High Grade Endometrial Stromal Sarcoma with Osteoclast-like Giant Cells: A Unique Subtype of a Rare Tumor

David Matthew Saulino, Noah Reilly, Bihong Zhao and Songlin Zhang*

McGovern Medical School, The University of Texas Health Science Center at Houston, USA

*Corresponding author: Dr. Songlin Zhang, MD, PhD, Department of Pathology and Laboratory Medicine, McGovern Medical School, The University of Texas Health Science Center, 6431 Fannin Street, Houston, Texas 77030, USA, Tel: (713)-500-5321, E-mail: Songlin.Zhang@uth.tmc.edu

Abstract

Osteoclast-like giant cells have been reported in variety of different tumor types including breast carcinoma, pancreatic carcinoma, ovary tumor, uterine leiomyosarcoma, hepatocellular carcinoma, renal cell carcinoma. To the best of our knowledge, there are only two previous case reports in the literature showing endometrial stromal sarcoma with osteoclast-like giant cells. Endometrial stromal sarcoma is a rare uterine malignant neoplasm and includes low grade and high grade types. Here, we report a case of high grade endometrial stromal sarcoma with osteoclast-like giant cells. A 54-year-old female presented with a recent history of postmenopausal bleeding, a subsequent computed tomography scan revealed a large uterine/pelvic mass. Histologic examination revealed a malignant spindle cell neoplasm with tumor necrosis and some areas with clusters of osteoclast-like giant cells interspersed in the tumor. The neoplastic spindle cells were positive for CD10 and cyclin D1, negative for desmin, caldesmon and SMA. The osteoclast-like giant cells were also diffusely positive for cyclin D1.

Keywords

Endometrial stromal sarcoma, Osteoclast-like giant cells, Cyclin D1

Introduction

Endometrial stromal sarcomas are rare uterine neoplasms arising in the uterine corpus. Currently, endometrial stromal sarcomas are further separated into low and high-grade subtypes. Low grade endometrial stromal sarcoma is distinguished with its low mitotic activity and its resemblance to typical endometrial stroma [1]. In contrast, high grade endometrial stromal sarcoma has pleomorphic cells with vesicular nuclei and prominent nucleoli [2]. Immunohistochemical staining is very helpful in distinguishing these tumors. Low grade endometrial stromal sarcoma will stain for CD10, ER, and PR [3]. The high-grade subtype is typically negative for these markers and curiously also displays positivity for cyclin D1 [2].

Osteoclast-like giant cells have been reported in an extremely wide variety of different tumor types including melanoma [4], breast carcinoma [4], pancreatic carcinoma [5], ovary [4], uterine leiomyosarcoma [6], hepatocellular carcinoma [4], uterine leiomyosarcoma [7], renal cell carcinoma [4], osteosarcoma [8], and lung carcinoma [4]. While the significance and cause of this phenomenon is poorly understood, recent studies have shown that expression of chemotactic factors by the tumor may be necessary to attract these giant cells [6,8].

Here we report a case of high grade endometrial stromal sarcoma with osteoclast-like giant cells. To the best of our knowledge, only two previous case reports of endometrial stromal sarcomas with osteoclast-like giant cells are present in the literature [9,10].

Case Presentation

Our report is of a 54-year-old female with a 1.5-month history of postmenopausal bleeding. The patient had been postmenopausal for 7 years (since 2010) but noted new vaginal bleeding. The patient also began experiencing severe pelvic and abdominal pain, as well as difficulty with urination. She went to an outside hospital and a CT scan revealed findings concerning for metastatic uterine cancer. In addition to a large 15 cm pelvic
were positive for CD68 (Figure 1E), supporting a histio-
cytic origin. The giant cells were also positive for cyclin
D1 (Figure 1F).

Discussion

During pathology gross examination, the large mass
was located at the lower uterine segment but did not
involving the uterine cervix, which was different from
the clinical impression of large uterine cervix mass. The
cause of this discrepancy is more likely to be the large
tumor mass located at the lower uterine segment made
clinical impression of cervical mass. The clinical differen-
tial diagnosis includes large leiomyoma, leiomyosar-
coma, endometrial stromal sarcoma and carcinosarcoma.
The differential diagnosis of the histology includes leio-
myosarcoma, high grade endometrial stromal sarcoma,
carcinosarcoma with sarcoma overgrowth, and undiffer-
entiated uterine carcinoma. The negative pankeratin
AE1/AE3 rules out carcinosarcoma and undifferentiat
ed uterine carcinoma. The positive CD10 and negative
smooth muscle markers (Caldesmon, desmin and SMA)
make the leiomyosarcoma unlikely and support the di-
agnosis of high grade endometrial stromal sarcoma.

Although endometrial stromal sarcoma is a rare tu-
mor, a variety of different histological features have
been described [11-13]. These features range from
relatively common (sex-cord like pattern, fibromyxoid
pattern) to rare (skeletal muscle differentiation, ossifi-
cation, rhabdoid features) [9]. Only two previous case
reports describe the presence of osteoclast-type giant
cells in association with these tumors [9,10]. It appears
that chemotactic factors expressed by the tumor itself may be responsible for attracting these giant cells. One such factor, receptor activator of nuclear factor kappa-β ligand (RANKL), has been shown to be highly expressed in another case of uterine sarcoma with osteoclast-like giant cells [6]. Additionally, the expression of cyclin D1 has been shown to be upregulated in osteoclast-like giant cells [14,15]. Cyclin D1 expression has also been shown to be associated the formation of giant cells in some studies, including multinucleation and increased chromosome content [15]. Owing to the extremely small sample size, it is currently unknown if this histologic variation has any clinical or treatment consequences. It is our hope that this article inspires future reporting of the rare variation, and that eventually enough data is collected to make effective treatment decisions for the affected patients. We also believe it is very useful to describe histologic variations to avoid inappropriate diagnosis, and perhaps shed more light on the tumor’s biological makeup.

Conclusion
Endometrial stromal sarcoma with osteoclast-like giant cells is a rare histologic variant of endometrial stroma sarcoma, and this is the third reported cases in the literature. The significance of the osteoclast-like giant cell component is not clear, and the cyclin D1 expression in the osteoclast-like giant cells may contribute to the giant cell formation.

References