Rev Esp Patol 1997; Vol. 30, Nº 4: 317-320

Caso aislado

Low-grade endometrial stromal sarcoma with focal sex-cord-like differentiation, and endometriosis

J.A. Aramburu-González*, R. Granados-Carreño and P. Ortega-Muñoz

Department of Pathology, Hospital Universitario de Getafe, Madrid, Spain.

RESUMEN

Presentamos un caso de sarcoma del estroma endometrial asociado a endometriosis y diferenciación de cordones sexuales, y revisamos la literatura. El tumor estaba presente en el útero, ovarios. epiplón y apéndice. Histológicamente se caracterizaba por áreas de diferenciación hacia cordones sexuales en el útero, por zonas de sobrecrecimiento estromal sin elementos glandulares, y por la presencia de endometriosis en el ovario derecho y epiplón. Histogenéticamente, valoramos la posibilidad de componente síncrono originado a partir de focos endometriósicos, frente al origen metastásico. También hemos considerado la diferenciación glandular dentro del tumor. Este tipo de presentación histológica es poco común, existiendo pocas referencias en la literatura. **Rev Esp Patol 1997; 30(4): 317-320.**

Palabras clave: Sarcoma del estroma endometrial - Endometriosis - Tumor de cordones sexuales - Diferenciación glandular

SUMMARY

A case of low-grade endometrial stromal sarcoma associated with endometriosis and focal sex-cord-like differentiation is reported here in a review of the literature. The tumor was present in the uterus, both ovaries, omentum, and appendix. The histological appearance of the tumor was that of a low-grade endometrial stromal sarcoma with epithelial differentiation in the form of sex-cord within the uterus, extensive intravascular stromal overgrowth without glandular elements, and the presence of endometriotic tissue in the right ovary (endometriotic cyst) and omentum. For the histogenesis, we favor the possibility of synchronous component arising from different foci of endometriosis versus a metastatic tumor. The case of glandular differentiation within the tumor has also been considered. The presence of sex-cord-like structures and benign-appearing glands of endometrial type, as seen in our case, is an unusual presentation with few references in the literature. **Rev Esp Patol 1997; 30(4): 317-320.**

Key words: Endometrial stromal sarcoma - Endometriosis - Sex-cord tumor - Glandular differentiation

INTRODUCTION

Endometrial stromal tumors are among the rarest primary neoplasms of the uterine corpus. Occasionally, they contain epithelial elements that resemble sex-cord (1). Foci of endometriosis or endometrial gland-like structures can coexist in an area adjacent to the tumor (2-4).

In this article, we describe the pathological and immunohistochemical findings of a unique tumor that combines sex-cord epithelial elements and several foci of endometriosis.

CASE REPORT

A 38-year-old nullipara presented with a two-year history of vaginal bleeding. Radiological studies revealed many intramyometrial masses, a large and cystic right ovary, and multiple nodules in the omentum with a soft tissue density. A hysterectomy with bilateral salpingo-ophorectomy, an appendectomy, and partial omentectomy were performed.

On gross examination, an ill-defined, firm, yellow-to-orange mass of 7 to 9 cm was observed in the uterus. Many small gray to black nodules were present in omentum. The right ovary contained a small endometriotic cyst.

Microscopic examination revealed irregular islands of tumor replacing the endometrium and invading the myometrium. They were composed of sheets of endometrial-type stromal cells and a network of small vessels with mild nuclear pleomorphism and a low mitotic rate (4m/10 HPF). The right ovary showed areas of low-grade endometrial stromal sarcoma that seemed to arise from the wall of an endometriotic cyst. Foci of endometriosis were found in areas adjacent to and within the tumor in the omentum (Fig. 1). The appendix and left ovary were also affected by tumor. Intravascular tumor composed solely of stroma was frequently observed in every affected organ. Focal differentiation into epithelial-like elements resembling an ovarian sex-cord tumor was observed within the endometrium (Fig. 2).

Immunohistochemical studies (Table 1) showed that the stromal cells were immunoreactive for vimentin, and estrogen and progesterone receptors. They were also focally positive for a keratin cocktail (EA1-EA3), and Ki-67 (less than 2%). The epithelial-like elements were only weakly positive for vimentin.

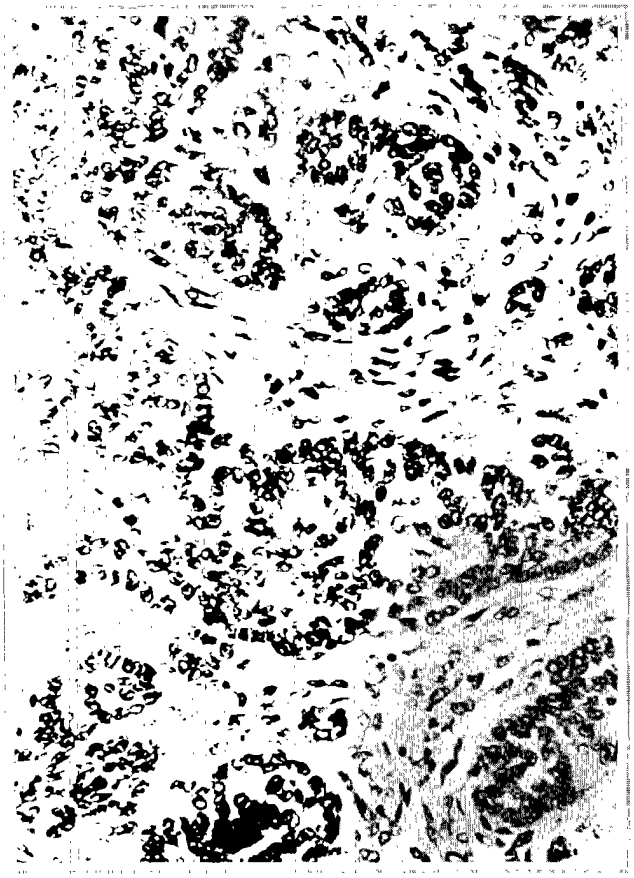

Figure 1. Foci of endometriosis in an area adjacent and within the tumor in omentum (original, HE ×10).

DISCUSSION

The presence of focal sex-cord-like differentiation within otherwise typical endometrial stromal tumors has been reported before (1, 5). Their frequency has varied from 1% (6) to almost 60% in different series (7). Recent immunohistochemical studies have found that sex-cord-like foci are frequently immunoreactive for vimentin, desmin, and muscle-specific actin, suggesting that they may represent foci of myoid, rather than epithelial differentiation (8). Other authors have found immunoreactivity for cytokeratines more commonly than for desmin (3). In our case, we only observed immunoreactivity for vimentin. Sex-cord differentiation is generally found in tumors from patients in the reproductive age (1), like the patient presented here. This finding does not seem to have an appreciable effect on tumor behavior (1, 7), even though some authors have observed highly significant differences in recurrence of disease, but not in mortality (9).

Figure 2. Endometrium: Focal differentiation into epithelial-like elements, resembling ovarian sex-cord tumor (original, HE ×175).

The case presented here brings up the controversial histogenesis of the endometriotic foci. A variety of malignancies arising from endometriosis have been reported (10-13). The occurrence of endometrial stromal tumors from foci of endometriosis has rarely been reported (12, 13), perhaps because some authors refer to this issue as a concurrent event of both entities (4). The case presented here involved an endometriotic cyst in the right ovary and many small nodules of endometriotic tissue adjacent and within the tumor in the omentum. The stromal overgrowth arising from the wall of the endometriotic cyst, and the close relationship between the components of the tumor and the endometriotic tissue in the omentum, suggests the possibility that the tumor originate from foci of the endometriosis in this case. However, metastatic origin from uterus is also possible.

Finally, some authors (3, 13, 14) have described low-grade endometrial stromal sarcomas with glandular differentiation. The presence of these glands separated by bland endometrial-type stroma may be, in areas, indistinguishable from endometriosis. Although such glands have been documented in 5% to 40% of the cases by several authors (3-5, 12, 14), this change is uncommon and has not been specifically noted in most published series of these tumors. Their occurrence has been variably attributed to focal epithelial differentiation (epitheliogenesis) or to the entrapment of non-neoplastic endometrial glands, as would occur in a low-grade endometrial stromal sarcoma supposedly arising from endometriosis. The occasional presence of those glands within intravascular tumor (3) supports the theory that the glandular component is part of the neoplasms. However, the histological appearance of these areas may be indistinguishable from that of a low-grade endometrial stromal sarcoma arising from endometriotic foci elsewhere. In our case, we interpret the finding of endometrial glands in the ovary and omentum in a continuum with the tumor, as part of endometriosis rather than as a glandular differentiation of the tumor. The presence of an endometriotic cyst and intravascular stromal overgrowth without glandular elements also support this point of view.

Table 1. Primary antibodies used.

Antibodies	Source	Dilutions
Vimentin	Biomeda	Prediluted
EA1-EA3	Biomeda	Prediluted
EMA	Biomeda	Prediluted
HHF-35	Biomeda	1:50
S-100	Biomeda	Prediluted
Progesterone receptor	Novocastra	1:20
Estrogen receptor	Novocastra	1:30
Ki-67	Novocastra	1:10

ACKNOWLEDGEMENTS

We thank Miss Aurora Suárez for her help with the immunohistochemical techniques.

REFERENCES

1. Clement PB, Scully RE. *Uterine tumors resembling ovarian sex-cord tumor. A clinicopathological analysis of 14 cases.* Am J Clin Pathol 1976; 66: 512-525.

2. Shiraki M, Otis CN, Powell JL. *Endometrial stromal sarcoma from ovarian and extraovarian endometriosis –report of two cases and review of the literature.* Surg Pathol 1991; 4: 333-343.
3. Clement PB, Scully RE. *Endometrial stromal sarcomas of the uterus with extensive endometrioid glandular differentiation: A report of three cases that caused problems in differential diagnosis.* International Journal of Gynecol Pathol 1992; 11: 163-177.
4. Young RH, Prat J, Scully RE. *Endometrioid stromal sarcomas of the ovary. A clinicopathologic analysis of 23 cases.* Cancer 1984; 53: 1143-1155.
5. Norris HJ, Taylor HB. *A clinical and pathological study of 53 endometrial stromal tumors.* Cancer 1966; 19: 755-766.
6. Symmonds RE, Dockerty MB, Pratt JH. *Sarcoma and sarcoma-like proliferations of the endometrial stroma. III. Stromal hyperplasia and stromatosis (stromal endometriosis).* Am J Obst Gynecol 1957; 73: 1054-1070.
7. Fekete PS, Vellios F. *The clinical and histologic spectrum of endometrial stromal neoplasm: A report of 41 cases.* Int J Gynecol Pathol 1984; 3: 198-212.
8. Franquemont DW, Frierson HF, Mills SE. *An immunohistochemical study of normal endometrial stroma and endometrial stromal neoplasm.* Am J Surg Pathol 1991; 15: 861-870.
9. Chang KL, Crabtree GS, Lim-Tan SK, Kempson R, Hendrickson MR. *Primary uterine stromal neoplasm. A clinicopathologic study of 117 cases.* Am J Surg Pathol 1990; 14(5): 415-438.
10. Brooks JJ, Wheeler JE. *Malignancy arising in extragonadal endometriosis. A case report and summary of the world literature.* Cancer 1977; 40: 3065-3073.
11. Mostoufizadeh M, Scully RE. *Malignant tumors arising in endometriosis.* Clin Obstet Gynecol 1980; 23: 951-963.
12. Heaps JM, Wieberg RK, Berek JS. *Malignant neoplasm arising in endometriosis.* Obst Gynecol 1990; 75: 1023-1028.
13. Chang KL, Crabtree GS, Lim-Tan SK, Kempson RL, Hendrickson MR. *Primary extrauterine endometrial neoplasm: A clinicopathologic study of 20 cases and review of the literature.* Int J Gynecol Pathol 1993; 12: 282-296.
14. Tavassoly FA, Norris HJ. *Mesenquimal tumors of the uterus. VII. A clinicopathological study of 60 endometrial stromal nodules.* Histopathology 1981; 5: 1-10.