CASE REPORT

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Müllerian adenosarcoma of cervix in a young nulliparous woman

Sameera Begum Kader Ibrahim, Prasanta Kumar Deka, Tham Seng Woh

ABSTRACT

Introduction: Müllerian adenosarcoma is a rare tumor of the cervix. Typical adenosarcoma presents as a large polypoid mass occupying the endometrial cavity. It occurs mainly in postmenopausal women. It is a very rare occurrence in adolescent girls and young adults. To date, this neoplasm has been reported in only 10 young adult and adolescent girls. The tumor tends to recur locally rather than to disseminate to distant areas. **Case Report:** A 23-year-old female, single, sexually not active and nulliparous presented to a private hospital for complaints of prolonged menstruation of six months duration. Speculum examination revealed cauliflower like growth arising from the cervix. Polypectomy and hysteroscopic diagnostic dilatation and curettage was done. Histopathology of the specimens showed cervical polyp with features of well-differentiated Müllerian adenosarcoma of low grade with endometrium in secretory phase. A total abdominal hysterectomy, bilateral salpingo-oophorectomy, omentectomy, pelvic and

para-aortic lymphadenectomy, appendectomy were done. **Conclusion:** Adenosarcoma most commonly arises from the endometrium, but some cases from the endocervix. In postmenarchal adolescents, these polypoid tumors usually arise from the cervix, in contrast to postmenopausal women in whom they usually arise from the endometrium. Cervical Müllerian adenosarcoma usually treated by radical hysterectomy even in younger age group as there is higher chance of recurrence later following conservative surgical management. The presence of malignancy should be always kept in mind while resecting a cervical polyp.

Keywords: Müllerian adenosarcoma, Endometrium, Cervix, Polyp, Vaginal bleeding

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INTRODUCTION

Müllerian adenosarcoma of the uterus is a rare tumor containing benign glandular epithelial and malignant mesenchymal elements. Typical adenosarcoma is a low-grade tumor, presents as a large polypoid mass occupying the endometrial cavity and may protrude into the vaginal cavity [1]. This tumor was first described by Clement and Scully in 1974 as Müllerian adenosarcoma. It occurs mainly in the uterus of postmenopausal women but can occur in adolescents and young adults [2]. Extrauterine

locations such as ovaries, cervix, vagina, peritoneum and Pouch of Douglas also had been reported [3–6]. To date, this neoplasm has been reported in only 10 young adult and adolescent girls. There is a case report of a 10-year-old girl, the youngest female ever reported, who was diagnosed with Müllerian adenosarcoma arising from the endocervix [7]. We report one case of Müllerian adenosarcoma of the cervix because of its rarity.

CASE REPORT

A 23-year-old female, single, sexually not active and nulliparous presented to a private hospital for complaints of prolonged menstruation of six months duration associated with continuous dull suprapubic pain with gradual abdominal distension. There was a history of scanty, thick, yellowish, foul-smelling vaginal discharge with pruritus on and off not responded to medications. She was admitted in a private hospital three times in last six months due to symptomatic anemia and received blood transfusion during each admission. There was no significant past medical or surgical illnesses and no family history of malignancy.

On general examination, severe pallor was present. Other vital signs were normal. Breast and thyroid examinations were normal. On per abdominal examination, no abnormality was detected. Initial trans-abdominal ultrasound scan did not reveal any abnormalities.

During the third admission, a trans-abdominal ultrasound scan was repeated which showed endometrial thickness of 18 mm. Consent was taken for vaginal examination, hysteroscopic diagnostic dilatation and curettage under general anesthesia. Speculum examination revealed a cauliflower like growth measuring about 6x6 cm arising from the cervix and a diagnosis of cervical polyp was made. Bimanual examination revealed normal sized uterus with no adnexal pathology. Polypectomy and hysteroscopic dilatation and curettage was done in view of cervical polyp and thickened endometrium (Figure 1). Histopathological report showed a polypoid fragment with leaf like structures which is composed of hypercellular stroma lined by single layer of benign looking epithelial cells. There are scattered glands within a hypercellular stroma with the cells displaying oval to spindle-shaped nuclei with mild pleomorphic and distinct nucleoli (Figures 2 and 3). Around three mitotic figures were noticed in ten high power fields. Immunohistochemical stains showed the mesenchymal cells are diffusely positive for CD10 (Figure 4), Actin (Figure 5) and Desmin panCK (Figure 6) which highlighted the epithelial component. No tumor necrosis, heterologous components or lymphovascular permeation were identified. The diagnosis of cervical polyp with features of well differentiated Müllerian adenosarcoma of low grade with endometrium in secretory phase was given. She was then referred to our hospital—Government

hospital Melaka— for further management. Computed tomography (CT) scan of abdomen and pelvis was done which showed an enlarged uterus with hydrometra and involvement of cervix could not be ruled out as it was bulky. There were bilateral ovarian cysts measuring less than 3x3 cm and no ascites. No evidence of distant metastasis.

The diagnosis of adenosarcoma was revealed to the patient and her parents. After a thorough evaluation of available literature and extensive discussions with the family members, a decision was made to perform radical

Figure 1: Tissue after polypectomy. It was removed in pieces.

Figure 2: Microscopic picture of the polyp with scattered glands within a hypercellular stroma with the cells displaying oval to spindle-shaped nuclei with mild pleomorphic and distinct nucleoli (H&E stain, x400).

Figure 3: Higher magnification of the tumor showing mild nuclear atypia (H&E stain, x400).

Figure 4: Immunohistochemical stains showing the mesenchymal cells are diffusely positive for CD10 which highlights the epithelial component (CD10, x400).

Figure 5: Immunohistochemical stains showing the mesenchymal cells are diffusely positive for Actin which highlights the epithelial component (Actin, x400).

Figure 6: Immunohistochemical stains showing the mesenchymal cells are diffusely positive for Desmin, panCK highlights the epithelial component (panCK, x400).

surgery of the patient. After consented for operation, exploratory laparotomy was carried out. Intraoperatively, a bulky uterus corresponding to 8–10 weeks size of gravid uterus, left endometriotic cyst measuring about 2.6x3.0 cm with normal looking right ovary and a small tumor mass measuring about 3 cm on the surface of small bowel, which is 4 cm from the ileocecal junction were found. Intraabdominal findings were suggestive of an extensive disease/involvement, which warranted our team to proceed with radical surgery in this patient. A total abdominal hysterectomy, bilateral salpingo-oophorectomy, omentectomy, pelvic and paraaortic lymphadenectomy, appendicectomy, a 5-cm resection of small bowel and end-to-end anastomosis were done and peritoneal fluid was sent for cytological study. Her postoperative period was uneventful. She had been clinically free of the disease for 18 months since surgery. Currently, she is on Premarin 0.625 mg once daily and other supplements. Although there are complaints of vague body ache, fatigue and mood-swings, patient is coping well with the treatment given.

Patient is being regularly followed-up once in every 4 months with pelvic examination and ultrasound of abdomen. Histopathological report showed the following findings:

Gross: A brownish mass of 15 mm size at the right endometrial wall with no obvious myometrial invasion, gross involvement of lower uterus, endocervix or parametrium. There are two polyps at the uterine fundus measuring 15x5x5 mm and 10x5x5 mm. Left ovary is slightly enlarged measuring 30x30x25 mm. Other structures were normal.

Microscopy: No residual tumor mass in uterus and cervix. Two benign endometrial polyps, left follicular ovarian cyst, right benign inclusion cyst of ovary and heterotopias of pancreatic tissue over the surface of small bowel. No evidence of malignancy. Peritoneal fluid study is negative for malignancy.

DISCUSSION

A malignant mixed Müllerian tumor is a malignant neoplasm found in the uterus, ovaries, fallopian tubes and other parts of the body that contains both carcinomatous (epithelial tissue) and sarcomatous (connective tissue) components. It is divided into homologous and a heterologous type. A malignant mixed Müllerian tumor (MMMT) account for 2–5% of all tumors derived from the uterus. It is found predominantly in postmenopausal women. Risk factors are similar to those of adenocarcinomas and include obesity, exogenous estrogen therapies and nulliparity. Uterine adenosarcoma occurs in all age groups, but is most common in women after the menopause [8]. Adenosarcoma most commonly arises from the endometrium, but some cases are situated in the endocervix [9]. In postmenarchal adolescents, these polypoid tumors usually arise from the cervix,

in contrast to postmenopausal women in whom they usually arise from the endometrium. Tumors that arise in the cervix account for 2% of all adenocarcinoma of the female genital tract (71% endometrium, 15% ovary and 12% the pelvis) [10]. The common presenting symptom is abnormal vaginal bleeding followed by pelvic pain, an abdominal mass or vaginal discharge. Its association is also described in patients taking tamoxifen and occasional cases have arisen in association with hyperestrogenism or with prior pelvic irradiation [11, 12]. Patient may present with intermenstrual and postcoital bleeding [13]. Clinical diagnosis may be challenging due to the benign gross appearance of the polyps [14]. Pathological diagnosis of low-grade adenocarcinoma is often difficult in a tiny histological specimen. Most of the time, the sarcomatous stroma may be of very low grade and may be misdiagnosed as a benign disease such as adenofibroma [15, 16].

The differential diagnosis of adenocarcinoma includes benign polyp, adenofibroma, embryonal rhabdomyosarcoma and endometrial stromal sarcoma. Cervical Müllerian adenocarcinoma is usually treated by radical hysterectomy even in young age group as there is high recurrence rate following conservative surgical management [7]. Recurrences may occur late. The role of chemotherapy and radiation is limited in the absence of extensive pelvic or residual disease [17]. Since the CT scan finding and intra-abdominal findings during surgery were suggestive of an extensive disease/involvement, which warranted radical surgery in our patient. However, histopathology report revealed no residual tumor mass in the cervix and uterus. So she has not received any adjuvant chemotherapy or radiotherapy. The patient and her family members were informed about the histopathological findings. Poor prognostic factors include sarcomatous overgrowth and high-grade malignant features in the stromal component. The presence of malignancy should be kept in mind always while resecting a cervical polyp.

CONCLUSION

Müllerian adenocarcinoma of the endocervix is rare in young women. The appropriate treatment of this tumor is uncertain because of its rarity in this age group, the malignant potential is not clearly defined and the evidence about the management is limited. On literature review, conservative surgical management such as polypectomy, cone biopsy or trachelectomy had more chance of recurrence. Hence, most of the experts recommend definite or radical surgical management irrespective of the age and parity. Chemotherapy and radiation is not recommended in the absence of extensive pelvic and/or residual disease. From literature review, recurrences have been reported 11 years after conservative treatment. Recurrences may occur late and thus long-term follow-up of these patients is recommended.

Author Contributions

Sameera Begum – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Prasanta kumar Deka – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Final approval of the version to be published

Tham Seng Woh – Substantial contributions to conception and design, Acquisition of data, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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