Endometrial stromal nodule of the vaginal wall with a review of vulvovaginal endometrial stromal neoplasms

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Introduction

Endometrial stromal tumors (ESTs) are usually found in both uterus and ovary. Histologically, they can exhibit a wide range of differentiations (Chew and Oliva, 2010), and their malignant potential is often defined by their mitotic activity and presence of invasive borders and eventual lymphovascular invasion. Low grade sarcomas represent the more frequent types, while benign endometrial stromal nodules (ESNs), which are non-invasive, are less common (Chew and Oliva, 2010).

We have done a systematic review on PubMed with the key words “extraterine endometrial stromal tumor”; 77 results were found and 15 were selected (4 of them are included in our references), we also read 11 articles regarding other extrauterine locations. Indeed extraterine locations are also rare but not exceptional (Chang et al., 1993) with most cases originating in endometriotic foci. Thus, primary cases of them being metastases from other sites. Bibliographical analysis demonstrated various types of endometrial stromal sarcoma, often associated to endometriosis, but no cases of stromal nodules like the one we are submitting.

This paper reports, for the first time, the occurrence of an asymptomatic, non-recurring, polyloid primary endometrial stromal nodule (ESN) in the vagina of a 47-year-old female that was not associated with endometriosis and that was successfully treated by local resection.

We also review the available cases of vulvovaginal ESTs.

Case report

A 47-year-old nulliparous patient, with an otherwise unexceptional gynecological history, consulted for a sensation of foreign body in the vagina. On clinical examination the vulva was unremarkable and a polypoid, non-recurring, pedunculated mass of approximately 2 cm on the posterior aspect of the introitus was found. The rest of the female genital tract was unexceptional. The nodule was completely resected. No other lesions were detected in either vagina or vulva. Hysteroscopy and abdomino-pelvic MRI performed after resection were normal. Ca125 serum levels were within normal range. The patient has been followed regularly for a period of six years without recurrence.

Pathology

On gross examination, the 2 cm round, white yellow mass was homogeneous and elastic. Microscopically the vaginal epithelium lined the external circumference of the nodule which enclosed a homogeneous proliferation of endometrial stromal-type cells that had clear-cut fine borders. No invasion of the pedicle or any surrounding vessels (Fig. 1A) was seen. Cells without atypia and lacking mitoses grew in diffuse sheets and had abundant collagen tracts and extensive hyalinization of perivascular distribution (Fig. 1B). Minor foci of calcification and foamy macrophages were present. Immunohistochemistry confirmed the endometrial stromal nature of the tumor by its

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conspicuous coexpression of CD10 and estrogen and progesterone receptors (Fig. 2A–B). Smooth muscle markers such as h-caldesmon and desmin were negative.

Discussion

We report a case of an endometrial stromal nodule in the vagina in a premenopausal patient. Our case is unique, since no instances of this variant of EST, a benign endometrial stromal lesion, have been previously reported outside the uterus. Histologically, ESNs have lineal, smooth, pushing margins, minimal atypia or mitoses and often present a yellow color; possibly due to the presence of foamy macrophages. Characteristically they are non-invasive and do not permeate the capsule or adjacent vessels. The present case exemplifies such a lesion in a highly unusual location. Its benign nature is confirmed by the absence of local recurrence after a long follow-up. However, it must be borne in mind that some ESSs may experience very late recurrences (Chew and Oliva, 2010).

In contrast, all cases of ESNs reported in the vulvovaginal region (Table 1) have corresponded to endometrial stromal sarcomas of various grades of differentiation. In at least five cases (Androulaki et al., 2007; Berkowitz et al., 1978; Corpa et al., 2004; Kondi-Paphitis et al., 1998; Liu et al., 2013; Masand et al., 2013) the tumors appeared to be primary neoplasms, while the remaining ones were likely to be metastatic in nature (Irvin et al., 1998; Kondi-Paphitis et al., 1998; Liu et al., 2013; Masand et al., 2013), since tumor was also found in pelvic locations, colon and lung. Association with endometriosis, locally or elsewhere, was found in 6 cases.

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In the present case, a metastasis from any other site was discarded since there was no evidence of either utero–ovarian primaries. Furthermore, an origin from endometriosis is unlikely since no such areas were detected in the vicinity of the neoplasm or elsewhere. This would support the possibility of a locally originated endometrial stromal neoplasm that could represent a type of Müllerian differentiation in the vagina, an organ whose partly Müllerian origin is well known (Sanchez-Ferrer et al., 2006).

Histopathological diagnosis was relevant to the management of this case, with characterization of its endometrial stromal cellularity based on both cell morphology and coexpression of characteristic markers such as CD10, estrogen and progesterone receptors. In the differential diagnosis, the immunohistochemical absence of smooth muscle markers excluded the more commonly found leiomyoma of the vagina (Imai et al., 2008), a tumor, especially their cellular variants, that may resemble ESN. An accurate diagnosis of ESN is important since local conservative surgery is curative. Core biopsy is not indicated since the diagnostic features of a pushing, non-invasive margin cannot be diagnosed with this procedure.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the
written consent is available for review by the Editor-in-Chief of this journal on request.

Conflict of interest

The authors declare that they have no conflict of interest.

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Table 1

Clinicopathological features of reported cases of endometrial stromal sarcoma of the vulvovaginal region.

| Nr  | Site  | ESS type | Organ involvement | Associated endometriosis |
|-----|-------|----------|-------------------|--------------------------|
| 1   | Vagina| Sarcoma NOS | —                 | +                        |
| 1   | Vagina| Sarcoma NOS | —                 | +                        |
| 1   | Vagina| LGESS    | —                 | —                        |
| 1   | Vagina| Sarcoma NOS | Pelvic            | +                        |
| 1   | Vagina| Sarcoma NOS | Pelvic            | —                        |
| 1   | Vagina| Sarcoma NOS | Metastases        | NA                       |
| 1   | Vagina| Sarcoma NOS | Colon             | +                        |
| 1   | Vagina| Sarcoma NOS | Pelvic            | +                        |
| 1   | Vagina| Sarcoma NOS | Lung              | +                        |
| 1   | Vagina| LGESS    | Late metastases   | NA                       |

Key: LGESS: low grade endometrial stromal sarcoma. NA: not available.