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Case Report

Pulmonary metastases of uterine endometrial stromal sarcoma in a young patient: An extreme rarity

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ABSTRACT

Endometrial stromal sarcoma (ESS) is a rare malignant tumor of the endometrium, usually seen in perimenopausal females. We report a case of ESS in a 24 year old woman, presenting as rapid enlargement of a uterine fibroid associated with irregular and excessive vaginal bleeding along with a lung nodule. Hysterectomy was performed. Histopathological examination and immunohistochemistry confirmed ESS. A proper preoperative diagnosis is difficult and in most cases the diagnosis is confirmed after hysterectomy for a presumed benign disease. Simultaneous pulmonary metastasis of ESS is an extremely rare event. As the tumor is rarely encountered, management protocols are still questionable. Although rare, ESS should be considered in the differential diagnosis of all women who present with a rapid enlargement of a uterine leiomyoma. This report is aimed to present a case of endometrial stromal tumor in a young female with simultaneous lung metastasis because of its rare existence and difficulties in establishing histological diagnosis.

1. Introduction

Endometrial stromal sarcoma (ESS) accounts for 0.2% of all the uterine malignancies [1]. The mean age of women with ESS is 42–58 years; 10–25% of the women are premenopausal [2]. The World Health Organization (WHO) classifies endometrial stromal tumors as benign endometrial stromal nodule (ESN) and ESS. ESNs are termed benign, as they do not infiltrate myometrium. In contrast, ESSs are characterized by infiltration of myometrium. The histopathology reveals uniform small cells bearing resemblance to the proliferative stage endometrial stroma. ESSs are classified based on cell morphology and mitotic count into either low-grade (LGESS) or high-grade (HGESS) tumors [3]. In comparison with HGESS, the age group of LGESS is usually younger (45–55 years). LGESS is composed of cells resembling normal endometrial stromal cells, usually with a low mitotic count, although it can be higher and this does not negate the diagnosis. These, like ESN, are associated with a (7;17) translocation. HGESS consists of mostly high-grade round cells with moderate cytoplasm, admixed with variable amounts of low grade spindled cells. Mitotic activity is usually high. The high-grade cells are negative for ER, PR, and CD10, positive for Cyclin D1, and have a characteristic (10;17) translocation (*YWHAE-FAM22* fusion).

The pathogenesis of these tumors is yet to be delineated. Identified risk factors are past exposure to pelvic radiation therapy, long-term tamoxifen use and unopposed estrogen use [4]. We report a case of high grade ESS presenting as rapid enlargement of a uterine leiomyoma. Simultaneous lung metastasis is an extremely rare event. Distant metastasis to lungs may occur after several years [5,6]. Multiple pulmonary nodules are common patterns of metastases of uterine ESS. But a solitary lung nodule is an unusual entity and only a few cases have been documented histologically [7,8].

2. Case report

A 24 years old woman P1L1 presented to the gynaecology outpatient department. She had a history of irregular vaginal bleeding for last 6 months with menorrhagia and dysmenorrhoea, gradually increasing in intensity. On examination, the uterus was enlarged to 28 weeks size. The patient was pale and haemoglobin (Hb) was 7 g%. Vitals were stable. All laboratory tests including hormonal tests were unremarkable. Ultrasonography confirmed a bulky uterus with a well-defined heteroechoic mass of 14 × 9 cm, with cystic degeneration in it. Both adnexa were normal and no free fluid was seen in the pouch of Douglas. Dilatation and curettage (D & C) was done which revealed proliferative endometrium on histopathological examination (Fig. 1). For past 15 days patient also complained of dyspnoea. Chest × ray revealed a radio-opaque lesion of size 3 × 2 cm in right middle zone (Fig. 2) and Computed Tomography Chest showed a well-defined lobulated moderately enhancing lesion in middle zone of right lung. Keeping in view her CT findings, CT guided FNAC was done from lung mass and it revealed features of malignancy (Fig. 3).She underwent total abdominal hysterectomy with bilateral salpingo-oophorectomy for bulky uterus with

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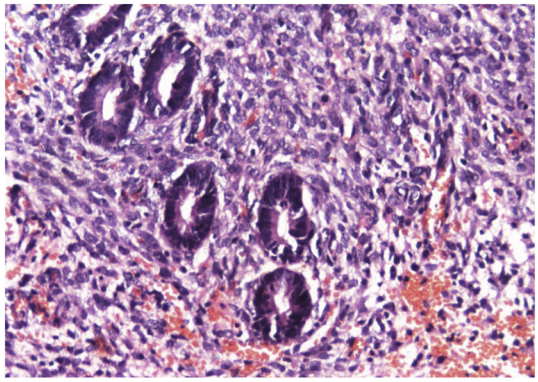

Fig. 1. Dilatation & curettage showing endometrium in proliferative phase (H&E-200×).

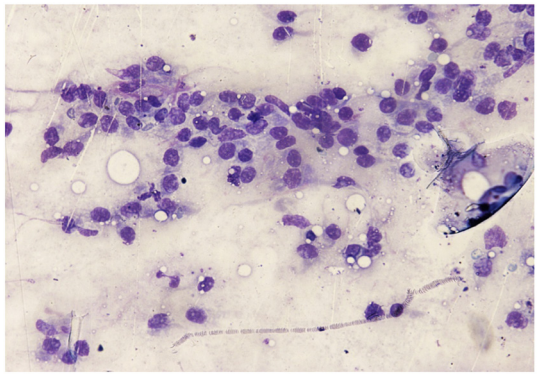

Fig. 3. FNAC showing features compatible with malignant lesion (LEISHMAN-100×).

suspicion of fibroid clinically. The lung nodule was excised through a right posterolateral thoracotomy incision and sent for histopathological examination.

On gross examination, the uterus with cervix measured 13 × 11 × 6 cm. Cut section showed the endometrial cavity filled with multiple nodular fleshy growths with areas of hemorrhage, largest nodule measuring 9 × 5 cm. Tumor was infiltrating more than half of myometrial thickness and extending approximately 3–4 mm from the serosa. The lung nodule was globular measuring 4 × 3 × 2 cm (Fig. 4A–C). Cut section was gray white. Histologically, the sections from various parts of nodule showed densely cellular tumor made up of round/oval/oblong uniform cells with scanty cytoplasm. (Fig. 5A, B) Cells were arranged in diffuse sheets, at places intersecting and anastomosing cords arranged around spiral arteriole. >10 mitotic figures were seen per ten high power fields in the mitotically active areas of the tumor. Irregular shaped, tongue shaped, and circumscribed nests of tumor cells were seen infiltrating the bundles of myometrium. Foci of necrosis and hemorrhage were also seen. The immunohistochemical study showed CD 10 and CK were positive and ER and PR (Fig. 6A & B) negative.

Histopathologic examination revealed a metastatic ESS, which was confirmed by immunohistochemistry (IHC). The estrogen receptor (ER), progesterone receptor (PR) were negative and CD10 was positive in the tumor cells. The histologic features of the lung nodules were identical to that of high grade uterine ESS obtained from hysterectomy specimen. Immunohistochemically, these neoplastic cells showed strong diffuse cytoplasmic staining for CD10 as well as for EGFR (Fig. 7).

The conclusion was that it was ESS of high grade malignancy. She underwent radiotherapy. Postoperative period was uneventful and the patient was discharged on 7th postoperative day.

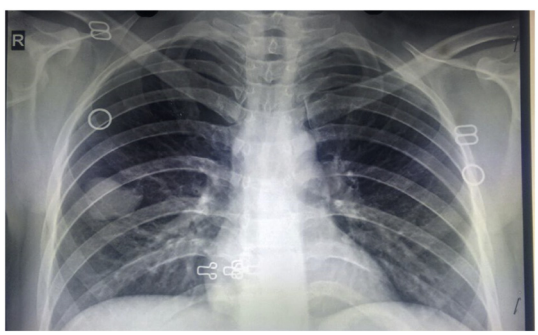

Fig. 2. Chest X ray showing radio opaque lesion in right middle lobe.

3. Discussion

Endometrial stromal sarcomas (ESS) are very rare malignant tumors accounting for 0.2% of all uterine malignancies. The Uterine sarcomas most often affect postmenopausal women, with a median age of 45 and 55 years. Our patient presented at 24 years, which is a rarity in itself [1]. It is an indolent tumor with local recurrences and distant metastasis can occur even years later after initial diagnosis.

The origin and biology of these tumors are poorly understood. A specific translocation t(7;17) (p15;q21) with involvement of two zinc fingers are described in most of the ESS. HGESS have a characteristic (10;17) translocation (*YWHAE-FAM22* fusion).

There is a relation between chromosomal aberrations and endometrial sarcomas. Chromosomal deletion on 7p is the most common finding (55.6%) in ESS and plays a role in tumor development and progression [9].The pathogenesis of ESS is unknown, but exposure to tamoxifen, unopposed estrogens and polycystic disease of ovary have a role to play.

The usual clinical presentation is abnormal uterine bleeding followed by uterine enlargement. They can present with pelvic pain and dysmenorrhoea. Although, the main tumor mass is almost always intramyometrial, most ESS involve the endometrium and uterine curettage may be helpful in preoperative diagnosis [10]. However, when the lesion is completely within the myometrium, the scrapings may not be helpful. Due to the great similarity of ESS with normal endometrium, it may be impossible to diagnose it with certainty on curettage fragments, and the definitive diagnosis can be made only on a hysterectomy specimen. Rarely ESS is initially present at an extra uterine site, most commonly the ovary. It can be a primary or metastatic lesion often from an occult tumor of the endometrium or from a previously undiagnosed case where a hysterectomy was done for a benign leiomyoma of the uterus.

Ultrasound is not reliable and can lead for the incorrect diagnosis of adenomyosis or uterine leiomyoma. Trans-vaginal color Doppler shows low impedance flow compared to other benign tumors. Magnetic resonance imaging can be useful for a preoperative diagnosis. The important imaging features are the presence of bands of low-signal intensity within the area of myometrial invasion. This is due to the worm-like permeation of tumor cells into the myometrium [11].

Immunohistochemistry will help in the detection of tumor. Strong and/or diffuse positivity for CD10 is found in ESS, which are helpful in distinguishing these tumors from histological mimics like cellular leiomyoma that are generally negative. LGESS tumor cells are typically positive for CD10, vimentin, actins, WT-1, IFITM1, ER, and PR. In contrast, the high-grade component is typically negative for ER, and PR, and shows strong and diffuse positivity (>70% tumor cell nuclei) for cyclin D1; CD117 is often positive [12].

The prognosis is dependent on the stage of the disease at the time of diagnosis. Prognostic factors are still controversial. Clinical factors such as age, race, parity, menopausal status, and pathological factors including