Chirurgia (2014) 109: 546-549
No. 4, July - August
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Primary Umbilical Endometriosis (Villar's nodule) – Case Study, Literature Revision

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Rezumat

Endometrioza ombilicală primară (nodulul Villar) – prezentare de caz, revizia literaturii

Endometrioza ombilicală spontană cunoscută și ca nodulul Villar, este o localizare neobișnuită a țesutului endometrial. Dacă în cazul localizărilor ombilicale secundare intervențiilor chirurgicale endometrioza nu este o raritate, localizarea ombilicală primară are o frecvență de sub 1% din totalitatea localizărilor acestei afecțiuni. Autorii prezintă un astfel de caz diagnosticat la o pacientă tânără, nulipară, fără intervenții chirurgicale în antecedente, la care diagnosticul este sugerat de prezența unei formațiuni tumorale ombilicale a cărei simptomatologie se suprapune cu perioadele menstruale. Sunt discutate aspectele privind diagnosticul diferențial și opțiunile terapeutice privind rezolvarea chirurgicală a acestui tip de leziune.

Cuvinte cheie: endometrioza spontană, hernie ombilicală, nodulul Villar

Abstract

Spontaneous umbilical endometriosis, also known as Villar's nodule, is an unusual location of the endometrial tissue. Whilst in the case of umbilical locations secondary to surgical procedures endometriosis occurs quite frequently, the frequency of the primary umbilical location is lower than 1% of all endometriosis locations. The authors present such a case diagnosed in a young, nulliparous patient with no history of surgical procedures, the diagnosis being suggested by the presence of an umbilical tumor mass whose symptomatology concurs with menstrual periods. The aspects regarding differential diagnosis and the therapeutic options for the surgical cure of this type of lesion are discussed.

Key words: endometriosis, umbilical hernia, Villar's nodule

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Introduction

Defined as the location of endometrial (glandular or stromal) tissue outside the uterus, endometriosis occurs at a rate of 2-22% in asymptomatic cases, depending on the diagnosis methods used, in 20-30% of infertility cases, and in 40-60% of dysmenorrhoea cases. Whilst the common locations include the ovaries, the uterosacral ligaments, the pouch of Douglas and the other pelvic organs, extraperitoneal locations (cervix, vagina, vulva, lung, navel or postoperative scars) are exceptions. Whilst umbilical endometriosis occurring following surgical interventions is not unusual, primary umbilical location has a frequency of 0.5-1% of all endometriosis locations. (1,2,3). The clinical manifestations and the presence of umbilical tumor masses associated with the low

frequency of this disorder, may determine difficulties or errors in the differential diagnosis of any umbilical tumor mass.

We shall present below the clinical manifestations, radiologic aspects, diagnosis algorithm and surgical treatment of a primary umbilical endometriosis case.

Case report

Female patient, aged 26, with menarche at the age of 13, nulliparous, has had, for approximately 1 year, umbilical pain, sanguinolent discharge located in the umbilical area, preceded by the occurrence of a tumor mass, unpainful upon palpation outside the menstrual periods. Regular menses accompanied by dysmenorrhoea in the last 6 months. Please note that the patient has never undergone surgical procedures.

The clinical exam reveals a nodule located deep in the umbilical scar of approximately 1 cm, discharging sanguinolent fluid, painful upon palpation (Fig. 1).

The computed tomography (CT) in the abdominal-pelvic area, carried out with contrast agent, reveals in the right adnexal projection area a cystic mass 4.6/3.6 cm suggestive for an endometrioma and a small umbilical tissular nodule of 1/1.5 cm, not exceeding the muscle plane (Fig. 2).

As the patient refuses a laparoscopic intervention meant to diagnose and treat the pelvic endometriosis foci, conservatory treatment of the right ovary endometrioma is decided, in order to be subsequently subjected to another gynaecologic evaluation.

Under general anaesthesia with orotracheal intubation the surgery is performed and the umbilical scar is removed, including all anatomical planes, the including the parietal peritoneum, with a tissular excess of approximately 0.5 cm as compared to the nodule, followed by parietal reconstruction with separate polypropylene threads and reconstruction of the umbilical scar, with an excellent aesthetic result.

The postoperative evolution is favorable, the patient being discharged the second day after the surgery. The histopathology exam confirms the umbilical endometriosis (Figs. 3, 4, 5). Re-evaluated after 6 months and one year after the surgery, the patient did no longer have pain or umbilical discharges, dysmenorrhoea greatly improved under hormonal treatment (Fig. 6).



Figure 1. *Umbilical nodule discharging sanguinolent fluid, concurrently with menstrual periods*

Discussions

Extraperitoneal endometriosis is an extremely rare location of this disorder (under 1% of total sites) but which should be considered in women during fertile period and having cutaneous tumor masses which determine painful symptoms during menstrual periods.

Described for the first time in the medical literature in 1886 by Villar, spontaneous umbilical endometriosis is an extremely rare lesion whose cause is not yet very clear, and there are different theories as to the extraperitoneal locations, theories that include coelomic metaplasia, lymphatic or vascular dissemination, genetic predisposition, immunological defects or retrograde menstruation.

The high specificity clinical characteristic which should suggest this diagnosis to the surgeon is represented by the

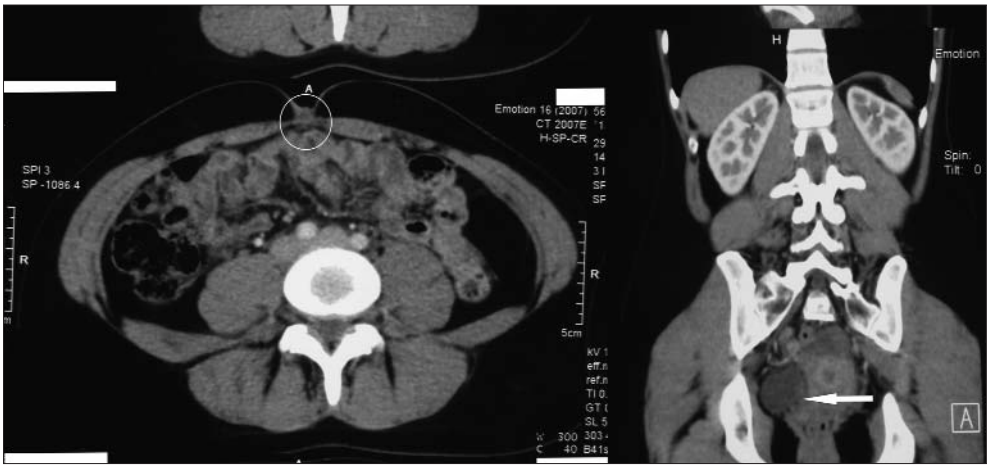


Figure 2. *The CT exam reveals a tissular umbilical nodule not exceeding the muscle plane. Right-sided adnexal endometrioma (pointed by the arrow)*

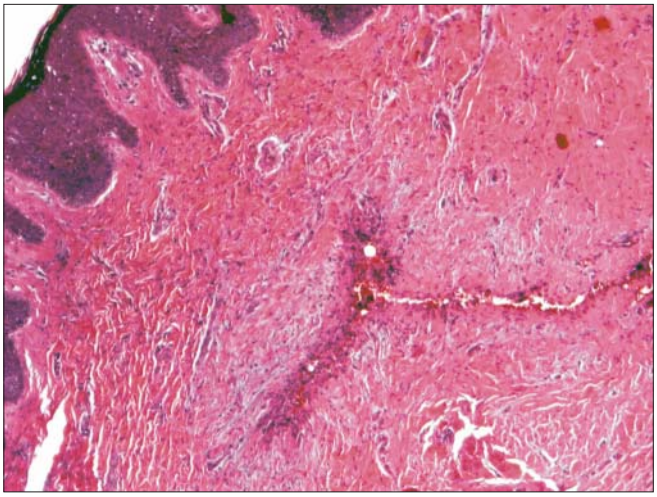


Figure 3. *Tegument (epidermis- upper left) having in the deep reticular dermis and in the hypodermis (Fig. 5) ectopic endometrium islands (endometriosis); haematoxylin-eosin staining, increase x 40*

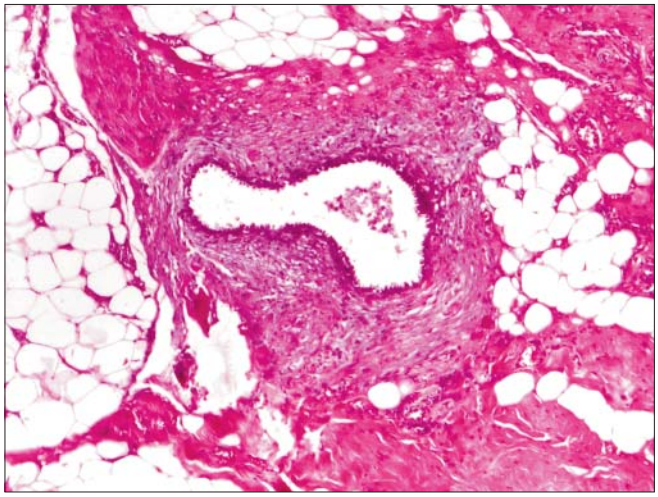


Figure 4. *Tegument having in the hypodermis a gland surrounded by cytogenic stroma or ectopic endometrium (endometriosis); haematoxylin-eosin staining, increase x 40*

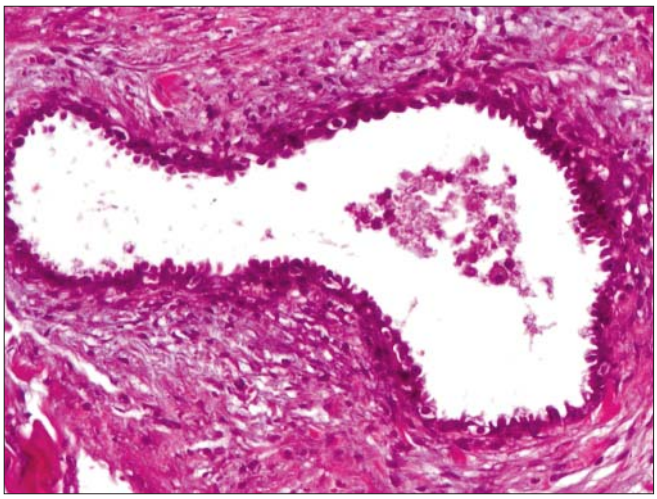


Figure 5. *Detail highlighting the endometrial gland (centrally) surrounded by the cytogenic stroma (endometriosis); haematoxylin-eosin staining, increase x 100*



Figure 6. *Aspect 30 days after the surgery. "Per primam" reconstruction of the umbilical scar.*

umbilical sanguinolent discharges concurrent with the menstruation period, a pathognomonic sign for this type of lesion, as in the case presented above, sign whose presence is, however, inconstant.

The presence of an umbilical tumor mass, for which the female patients refer, usually, to the general surgeon, may determine diagnosis errors, particularly if bleeding is not present, the differential diagnosis with secondary metastatic nodules (Sister Mary Joseph sign) being clarified only by histopathological exam (5,6). Furthermore, spontaneous umbilical endometriosis can be mistaken for or associated to umbilical hernia (7,8); there are, in the literature, examples of cases regarding the association of the two types of lesions, the assignment of the symptoms exclusively to the hernial lesion having as consequence the incomplete removal of the tumor, with the reoccurrence of the painful manifestations after the surgery. (7,9).

Imaging exploration, particularly MRI or CT exam are crucial in the preoperative evaluation of the tumor volume, of its intraabdominal extension and in the identification of other related intraperitoneal lesions, suggestive for endometriosis (3,6,7,8).

Conclusions

Primary umbilical endometriosis (Villar's nodule) should be

considered in the differential diagnosis of umbilical tumors; the large excision of the lesion including all sections of the abdominal wall with the concurrent reconstruction of the umbilical scar has good long term results.

References

1. Falcone T, Lebovic DI. Clinical management of endometriosis. Obstet Gynecol. 2011;118(3):691-705.
2. Leyland N, Casper R, Laberge P, Singh SS; SOGC. Endometriosis: diagnosis and management. J Obstet Gynaecol Can. 2010;32(7 Suppl 2):S1-32.
3. Mounsey AL1, Wilgus A, Slawson DC. Diagnosis and management of endometriosis. Am Fam Physician. 2006;74(4):594-600.
4. Vessey MP, Villard-Mackintosh L, Painter R. Epidemiology of endometriosis in women attending family planning clinics. BMJ. 1993;306(6871):182-4.
5. Loverro G, Mei L, Vicino M, Cormio G, Selvaggi L. Umbilical endometriosis. J Gynecol Surgery. 2001;17(2):65-68.
6. Weng CS, Yang YC. Images in clinical medicine. Villar's nodule - umbilical endometriosis. N Engl J Med. 2011;364(21):e45.
7. Yuen JS, Chow PK, Koong HN, Ho JM, Girija R. Unusual sites (thorax and umbilical hernial sac) of endometriosis. J R Coll Surg Edinb. 2001;46(5):313-5.
8. Farhi D, Zimmermann U, Chapron C, Dupin N. Umbilical endometriosis. J Eur Acad Dermatol Venereol. 2007;21(2):280-1.
9. Carter S, Davidson T, Hussain N, McLaughlin J. A case of spontaneous umbilical endometriosis. J Obstet Gynaecol. 1998; 18(5):490-1.