AFRICAN JOURNAL OF MEDICINE and medical sciences

VOLUME 31, NUMBER 3, SEPTEMBER 2002

EDITOR:
B. O. OSOTIMEHIN

ASSISTANT EDITOR:
A. O. UWAIFO

ISSN 1116 — 4077

Afr. J. Med. med. Sci. (2002) 31, 281-282

Isolated umbilical endometriosis – a rare finding

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Summary

A 32 yr old multiparous Nigerian woman presented with histological confirmed umbilical endometriosis. There was no evidence of endometrotic deposits elsewhere. Apart from cyclical bleeding from the umbilicus, she was relatively free of any of the other signs and synptoms of Endometriosis. She had excision of the endometeriotic lesion and responded favourably to treatment. A high index of suspicion is required in making a diagnosis of endometriosis in remote extra pelvic sites especially with little or no characterisitic pointers to the disease.

Keywords: *Endometriosis, cutaneous, umbilicus, excision biopsy*

Rèsumè

Une femme Nigerianne age de 32 ans qui a en auparavent plus d'un enfant an cours de l'acconchement s'ert presentee avec l'endometriose ombilicale histologiquement confirmee. Il n'yavait pas d'antre evidence de depots endometriotique ailleurs. En dehors de la perte de sang cyclique an nirean du nombric, ele etait relativement libre de tont outré signe et symptome de l'endometriose. Elle a en une excision de la lesion endometriolique et repondait favourablement an traitement. Un index eleve de suspicion est exige lors du diagnostic de l'endometriose des sites isoles extra pelvique specialement avec pen on sans index characteristique de la maladie.

Introduction

One of the most interesting lesions encountered in gynaecological practice is endometriosis, a clinical and pathological entity described by Von-Recklinghausen in 1860 and publicized primarily by the classic contributions of Sampson in 1921. Since then, the pathogenesis of endometriosis has been a subject of debate. Although endometriosis is usually confined to the pelvis in the region of the ovaries cul de sac, the uterosacral ligaments and the utero-vescical peritoneum resulting almost always as a rule in intractable infertility and chronic pelvic pain, occasionally remote sites such as the lungs, abdominal wall, gastro-intestinal tract and nostrils may be rarely affected coexisting with pelvic endometriosis. This communication reports a rare case of isolated umbilical endometriosis in a 32-year-old Para 3+0 Nigerian woman without any associated pelvic endometriotic lesion and also presenting without any of the cardinal symptoms of endometriosis.

Case report

A 32-year-old para 3+0 petty trader was referred to the gynaecological out patient clinic of the University College Hospital, Ibadan on the 1st of March 2001, with a 2-year history of painless cyclical bleeding from the umbilicus, which coincided with her menstrual periods. She attained menarche at 16 years and menstruates for four days in regular 28-day cycles. There was no history of dyspareunia or infertility. She had three previous deliveries and they were all spontaneous vaginal deliveries and hospital supervised. Her past medical history was uneventful and she had never undergone any surgery.

On examination, she was a healthy looking young lady, not pale or jaundiced and she was moderately built. She was 1.67 meters tall and she weighed 56kg. The chest was clinically clear and cardiovascular system was intact. The abdomen was full and not tender. There were no palpable masses. The umbilicus was everted and dark brown in colour. It appeared split down the middle longitudinally by a crevice that was smeared with altered blood (figure 1). It felt indurated but was not tender.

Pelvic examination revealed a normal female lower genital tract. The uterus was of normal size, anteverted and was freely mobile. The adnexea and pouch of Douglas were normal. There was no area of induration and there were no palpable masses. A full blood count and urine microscopy, culture and sensitivity were within normal limits. Pelvic ultrasound revealed no abnormality. A double puncture laparoscopy showed a clean pelvis and there were no endometriotic deposits. Hysterosalpingography revealed no utero-cutaneous communication. On the 13th of April 2001, she had excision biopsy of the umbilical lesion under general anaesthesia. Histology of the specimen revealed the presence of ectopic endometrial glands associated with endometrial stroma consistent with endometriosis. (fig. 2)

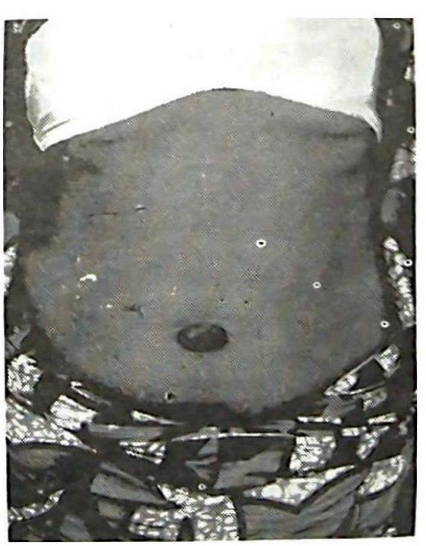

Fig. 1 Enlarged everted umbilicus of the patient. Note the longitudinal slit in the middle. The surrounding scarification marks indicate chronicity of the condition

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Fig. 2: Umbilical endometriosis (X 40 magnification) showing tubular and cystically dilated endometrial glands with some glands demonstrating intra luminal haemorrhage.

Discussion

Although endometriosis is thought to be most common amongst white women of higher socio-economic status who postpone childbearing, recent laparoscopic evidence has dispelled the notion that endometriosis is rare in other races. About 10% of Japanese women hospitalised for gynaecological symptoms had endometriosis confirmed by laparoscopy for pelvic pain [1]. Unlike in Caucasians however, experience so far among the blacks shows clearly that the condition is commoner among women of low socio-economic group [2]..

It is also interesting to note that there is a geographical difference in sites between the north and southern parts of Nigeria with the majority of endometriosis in the northern savannah region in Nigeria rarely exhibiting cutaneous and intestinal types found in southern Nigeria [3]. Villar'is credited with the first description of umbilical endometriosis in 1886. Umbilical endometriosis provides a test which theories of retrograde menstruation, coelomic metaplasis, or lymphatic/ haematogenous dissemination have difficulty reconciling.

As was the case in this patient, a dark brown/bluish nodule with history of bloody or brown discharge during menstrual flow is considered to be pathognomonic for cutaneous endometriosis. However, this classic presentation is believed to be the exception rather than the rule. Bleeding is observed in only 15% of the lesions and cyclic symptoms associated with menstruation is 52% of cases [4]. It is unusual for umbilical endometriosis to occur without any associated intrapelvic endometriotic lesion [5]. This rarity is exhibited in this patient. Thus, a negative history and review of systems for abdominal endometriosis does not eliminate the possibility of cutaneous endometriosis from the differential diagnosis. Pain is the most frequently reported symptom, but is also a clinical finding of many diseases in the differential diagnosis of an umbilical mass such as granulomas, melanoma, dermatofibroma, angiomas and epithelial inclusion cysts.

Both surgical and medical treatments have been used for cutaneous endometriosis. Excision biopsy with clinically guided margins that was performed in this patient offers a recurrence rate of 11%, most recurrences being evident within 1 year [4]. Chatterjee attempted medical management for cutaneous endometriosis with daily oral norethisterone resulting in no clinical improvement [6]. Studies comparing the concentration of hormone receptors in ectopic and intrauterine endometrium note a lower concentration of hormone receptors in ectopic endometrium, which may explain the poor response rates to pharmacological therapy [7]. Also, Mostoufizadeh and Scully [8] noted that excess endogenous or exogenous oestrogens may have a role to play in the development of malignancy in endometriosis and stressed caution in the decision to use oestrogens. Given the ineffective results and potential risks of medical management, combined with the frequent need for an excision biopsy to aid in the diagnosis, surgical intervention appears to be the treatment of choice for cutaneous endometriosis.

There have been two reported cases of malignancy arising in umbilical endometriosis. Brooks and Wheeler [9] noted a site-dependent relative risk of malignancy with ovarian endometriosis displaying a greater malignant propensity compared with extragonadal endometriosis. With two reported malignancies out of a total of about 200 reported cases of umbilical endometriosis, the potential for malignant transformation appears low.

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