

Case report

Endometrial stromal tumors with sex cord-like elements: a case report

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Abstract

Endometrial stromal nodules are rare. They represent less than a quarter of endometrial stromal tumors. Clement and Scully described as variants of endometrial stromal nodules two types of tumor resembling ovarian sex cord tumors. Type I is tumor that resembles focally an ovarian sex cord tumor which can be abbreviated ESTSCL, or endometrial stromal tumors with sex cord-like elements and type II; uterine tumors with more than 40% sex cord-like differentiation so called UTROSCT for uterine tumor resembling ovarian sex cord tumor. Uterine tumours with sex cord-like differentiation are very rare with only 50 cases reported in the literature so far. The diagnostic is based on microscopic findings. Immunohistochemical tests to elucidate their nature yield variable and polyphenotypic with coexpression of markers of epithelial, myoid and sex cord lineage as well as hormone receptors. We report an additional case of an endometrial stromal tumors with sex cord-like elements in a 48-year-old woman presenting with abnormal mass.

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Introduction

Endometrial stromal nodules are rare. They represent less than a quarter of endometrial stromal tumors [1]. Exceptionally, this benign tumour can resemble, at least focally, an ovarian sex cord tumor. Endometrial stromal tumors with sex cord-like elements (type I) have an unfavorable prognosis; and the uterine tumor resembling ovarian sex cord tumor UTROSCT (type II) containing more than 40% sex cord-like differentiation and less endometrial component are biologically less aggressive.

Patient and observation

A 48-year-old multiparous woman presented with pelvic mass with abdominal pain, urinary tract symptoms, abnormal vaginal bleeding and palpable uterine mass. Ultrasound examination showed a heterogeneous uterine tumor composed of cystic and solid parts leading to abdominal hysterectomy.

Gross finding (**Figure 1**) a polypoid and protrude tumor involving in the myometrium and tend to bulge above the surrounding myometrium. The tumor is well circumscribed contour, measuring 11x10x8cm and have fleshy yellow surface (**Figure 2**). This tumor is intramural with no connection to the endometrium.

In microscopic findings the tumor consist of cells that closely resemble normal proliferative-phase endometrial stromal cells with areas of epithelial-like structures that have an appearance reminiscent of an ovarian sex cord-stromal tumor. The tumor cells have uniform, small, darkly staining round or oval nuclei with granular chromatin and inconspicuous nucleoli. Mitotic activity is less than 3MF/10HPF.

The epithelial-like cells grow in cords and trabeculae, they are cuboidal with scanty amphophilic cytoplasm and nuclei resemble those of the surrounding stromal cells (**Figure 3**). The tumor presents expansile, non-infiltrative margins that compress the surrounding myometrium (**Figure 4**). The tumor was immunoreactive to CD10 (**Figure 5**) and hormonal receptors (ER (**Figure 6**) and PR (**Figure 7**). Immunostaining for AML, desmine, calretinin, cytokeratin AE1/AE3 and inhibin was negative.

Discussion

Tumors of the uterus resembling ovarian sex cord tumors were reported by Clement and Scully in 1976 and were divided in 2 groups [2] accounting of the share of sex cord-like elements. Type I is tumours with only focal sex cord-like differentiation and type II with predominant differentiation with more than 40% of sex cord-like differentiation so called UTROSCT for uterine tumor resembling ovarian sex cord tumor.

Uterine tumours with sex cord-like differentiation are rare occurring in middle-aged women, the average age is around 50 with no racial predisposition. The main symptoms are abnormal vaginal bleeding and menorrhagia [3] that can cause anemia, and pelvic or abdominal discomfort. The average duration of symptoms before diagnosis is about 2 months. The pelvic mass in our case is due to the latest of consultation. Most patients have an enlarged uterus or a palpable uterine mass.

About 10% of tumours are found incidentally after hysterectomies performed for other conditions.

Macroscopically, the tumor is characteristically a solitary, well delineated, round fleshy nodule with a yellow to tan sectioned surface. The median tumour diameter is 4 cm (range 0.8 to 15cm). It was 11cm in our case. About two-thirds are purely intramural without any apparent connections to the endometrium. Occasional tumors are cystic, but foci of necrosis and hemorrhage are rare.

The histological appearance found endometrial stromale nodules with areas of epithelial-like structures resembles an ovarian sex cord tumours. The stromale nodules have expansile, noninfiltrative margins that compress the surrounding endometrium and myometrium. Minor irregularities of the margin are common, but invasion of the surrounding myometrium indicates that the tumor is a stromale sarcoma, not a stromale nodule.

Endometrial stromal tumors with sex cord-like elements exhibit a polyphenotypic immunophenotype. There is most often a mixed epithelial-myoid phenotype, with immunoreactivity for cytokeratin and actin, and, in some cases, desmin [4]. Immune-stains for EMA are almost always negative. In accord with the resemblance to a sex cord tumor, immunoreactivity for inhibin and CD99 is detected in epithelial-like structures in about a third of type I tumors [5]. In our case the tumor expresses the CD10 and hormonal receptors. Immunostaining for AML, desmine, calretinin, cytokeratin AE1/AE3 and inhibin was negative.

Endometrial stromal nodules with focal sex cord-like differentiation tend to relapses and metastasing. In Clement and Scully's initial report, three of five patients, even with follow-ups, had recurrences and two died [2]. Baker et al found that 15% of reported cases were known to have recurred [5].

Because of the tumours' rarity, there are no randomized studies as to an optimal therapy. Nevertheless, hysterectomy is usually the appropriate therapy because it permit to evaluate the periphery of the tumor and to be certain that it's completely circumscribed and non invasive.

Conclusion

Endometrial stromal tumors with sex cord-like elements type I according to the features described by Clement and Scully is very rare tumor with a tendency of recurrences. The pathologist had a crucial role in diagnosis, often difficult due to the heterogeneity of the tumor. Correct diagnosis should avoid overtreatment. Hysterectomy is usually the appropriate therapy to evaluate margins. Endometrial stromale tumors with a focal sex cord pattern with circumscribed margins are benign but those with infiltrating margins behave as low-grade endometrial stromale sarcomas.

Competing interests

The authors declare that they have no competing interests.

Authors' contributions

Mouhcine Bendahou made contributions to the conception and design, and acquisition of data. Hind El Fatemi, Taoufiq Harmouch and Amal Benlemlih performed the histological examination and was a major contributor in writing the manuscript. Hind El Fatemi has been involved in drafting the manuscript and revising it critically for important intellectual content. Afaf Amartu has given final approval

of the version to be published. All authors read and approved the final manuscript

Figures

Figure 1: Polypoid and protrude tumor involving in the myometrium and tend to bulge above the surrounding myometrium with no connection to the endometrium.

Figure 2: The tumor has a fleshy yellow surface.

Figure 3: The tumor consists of cells that closely resemble normal proliferative-phase endometrial stromal cells with areas of epithelial-like structures grow in cords and trabeculae. HESx20

Figure 4: The tumor presents expansive, non infiltrative margins that compress the surrounding myometrium.HESx5

Figure 5: The tumor is immunoreactive to CD10. HESx20

Figure 6: The tumor is immunoreactive to hormonal receptors (ER). HESx20

Figure 7: The tumor is immunoreactive to hormonal receptors (PR). HESx20

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Figure 1: Polypoid and protrude tumor involving in the myometrium and tend to bulge above the surrounding myometrium with no connection to the endometrium



Figure 2: The tumor has a fleshy yellow surface.

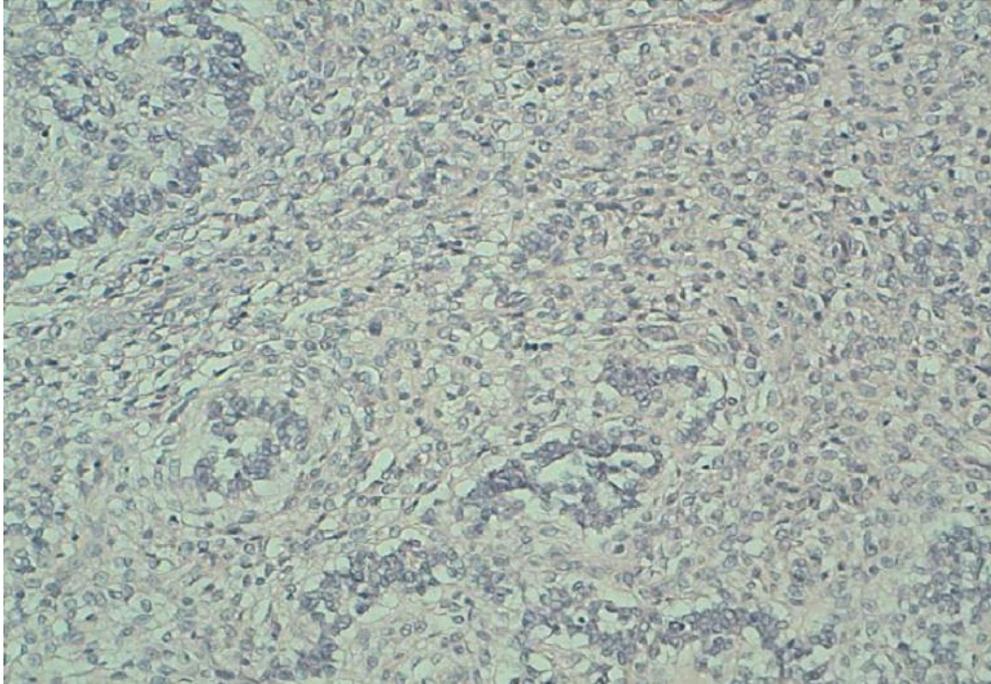


Figure 3: The tumor consist of cells that closely resemble normal proliferative-phase endometrial stromal cells with areas of epithelial-like structures grewed in cords and trabeculae. HESx20

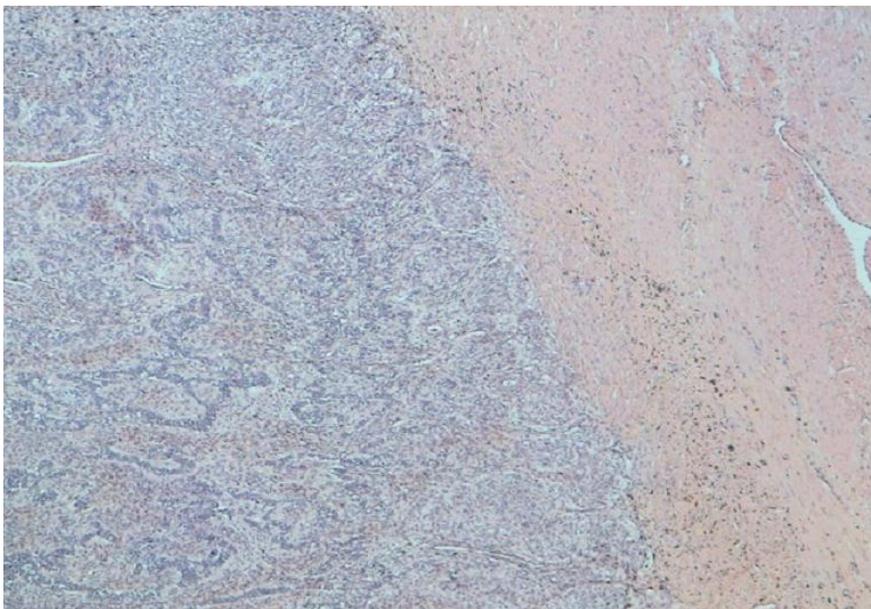


Figure 4: The tumor presents expansive, non-infiltrative margins that compress the surrounding myometrium.HESx5

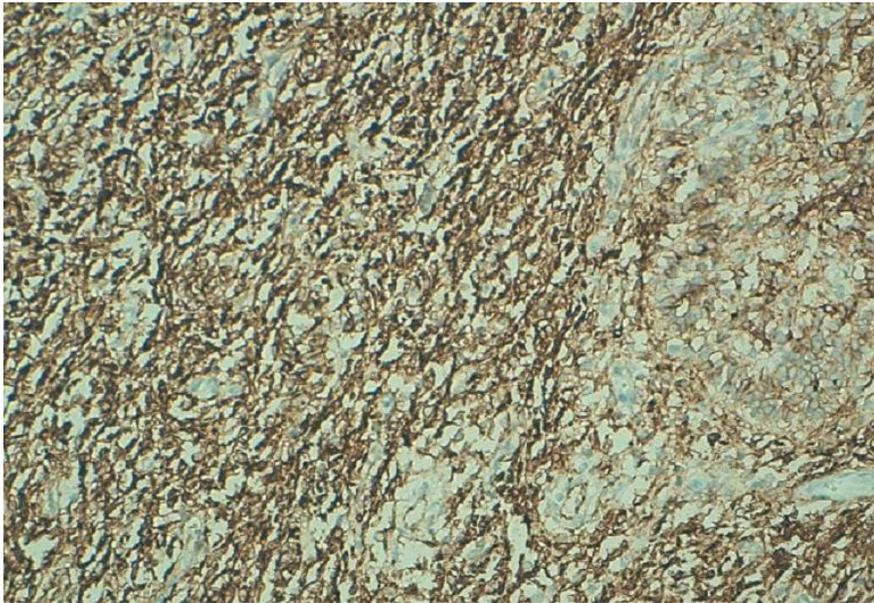


Figure 5: The tumor is immunoreactive to CD10. HESx20



Figure 6: The tumor is immunoreactive to hormonal receptors (ER). HESx20



Figure 7: The tumor is immunoreactive to hormonal receptors (PR). HESx20