Pyoderma gangrenosum, vesicovaginal fistula and endometriosis

A case report

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Summary

A 47-year-old multiparous woman presented with endometriosis, true urinary incontinence, and the mucosal and skin lesions of pyoderma gangrenosum. The subsequent investigations, treatment and course are discussed, as well as the possible causal interrelationship between the above three conditions.

Pyoderma gangrenosum (PG), originally described by Brunsting et al., 1 is a destructive inflammatory skin disease presenting clinically as necrotic ulcers with a characteristic purpurish undermined edge and surrounding erythema. These ulcers begin as erythematous papules, vesicles or pustules and heal on treatment, leaving an atrophic, 'cigarette-paper' scar. The histological appearance is nonspecific, the ulcer extending into the upper part of the dermis with an acute inflammatory cellular infiltrate at the centre. 2 Evidence of vasculitis may be present, although necrotizing vasculitis is not a conspicuous feature in most reported series. Early intact lesions are sterile and the pathogenesis of the disorder remains unknown. PG has been reported in association with diseases due to disturbed immune mechanisms, most frequently ulcerative colitis, 3 regional enteritis, 4 rheumatoid arthritis, 5 leukaemia 6 and myeloma. 7 However, many patients have no associated disease. Characteristically resistant to treatment, the ulcers respond only to systemic corticosteroids 8 or a combination of steroids and clofazime 9,10.

Vesicovaginal fistula is a disabling gynaecological condition which may be caused by obstetric or gynaecological trauma or may be a complication of advanced local malignancy or the treatment thereof. We report a case of PG associated with endometriosis and resulting in a vesicovaginal fistula.

Case report

A 47-year-old multiparous woman was referred from another centre, with true urinary incontinence and extensive skin ulceration.

In November 1974 she had developed lower abdominal pain exacerbated by menstruation. A cystic adnexal mass was detected and she underwent a total abdominal hysterectomy and left salpingo-oophorectomy; an endometroid cyst of the left ovary was found and endometriosis was confirmed on histological examination. Ethynodiol and mestranol (Metrulen; Searle) was prescribed postoperatively and she remained well for the next 6 months.

In April 1976 she developed true incontinence of urine. This was found to be due to a large vesicovaginal fistula, the edges of which showed histological evidence of endometriosis in one segment. The remaining ovary and tube were then removed and the fistula was repaired.

During July 1976 she developed scattered painful skin ulcers over the back and anterior abdominal wall. Repeated histological examinations of the ulcer edges revealed only nonspecific granulation tissue. Two months later the vesicovaginal fistula recurred but this time no evidence of endometriosis could be found on macroscopic examination or in repeated biopsy specimens. Repair was again achieved and continence was maintained for over a year, during which time the skin ulcers continued to spread.

In February 1978 when the fistula broke down for the third time she was referred to Groote Schuur Hospital. On admission she looked ill but was afebrile. Large, necrotic ulcers clinically characteristic of pyoderma gangrenosum (PG) were present over the abdomen, buttocks, legs and one ear (Fig. 1). Histological examination of the edge of one of the ulcers showed histological evidence of endometriosis in one segment. The remaining ovary and tube were then removed and the fistula was repaired.

In April 1978 the ulceric lesions continued to spread, so that by July she was totally incontinent, even with catheter drainage under negative pressure. At this point the extent of the damage to the genitalia was assessed by examination under general anaesthesia. The ulceration and granulation tissue had healed leaving widespread tissue destruction — the labia minora were reduced to a few strips of tissue, and the large vesicovaginal fistula had completely destroyed the anterior vaginal wall, the urethral sphincter and the base of the bladder (Fig. 2).
As regards surgical management, it was felt that primary closure of the fistula was not feasible owing to atrophy of the surrounding tissues and the poor local blood supply. Urinary diversion with an ileal conduit or by means of ureterosigmoidostomy was considered; however these procedures are irreversible and liable to many complications, particularly repeated attacks of ascending urinary infection. After it was ascertained that the patient had no further interest in sexual activity, it was decided to perform the lesser procedure of colpocleisis whereby the vaginal cavity is obliterated. She made a rapid postoperative recovery and on discharge from hospital 3 months after admission could retain a fair amount of urine, although she did not regain complete continence. The dose of methylprednisolone had by now been reduced to 12 mg/d, and was tapered off gradually over the next few months.

**Discussion**

Endometriosis is defined as the presence of ectopic endometrial tissue possessing the histological and physiological features of true endometrium. It is essentially a benign lesion, of which there are two types: internal endometriosis or adenomyosis which involves the myometrium, and external endometriosis involving tissues outside the uterus.\(^1\) The sites of endometriosis were investigated in 2686 patients by Masson;\(^2\) the majority of lesions were within the uterus (1858 patients); only 62 were in the bladder and 1 in the vesicovaginal septum. The ectopic endometrium responds to cyclical hormonal variations in exactly the same way as normal endometrium, with resultant menstrual bleeding and usually dense adhesion formation. Tissue destruction is extremely rare.

Vesicovaginal fistulas may be caused by malignant disease, radiation or obstetric or gynaecological trauma, including pressure necrosis from foreign bodies. A rare congenital form has been described. Very rarely, chronic granulomatous lesions such as schistosomiasis and lymphogranuloma venereum may be responsible.\(^3\) Endometriosis has not previously been described as a cause of vesicovaginal fistula. It is not a destructive disease and in fact could have been expected to regress in our patient rather than to cause further damage after bilateral oophorectomy. Furthermore, no histological evidence of residual endometriosis was found in the fistula edges at the time of the recurrences, although ectopic endometrial tissue was found in the original biopsy specimen.

The handling of an advanced fistula of this kind is a difficult matter. Even in post-radiation fistulas the urethral sphincter is intact - in this case it was completely destroyed. The traditional colpocleisis, as described by Chassar Moir,\(^4\) in which the vagina is obliterated at the level of the fistula, could not be performed because the urethra was absent. A tubular conduit was therefore fashioned from the vaginal wall to act as a urethra, and the perineal muscles were approximated around this narrow opening to form a sphincter. The rest of the vagina was obliterated. This procedure resulted in a fair degree of urinary control which was quite acceptable to the patient.

The problem presented by our patient relates to the course of events in her illness. We know that she initially presented with endometriosis (confirmed on histological examination) of the vesicovaginal fistula wall. She subsequently developed PG-like lesions in the vagina which spread onto the vulva. These ulcers are analogous to the lesions described in cases of oral PG complicating ulcerative colitis.\(^5\) These vulval lesions, as well as the skin ulcers, responded (as does PG) to adequate doses of

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**Fig. 1. Ulcers on the abdomen showing advancing edges with healing in the centre.**

**Fig. 2. Remains of the anterior vaginal wall.**
corticosteroids and clofazimine. It is likely that PG caused the fistula, but the role which endometriosis played in the pathogenesis of PG and the vesicovaginal fistula in our patient must remain speculative.

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REFERENCES


Nuus en Kommentaar/News and Comment

Bedoarmiddels vir vroulike pasiënte

Dit is alles goed en wel om die gebruik van bedoarmiddels deur die bevolking af te keur of die algemene praktissyns wat dit voorskryf te veroordeel, maar enigemand wat 'n algemene praktissyn was, sal weet dat dit baie maklik is om 'n pasiënt se probleem met 'n middel op te los as om sy moeilike en onvriendelike omgewing te verander. Daar word algemeen aanvaar dat meer vroulike as manlike pasiënte bedoarmiddels gebruik en twee onlangse studies uit Kanada bevestig dit. Bass en Baskerville (Canad. med. Ass. J., 1981, 125, 1225) het die verslagkaarte ontleed van pasiënte tussen die ouderdomme van 25 en 64 jaar wat by 'n departement van gesitgesondheid aangemeld het om verskillende behandeling wat voorskrif vir 'n ligte bedaarmiddel ontvang. Hierdie voorskrifte was gelyk aanvra of aanmoedig. Hierdie is egter slegs gissinge en ons verstaan nog nie die wat die ware beweegredes is van dokters wat hierdie pille meer geredelik aan vroulike pasiënte voorskryf nie.

Bacteriuria and mortality in women

About 3 - 4% of women have bacteriuria, but nobody knows whether this common condition has any adverse consequences for the general population. There had been suggestions from a couple of investigations that bacteriuria is associated with an excess mortality in women, and the latest report on the subject confirms this view. Evans et al. (Lancet, 1982, 1, 156) cite a number of surveys for bacteriuria which had been performed either in South Wales or in Jamaica. In South Wales there were two separate series of three surveys each, one series in the Rhondda Valley and the others in the Vale of Glamorgan, while three surveys had been conducted in Jamaica. The intervals between the first and the third survey ranged from 1958 to 1971, 1960 to 1971, and 1959 to 1969. The present investigators measured mortality over the entire study period from survey 1 to survey 3 in women classified as bacteriuric or not, according to results of the first survey. They found that 26 out of 94 women with bacteriuria had died between the first and third surveys, compared with 147 out of 1444 non-bacteriuric women, giving a crude risk ratio of 2,5 for the bacteriuric women. Further comparisons of the different surveys confirmed this increase in mortality associated with bacteriuria. It is of course possible that bacteriuria may be more common in persons with debilitating illness than in the general population, which would account for an increase in mortality. Alternatively, some of the women might have acquired their bacteriuria in hospital to which they had been admitted for some other illness. However, the authors believe that these data suggest a positive association between bacteriuria and mortality in women after adjustment for age and weight, and that biological mechanisms may explain the association. It looks as if a large-scale therapeutic trial to clarify the situation is warranted.