A New Case of Primary Signet Ring Cell Carcinoma of the Uterine Endometrium: A Case Report

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Abstract

The presence of signet-ring cell carcinoma (SRC) in an endometrium is extremely rare and it is always necessary to rule out a metastatic neoplasm. We report a 66-year-old woman FIGO stage IIIc2 endometrial carcinoma with a signet-ring cell component found in the endometrium, left ovary and pelvic lymph nodes of the radical hysterectomy with bilateral salpingo-oophorectomy and pelvic and para-aortic lymphadenectomy. The uterine neoplasm invaded more than one-half of the myometrium, left tube, left ovary and para-aortic lymph node. Alcian blue showed the presence of mucin in the SRC. The patient was alive and without evidence of recurrence 17 months after adjuvant chemotherapy. We have discussed the differential diagnosis of this kind of neoplasm and we have reviewed the literature on SRC carcinoma of the endometrium.

ABBREVIATIONS

SRC: Signet Ring Cell Carcinoma

INTRODUCTION

It is well recognized that poorly differentiated adenocarcinoma often accumulate in large or aggregated intracytoplasmic vacuoles that peripherally displace the nucleus and variably distort contours into a crescent shaped configuration. Signet-ring cell carcinoma (SRC) is defined as a tumor composed predominantly or exclusively of SRC, characterized by a central, optically clear, globoid droplet of intracytoplasmic mucin with an eccentrically placed nucleus. SRC are usually gastric, intestinal or rare in the female genital tract. To the best of our knowledge primary carcinoma of the endometrium with SRC has only been observed in three previous cases [1,2]. In fact primary pure endometrial SRC and caution that this tumor can closely mimic the pattern of extragenital carcinoma metastatic to the endometrium. In this paper we report a new case of primary endometrial SRC.

CASE PRESENTATION

A 66-year-old multiparous (gravid 3, para 3) woman was referred to the Department of Obstetrics and Gynecology for persistent abnormal vaginal bleeding of six-month duration. An endometrial curettage was performed, curettage interpreted as highly suggestive of metastatic cancers.
In the hysterectomy specimen, there was fleshy white nodule on the posterior wall of the endometrial cavity (Figure 3). On microscopic examination, the SRC comprised 90% of the tumor, while 10% was represented by poorly differentiated adenocarcinoma cells component. Much of tumor was arranged in sheets with many cells containing large intracytoplasmic vacuoles causing peripheral displacement of nuclei resulting SRC morphology (Figure 4). Nuclear atypia in these cells ranged from mild to severe, and mitosis were frequent. Many lymphoplasmacytic infiltrate accompanied the tumor. There was no transition from SRC pattern to endometrioid adenocarcinoma pattern. The malignant cells diffusely infiltrating endometrial stromal invaded more than one-half of the myometrium (Figure 5), left tube and left ovary. Isthmus and uterine cervix were free of disease. Immunohistochemical staining showed the SRC to be diffused positive for Keratin (AE1/AE3, CAM 5.2), PAS and mucin (alcian blue) (Figure 6). ER and PR immunostains were negative in the SRC component. The endometrial origin is documented by the histopathological examination of the hysterectomy specimen because uterine cervix and isthmus were free of disease.

DISCUSSION

The presence of SRC in a carcinoma of the uterine endometrium strongly raises the possibility of a metastasis. The most common extraterine carcinomas that metastasize to or extend into the endometrium arise in the ovary, breast, or gastrointestinal tract, especially the colon. SRC may also occur in primary ovarian carcinoma. [3,4]. In uterine cervical cancer, there are eleven cases of signet ring cell carcinoma revealed in literature [5,6]. Non neoplastic endometrial stromal signet ring cells may be observed during decidualization, when endometrial stromal cells acquire cytoplasmic vacuoles secondary to accumulation of
glycogen and glycoproteins. A lack of staining for mucin as well as immunohistochemical non-reactivity for epithelial markers typically allow recognition of these non-neoplastic elements [7].

The patient described in this report had an endometrial tumor that on biopsy showed a SRC morphology and a diffusely permeative pattern of shaped entrapment of normal endometrial glands. As noted above, this pattern had been widely emphasize in the literature as indicative of metastasis. Before surgery, during chemotherapy and post chemotherapy, extensive clinical investigation failed to show an extragenital primary malignancies. In this case, a hysterectomy was performed, showing again SRC of the endometrium. The myometrium is invaded by metastases more often than the endometrium. The extragenital malignancy that most frequently metastasizes to uterus is breast carcinoma; it is followed by primary gastrointestinal cancers, especially from stomach and colon. Metastatic breast carcinoma in the uterus is frequently characterized by polygonal cells or SRC, often in a linear or single-file arrangement. [8] We believe this case is of primary endometrial SRC based on several findings. Because, the absence of identified primary tumor elsewhere and absence widespread systemic disease, which is present in nearly all patients with extragenital metastasis to the endometrium.

The prognosis of primary SRC of the endometrium is not well known as a result of the little number of case reports. Only one case has documented survival time longer 6 months [1]. Other cases have not documented survival time [2]. In this case, survival time after operation and chemotherapy is 17 months, it may be longest survival time.

CONCLUSION

In conclusion, we report a primary SRC of the endometrium. Although an extremely rare phenomenon in endometrioid adenocarcinoma, the presence of SRC does not preclude an endometrium primary.

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REFERENCES