Endometrial stromal sarcoma presenting as an ovarian cyst – A case report

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Abstract

Introduction: Endometrial stromal sarcomas usually arise in the uterine corpus while it is rare in extra-uterine sites. The most common extra-uterine site is the ovary. However, primary endometrial stromal sarcoma of the ovary rarely arises in a serous tumor. We report a case of primary ovarian ESS presenting in the wall of a serous cyst along with a synchronous endometrial stromal nodule (ESN).

Case report: A 50 year old woman presented with lower abdominal pain and fullness. On radiological examination, a left ovarian, predominantly cystic mass about 10cm in diameter was found along with an uterine nodule, 1.5cm in diameter, presumed to be a leiomyoma. The ovarian cyst on histology was diagnosed as ESS arising in a serous cystadenoma. The uterine nodule was diagnosed as ESN since there was no myometrial or vascular invasion.

Discussion: Endometrial stromal sarcomas pose a diagnostic challenge at extra-uterine sites. Malignant mixed mullerian tumor and sex cord stromal tumor are close differential diagnoses. The other problem is deciding whether the tumor is primary or secondary. The problem of diagnosis is compounded if the presentation is cystic.

Conclusion: We concluded that a primary ovarian ESS should be diagnosed after excluding possible metastatic spread from uterine ESS. In cases with an uterine nodule, the periphery of the lesion should be thoroughly sampled to exclude ESS. We also suggest that an ovarian cyst is not necessarily epithelial.

Key-words: endometrial stromal sarcoma, endometrial stromal nodule, cyst

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Introduction:

There are three categories of endometrial stromal tumors: endometrial stromal nodule, low-grade endometrial stromal sarcoma, high-grade endometrial stromal sarcoma. Endometrial stromal sarcoma (ESS) usually develops in the uterine corpus, whereas extrauterine sites are extremely rare.[1] Only 86 cases have been reported in the literature and more than 50% of the cases were associated with pre existing endometriosis.[2] The primary site in 76% of cases of extra-uterine endometrial stromal sarcomas is the ovary.[3] Primary ovarian sarcomas comprise only 1-3%

of all ovarian neoplasms and very rarely arise in serous ovarian tumors.[4]

The rarity of ESS in the wall of an ovarian cyst along with a synchronous endometrial stromal nodule prompted this case report.

Case Report:

A 50 year old woman presented with lower abdominal pain and a feeling of fullness of lower abdomen. Par vaginal examination revealed a pelvic mass palpable through the left vaginal fornix, about 8cm in diameter. Physical examination was otherwise normal. Ultrasound examination and C.T. scan showed a cystic left ovarian mass measuring 10x8x8cm with slight peripheral enhancement. A provisional diagnosis of epithelial cystic neoplasm of ovary was made. Malignancy could not be ruled out. The other ovary was normal in size. The uterus showed a subserous nodule measuring 1.5x1.5 cm, presumed to be a fibroid. Other laboratory data were within normal limits. The patient underwent total abdominal hysterectomy and bilateral salpingo-oophorectomy.

Gross examination:

The specimen showed an ovarian tumour measuring 10x9x8cm with smooth outer surface. On cut section the tumour was predominantly cystic. The cyst was unilocular, containing serous fluid. The solid part was yellow-tan in colour (Fig 1). There were no areas of haemorrhage or necrosis. The uterus was normal in size and showed a nodule impinging on the serosa, measuring 2x2 cm. The other ovary and omentum were grossly normal.

Histopathological examination:

The ovarian tumour in the solid part was composed of small, closely packed cells with uniform oval nuclei and coarse, evenly dispersed chromatin. The cytoplasm was scanty with indistinct cell borders. A characteristic vascular pattern with perivascular proliferation of the tumour cells was seen (Fig 2). Mitotic figures were three per ten high power fields. Cellular atypia or Pleomorphism was minimal. . The uterine nodule showed a similar histological picture but was less cellular with rare mitoses. The entire nodule was processed. No focus of myometrial or vascular invasion was seen. The ovarian cyst in other areas was lined by a single layer of flattened cuboidal epithelium, resembling a serous cystadenoma. There were no foci of endometriosis. The endometrium was in proliferative phase. Multple sections from rest of endometrium, myometrium, other ovary and omentum showed no evidence of endometrial stromal sarcoma.

Reticulin stain showed reticulin deposit around individual tumor cells in both lesions.

Immunohistochemistry:

The tumor cells in both lesions were positive for CD10 (fig3), estrogen and progesterone receptor but negative for smooth musle actin, desmin and alpha inhibin.CD10 positivity however, was more intense in the ovarian lesion.

The case was diagnosed as low grade endometrial stromal sarcoma arising in ovarian serous cystadenoma with a synchronous endometrial stromal nodule.

The patient is undergoing chemoradiation therapy and is doing well.

Discussion:

Endometrial stromal sarcomas pose a diagnostic challenge, especially in extra-uterine sites.[1] Mourad et al found that out of 86 reported cases of extra-uterine ESS, 48 were in the ovary and the rest were extra-ovarian sites including vaginal septum, fallopian tube,broad ligament and abdominal cavity.[2]

There are two hypotheses regarding the pathogenesis of extra-uterine ESS. The first one is that it arises from foci of pre-existing endometriosis and the second one is that it arises from sub-mesothelial pluripotential cells.[5]

The microscopic differential diagnosis of ovarian ESS includes Malignant mixed mullerian tumor and sex cord stromal tumor.[6] Careful morphological assessment and a panel of immunohistochemical markers as in our case is helpful.

Any ovarian sarcoma poses a diagnostic problem as it may be primary or metastatic. In our case an uterine nodule was found. It was composed of endometrial cells, was well circumscribed with no evidence of myometrial or vascular invasion and had rare mitoses. It was only two centimetre in size compared to the big ovarian mass, was located in the myometrium towards the serosal aspect and had less intense CD10 positivity compared to the ovarian lesion. We diagnosed this as an associated endometrial stromal nodule (ESN). We have not found any other report of a case describing the synchronous occurrence of ESN and ovarian ESS. Thorough sampling of the periphery of the lesion is necessary in such cases to differentiate low grade ESS and ESN. A wrong diagnosis in the present case would signify a higher stage disease with ovarian spread. In a younger patient where uterine preservation is desirable ESN poses a greater problem since it cannot be reliably diagnosed without hysterectomy.

In many cases with sarcoma found in both the uterus and ovary, it might not be possible to ascertain which one is the primary or whether there are double primaries. Prognosis is poor once the disease has metastasised.[7]

An ovarian cyst is frequently epithelial in nature. The mere possibility of a

cyst being sarcomatous is scarcely mentioned in literature. Thus, a cystic sarcoma is a difficult pre-operative diagnosis. A sarcoma may outgrow its blood supply and break down to form a cyst. On the other hand, a simple cystic tumor such as a cystadenoma or a dermoid may develop sarcomatous tissue in its wall. The sarcomatous tissue may then be called 'adenocystoma' as in our case.[8]

Ovarian primary sarcomas have a poor overall prognosis.[9] They should be carefully considered in the differential diagnosis of ovarian lesions.

Conclusion:

ESS has better prognosis than other sarcomas, especially in lower stage. Our case highlights the importance of diagnosing primary ovarian ESS without wrongly upstaging it in cases where a benign uterine ESN may be associated. The case also suggests that a predominantly cystic ovarian lesion can be a rare presenting feature of a sarcoma.

References:

1. Tahmasebi M, Cina M. Bilateral endometrial stromal sarcoma originating from ovarian endometriosis: A case report. Iran J Radiol 2009; 6: 33-6.
2. Mourad W.A, Abdelgaffar B, Tulbah A, Tulbah M, Subhi J. Multifocal extrauterine endometrial stromal sarcoma: a very rare complication of endometriosis associated with elevated serum CA-125. Annals of Saudi Medicine 2003; 23: 181-3.
3. Mouthon A, Tiret E, Parc Y, De Saint Maur P, Parc R, Flejon JF. Endometrial stromal sarcoma of the rectosigmoid colon arising in extragonadal endometriosis and revealed by portal vein thrombosis. Arch Pathol Lab Med 2001; 125: 1088-90.
4. Clarke TJ. Sarcoma like mural nodules in cystic serous ovarian tumors. J Clin Pathol 1987;40: 1443-8.
5. Zaloudek C, Hendrickson MR, Soslow RA. Mesenchymal tumors of the uterus. In: Kurman RJ, Ellenson LH, Ronnett BM, editors. Blausteins pathology of the female genital tract. 5th ed. New York Springer- Verlay Inc 2002;586-93.
6. Joo Yeon Kim, Seong Yeon Hong, Hyun Jung Sung, Hoon Kyu, Suk Bong Koh. Acase of multiple metastatic low grade endometrial stromal sarcoma arising from an ovarian endometriotic lesion. J Gynecol Oncol 2009;20:122-5.
7. Tak Hong Cheung. Metastatic sarcoma of the ovaries. CME Journal of Gynecologic oncology 2004;9:153-6.
8. Frank E Taylor. On adenocystoma ovari sarcomatodes. BJOG: An international journal of obstetrics and gynaecology 2005; 9: 268-85.

9. Dai Y, Shen K, Lang H, Huang HF, Pan LY, Wu M et al. Primary sarcoma of the ovary: clinicopathological characteristics, prognostic factors and evaluation of therapy. Chin Med J (Engl) 2011; 124: 1316-21.

Figure 1: Gross Examination – shows an ovarian cyst and a small uterine p[illegible]ith a characteristic tan yellow colour.

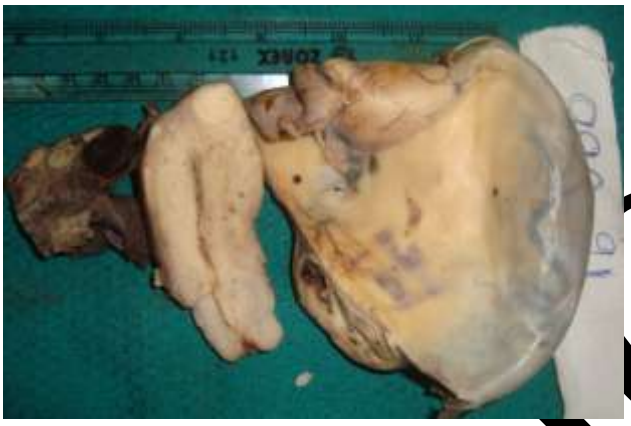

Figure 2: Histology shows th[illegible] of the cyst composed of small, closely packed cells with uniform nuclei and scanty cytoplasm. [illegible] characteristic vascular pattern with perivascular proliferation of the tumour cells is seen (H&E x100).

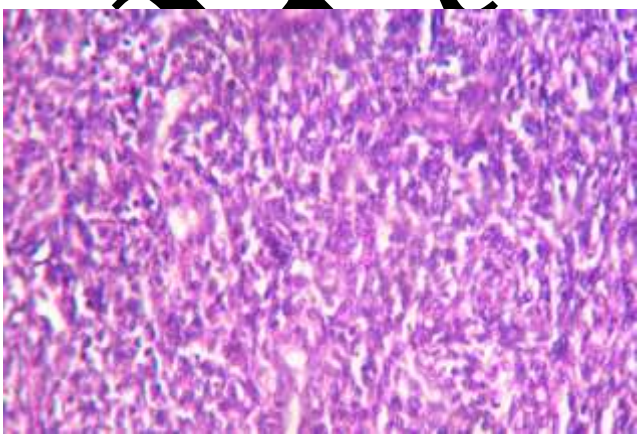

Figure 3: CD10 positivity in the uterine nodule. The periphery of the lesion shows absence of myometrial infiltration (x100)