High grade endometrial stromal sarcoma (HG-ESS) with metastasis to skin

1. Sheeba Bhardwaj (Junior resident, Radiation Oncology)
2. Peoli Mukutawat (Junior resident, Radiation Oncology)
3. Shweta Bhardwaj (Junior resident, Anaesthesiology)
4. Vivek Kaushal (Senior Professor and Head of Department of Radiation Oncology)

Abstract-
High-grade endometrial stromal sarcoma is a rare clinical entity and only few cases have been reported in the literature. Herein we present a case of 55 year old patient who presented with complaints of heavy menstrual bleeding from 2 years. Patient received hemostatic external beam radiotherapy (EBRT) to control the bleeding. 4-months after EBRT patient underwent total abdominal hysterectomy with bilateral salpingo-oophrectomy. Histopathologic examination revealed high-grade endometrial stromal sarcoma (HG-ESS), with myometrium invasion. Patient had progressive disease and developed skin metastasis and lost the follow up.

Key words- endometrial stromal sarcoma, skin metastasis.

Introduction-
Endometrial stromal tumors (EST) are rare tumors of endometrial stromal origin and account approximately 0.2% of all uterine neoplasm. Most common age of presentation is 42-58 years. As per 2014 World Health Organization (WHO) tumor classification system, EST can be classified into four categories: endometrial stromal nodule (ESN), low-grade endometrial stromal sarcoma (LG-ESS), high-grade endometrial stromal sarcoma (HG-ESS), and undifferentiated uterine sarcoma (UUS). HG-ESS is a rare entity. The usual clinical presentation of ESS is abnormal uterine bleeding that occurs in about 90% of women and 70% cases show uterine enlargement. They can present with pelvic pain and dysmenorrhea. An asymptomatic ESS occurs in 25% individuals. About 30 to 50% of the ESS has extra uterine spread at the time of the diagnosis. Herein, we report a case of HG-ESS in a female who presented with abnormal vaginal bleeding from 2 years.

Case report- A 55 year old P4L4 female presented with complaint of heavy menstrual bleeding form 2 years. Her past history was unremarkable. Patient was diagnosed as a case of uterine neoplasm stage IIIB. Patient received hemostatic EBRT (800- cGy single session) after which bleeding was controlled. 4-months after EBRT patient underwent total abdominal hysterectomy with bilateral salpingo-oophrectomy. Histopathologic examination revealed high-grade endometrial stromal sarcoma (HG-ESS), with myometrium invasion. IHC (immuno-histochemistry) examination showed CK (creatine kinase)- negative, vimentin- positive, cyclin D1- positive, Estrogen receptor/Progesterone receptor- negative. Patient had...
progressive disease thereafter and developed skin metastasis (figure) and lost the follow up after 3-months of treatment completion.

(figure- skin metastasis)

Discussion- Endometrial stromal tumors (EST) are rare tumors of endometrial stromal origin. HG-ESS is a rare entity. The common presenting symptoms of HG-ESS are abnormal vaginal bleeding, palpable mass, and pelvic pain. EBRT is an effective measure to control the intractable vaginal bleeding. The patient in our report had heavy vaginal bleeding which was controlled by EBRT. Our patient experienced rapid disease progression and developed cutaneous metastasis and lost the follow up.

Conclusion- ESS is a rare uterine tumor. Due to rarity of cases there is insufficient information about an optimal management of HG-ESS. Development of skin metastasis in our patient confirms the aggressiveness of the disease. The management of tumor requires multimodality treatment from surgery to EBRT and adjuvant chemotherapy. EBRT is an effective measure to treat the heavy vaginal bleeding.

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