and increasing medical-insurance premiums. To follow the American example and simply to opt out of practice will prove to be disastrous for the specialty within Australia. Solutions to this dilemma must involve a consideration of the many factors that are entailed.

Obstetricians must set and maintain high standards of care. Realistic expectations of modern obstetric care must be communicated to consumers. The media must be responsible in publicizing advances in obstetrics and must not engender false hopes. The legal profession must examine the means by which plaintiffs have access to proper and adequate compensation and consumers must be counselled properly as to the effective means by which proper litigation is instituted. The accountability of medical practitioners must be maintained.

Finally, we must address the views that are embodied in the following statement:

> The law is a clumsy instrument for dealing with dynamic human relationships and as such should be a last resort as a solution for the individual, although it has a valuable function as a deterrent and a setter of standards. Education of both patients and doctors, consumer pressure and alternative dispute resolution are powerful agents of change which should be fully utilised in the hope that litigation will rarely need to be resorted to given the human suffering such action reflects.

From: *Med J Aust* 1989; 150: 545-546.



A small endometriotic cyst filled with old blood, having an epithelial lining and being surrounded by reactive scar tissue and fibrosis. Aponeurotic diaphragmatic tissue is seen as a wavy band running across the lower part of the figure and striated muscle fibres are seen at lower left (between arrows) (haematoxylin and eosin stain; original magnification x30).

sive", causing significant scarring and fibrosis not unlike a malignant process.

Deeply invasive endometriosis is invariably difficult to treat. Traditional medical therapy has not been successful. The regimens have been largely suppressive rather than curative, and no drug has yet been shown to eradicate endometriosis or effect a long-term cure.[3] In addition, significant side effects may be expected from many of these treatments. Unfortunately, the newer gonadotropin-releasing hormone analogues, which had offered promise, may only be used for a maximum of six months, mainly because they are associated with the development of osteoporosis,[4] and have not been shown to eradicate endometriosis.[3]

Traditional surgical interventions have involved either ablation or total hysterectomy and bilateral salpingo-oophorectomy. Ablative techniques appear to assist with mild to moderate disease but are no help in severe endometriosis and have the potential to cause significant damage, largely because of the risk of lateral thermal damage. As the depth of involvement may be particularly difficult to assess, these techniques often result in inadequate treatment, with residual disease remaining underneath the zone of treatment leading to "disease recurrence".[5] The use of hysterectomy, which has been championed often, unfortunately has no theoretical basis, and may make the situation worse by allowing invasive endometriosis to erode both bladder and bowel.[6] Oophorectomy may be of value by reducing the hormonal impetus for disease progression, but does not remove the disease and substantially increases the risk of postmenopausal problems such as osteoporosis and ischaemic heart disease.

Excisional techniques for deeply invasive disease have been demonstrated to be highly effective for relieving symptoms, with substantially less risk of recurrence than traditional therapy.[3,7] Unfortunately, the surgery is difficult and time consuming, with the potential for substantial complications.

This patient exemplifies both the significant symptoms that can be caused by unusual endometriotic deposits and the impressive results that can be achieved by aggressive excisional treatment.

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