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Case Report

Endometrial Stromal Sarcoma with Cd56 Expression: A Case Report

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Abstract

Endometrial stromal sarcoma (ESS) is a rare malignant tumour of the endometrium, accounts for less than 1% of all uterine malignancies. Routinely, it is diagnosed morphologically, supported by immunomarkers of CD10 and vimentin. CD56 is used widely in neuroendocrine tumour. In our current practice, CD56 is not used to support the diagnosis of ESS. We present a case of a postmenopausal lady with advanced ESS who had expression of CD56 upon immunohistochemical study.

Keywords: CD56, endometrial stromal sarcoma, immunohistochemistry, uterine leiomyoma, vaginal neoplasm

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Introduction

Endometrial stromal sarcoma (ESS) is a malignant tumour that develops in a connective tissue of the endometrium. This entity accounts for 6% of all uterine sarcomas and only less than 1% of all uterine malignancies (1). The diagnosis is made morphologically, supported by the immunohistochemical positivity of CD10 and vimentin. CD56 is a membrane bound cell surface sialoglycoprotein, a neural-cell adhesion molecule that is expressed in adult neural, neuroectodermal and neuroendocrine tissue (2). Being a highly sensitive immuno-marker, CD56 is used widely for screening of neuroendocrine tumour (2). However, the diagnosis of neuroendocrine tumor should be made based on the morphology in combination with at least positive expression of two neuroendocrine markers. In current practice, CD56 is not utilized to support the diagnosis of endometrial stromal sarcoma. Herein, we present a case of ESS with an expression of CD56.

Case Report

A 65-year-old lady, para 3 presented with postmenopausal bleeding for 3 months duration. She denied family history of malignancy. Upon presentation, she was not anemic. Clinically, she had a good hemodynamic stability. The uterus was mobile at 20 weeks size. She was noted to have a mass over the anterior lower third of vagina. Colposcopy, cystoscopy and hysteroscopy revealed a fluffy endometrium with an endometrial mass seen resembling a degenerating uterine fibroid. There was another mass over the anterior lower third of the vagina measuring 4 x 3cm.

Magnetic resonant imaging of the pelvis revealed a small enhancing lesion measuring 2.8 x 2.7 x 1.8 cm with no clear fat plane with the anterior wall of the distal third of vagina. There was an enlarged iliac node measuring 1.8 x 2.4 cm. Computed tomography scan demonstrated fungating vaginal mass with multiple lobulated uterine masses and presence of multiple lung metastases.

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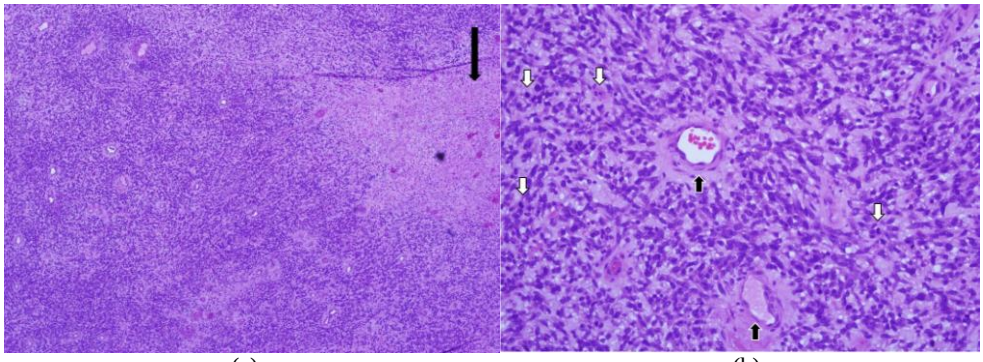
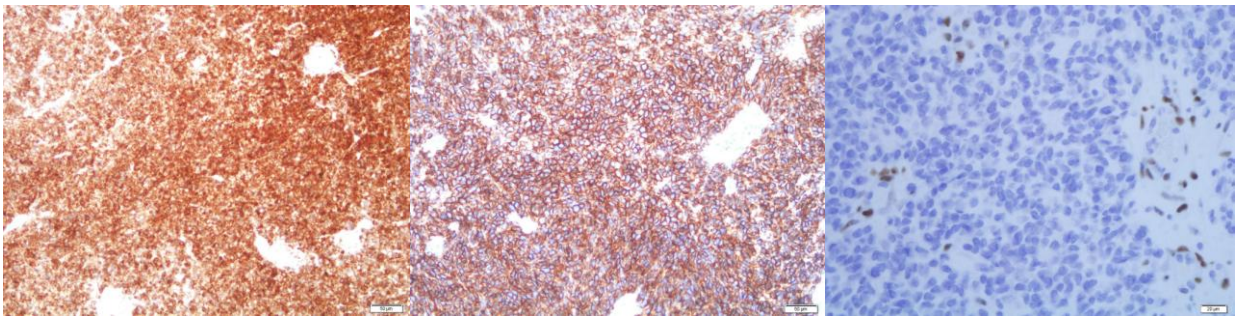


(a) (b)

Figure 1: a) Low power magnification of the tumor with presence of necrosis (arrow). (Hematoxylin and Eosin, original magnification x4); b) The tumor cells arranged in perivascular pseudorosette formation (black arrow). The cells show high grade nuclear atypia with frequent mitosis (white arrow). (Hematoxylin and Eosin, original magnification x20)



(a) (b) (c)

Figure 2: a) Diffused positive cytoplasmic staining of CD10. (Immunohistochemical staining, original magnification x20); b) Diffused strong positivity of CD56 with brownish cytoplasmic membrane staining. (Immunohistochemical staining, original magnification x20); c) Some of the cells show immunoreactivity towards estrogen receptor (brown nuclear staining). (Immunohistochemical staining, original magnification x40)

Initial core biopsy of the tumor suggested a neuroendocrine carcinoma. The tissue morphology showed a high-grade tumor cells with extensive crush artefact with nuclear moulding. The immunoreactivity with antibodies towards chromogranin, synaptophysin and neuron specific enolase (NSE) was negative. However, the tissues were exceptionally positive for CD56.

An excision biopsy of the vaginal mass showed a tumor tissue composed of malignant cells arranged in sheets and fascicular pattern with a whorling surrounding blood vessels forming a pseudorosette (Fig. 1a). The malignant cells were pleomorphic and display enlarged plump hyperchromatic nuclei with conspicuous nucleoli (Fig. 1b). The mitoses were brisk with presence of necrosis. The malignant cells were positive for vimentin, CD10 and focal positivity for ER with diffusely intense CD56 (Fig. 2a-2c). Otherwise, other neuroendocrine markers used (chromogranin, synaptophysin and NSE) were still negative. Broad-spectrum keratin and TTF-1 were also negative, hence excluded the diagnosis of primary neuroendocrine carcinoma of the lung since the patient also noted to have multiple lung masses. These features were consistent with the diagnosis of endometrial stromal sarcoma.

In view of the advance stage of tumor with multiple lung metastases, the patient was subjected for palliative care without active surgical intervention.

Discussion

ESS is a rare sarcoma of the uterus arising from the endometrium. It occurs in premenopausal lady with heavy, irregular menstrual bleeding mimicking leiomyoma. Uterine curettage mostly will lead to the diagnosis. In postmenopausal bleeding, this will create more alarming signs hence needing even more urgent intervention.

The World Health Organization 2014 has classified endometrial stromal tumor into 4 categories as endometrial stromal nodule (ESN), low-grade endometrial stromal sarcoma (LGESS), high-grade endometrial stromal sarcoma (HGESS) and undifferentiated uterine sarcoma (UUS) (3). These

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categories demonstrate molecular alterations ranging from a scarce chromosomal rearrangement to a complex cytogenetic aberration as well as tumor morphology and prognosis (3). ESN is a benign tumor resembling proliferative endometrial stroma (4). ESS nonetheless is a malignant tumor infiltrating into the myometrium and/or lymphovascular spaces with proliferative pattern. In the presence of minimal to no cytological atypia and low mitotic rate, it is categorized as LGESS (4). HGESS however has high-grade cells with high mitotic activity and presence of necrosis (4). UUS otherwise is lacking of a specific pattern and differentiation with high-grade nuclear features (4).

In this reported case, the initial diagnosis was made based on a limited biopsy; hence, it was dependent on the immunomarker solely. CD10 is an important marker for the diagnosis of ESS in which strong and positively diffuse CD10 was found in ESS (5). This marker is helpful in distinguishing ESS from benign tumor such as leiomyoma especially. In our case, it was not done initially as there was no clinical or morphological suspicion of ESS. These had led to a wrong diagnosis of neuroendocrine carcinoma. In practice, larger sample by excision biopsy provides better assessment of the tumour morphology will direct to a correct diagnosis. A good clinical history and the details on the origin of tumor are essential for a pathologist in providing a correct diagnosis.

CD56 alone is not a good indicator to diagnose neuroendocrine carcinoma. It must be supported by at least 2 immunomarkers such as chromogranin A, synaptophysin and/or NSE or CD56. Chromogranin A has a high specificity and sensitivity in neuroendocrine carcinoma (6). In cases with expression of CD56 or NSE alone without expression of chromogranin A, the diagnosis of neuroendocrine carcinoma should be down the list. As in this case, the first biopsy should not be interpreted as neuroendocrine carcinoma just based on CD56 expression alone.

Ohishi et al. has described the use of CD56 in diagnosing ESS in which the immunoreactive cells showed strong intensity of membranous and cytoplasmic staining (7). All of the tumors described were negative for chromogranin A and synaptophysin. In addition to CD10, CD56 can also be an immunomarker to support the diagnosis of ESS. However, in our current setting, CD56 is not been using as a marker for ESS.

Conclusion

CD56 can be one of the markers of endometrial stromal sarcoma in addition to the existence immunomarkers. The knowledge of this marker on ESS is limited as this marker is not routinely performed on this tumor. However, it can be a diagnostic pitfall of neuroendocrine carcinoma. The expression of CD56 alone with negativity of other neuroendocrine marker in female genital tract tumor should resist the diagnosis of neuroendocrine carcinoma.

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