

A CASE OF EMBRYONAL RHABDOMYOSARCOMA OF THE UTERINE CERVIX IN A MIDDLE-AGED WOMAN

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Embryonal rhabdomyosarcoma (RMS) of the cervix is rare and most commonly occurs in the late teens and early 20s. We report a case of cervical embryonal RMS in a 52-year-old woman. This patient presented with an abnormal vaginal bleeding for 2 months and a mass protruding from the introitus, measuring 7 × 6 cm. She underwent radical abdominal hysterectomy with bilateral pelvic lymph node dissection and radical vaginectomy. The final pathologic result was cervical RMS, consistent with the Intergroup RMS study group IIC. Immunohistochemistry was positive for desmin, myogenin, and myogenic differentiation 1. The patient received conservative management in a convalescent hospital without adjuvant treatment due to cerebral hemorrhage and relapsed septic condition after surgery.

Keywords: Embryonal rhabdomyosarcoma; Uterine cervix

Embryonal Rhabdomyosarcoma (RMS) is the most common soft tissue tumor in childhood and young adults. RMS is rare in adults, with soft-tissue sarcomas making up less than 1% of malignancies in adults and RMS accounting for 3% of all soft-tissue sarcomas. In University of Tehran data only 6 RMS were found (0.39%) among the 1,528 patients with genital tract malignancies [1]. In Korea, a total of 8 cases of RMS were reported in young adults but there has been no case in which the patient was older than 40 years. We report a case of cervical RMS in a 52-year-old woman who underwent radical surgery.

likely embryonal RMS. Magnetic resonance imaging and F-18 fludeoxyglucose (FDG) positron emission tomography/computed tomography also showed a 7 × 6 cm mass arising from the endocervix and protruding outside the vagina cavity to the perineal portion with FDG uptake (Fig. 2). Moreover, parametrial invasion was seen on posterior aspect of the uterine cervix. The serum level of CA-125 was 58.6 U/mL. Other laboratory tests showed no specific abnormality. Her height, weight, and body mass index were 156 cm, 53 kg, and 21.8 kg/m², respectively. Based on the results of these imaging studies and the pathologic result of biopsy, the patient was diagnosed with sarcoma of the uterine cervix.

Case Report

A 52-year-old multiparous woman presented with vaginal bleeding for several weeks and the feeling of a mass protruding from the introitus for 2 months. She has normal menstrual cycles. The patient was referred to our institute with a suspicion of a cervical myoma. In her medical history, she had been diagnosed with cerebral arteriovenous malformation and treated with embolization and gamma knife surgery a few months prior. On the vaginal examination, a 7 × 6 cm soft pinkish irregular mass arising from the uterine cervix and reaching up to the introitus was seen (Fig. 1). Biopsy was performed and the result was favoring sarcoma, most

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Fig. 1. A 7 × 6 cm size soft pinkish irregular mass arising from the uterine cervix and reaching up to the introitus.

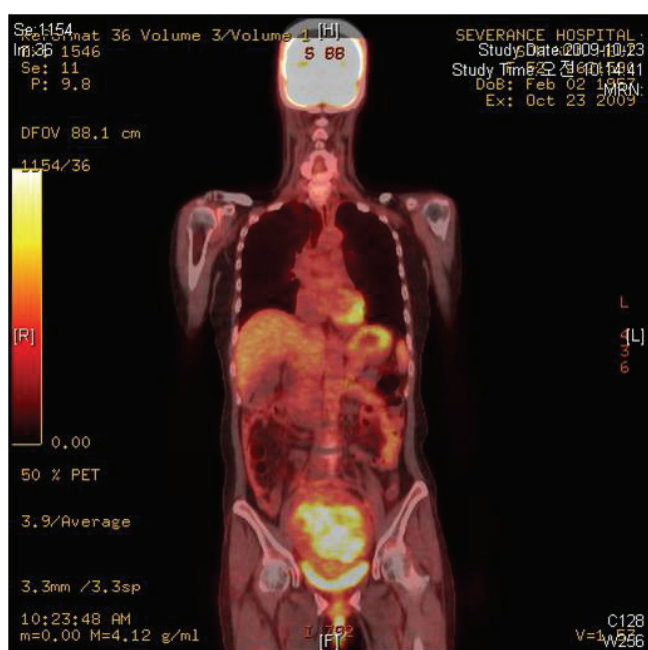


Fig. 2. F-18 fludeoxyglucose (FDG) positron emission tomography/computed tomography finding. There was intense uptake in the protruding vaginal mass, consistent with malignancy. FDG uptake extension to the cervix area was seen, suggestive of malignant involvement and periregional infiltration was suspected.

Therefore, radical abdominal hysterectomy with bilateral salpingo-oophorectomy, and bilateral pelvic lymph node dissection were performed. Additionally, radical vaginectomy was performed to ensure adequate resection margin. The final pathologic result was cervical RMS, consistent with the Intergroup RMS Study Group



Fig. 3. Gross finding after surgery.

IIC. Immunohistochemistry was positive for desmin, myogenin and myogenic differentiation 1 (myoD1). Concurrent chemoradiation therapy was planned. The regimens for chemotherapy was vincristine of 1.5 mg/m² on the first day plus dactinomycin of 0.3 mg/m² and cyclophosphamide of 150 mg/m² from first day until fifth day for 6 cycles. However, she has been observed without adjuvant treatment due to cerebral hemorrhage and relapsed septic condition after surgery. She received conservative management in a convalescent hospital.

1. Histopathologic results

There was a grossly huge pedunculated mass with surface nodularities, measuring 7 × 5.5 × 5 cm, arising closer to endocervix than the junction of the lower uterine segment (Fig. 3). On sections, it was pinkish gray myxoid and showed a multifocal hemorrhage. A well-demarcated bulging intramural mass, measuring 11 × 2 cm was noted in the myometrium. Microscopically, the intramural leiomyoma was involved by the RMS. The desmin immunoreactivity confirmed the myoblastic differentiation. The tumor consisted of oval to spindle shaped malignant embryonal rhabdomyoblasts with eosinophilic cytoplasm (Fig. 4A). Higher magnification of the tumor cells showed cross striation. Histopathological picture favoured embryonal RMS. The soft tissue of right ureteral, left uterosacral, parametrium, anterior and posterior vaginal wall, vaginal cuff, and right uterine artery were also infiltrated by tumor. The lymphovascular space invasion was noted. Pelvic lymph nodes were free of tumor. Immunohistochemistry was positive for phosphotungstic acid-hematoxylin, myoD1, myogenin, desmin, and smooth muscle actin (Fig. 4B).

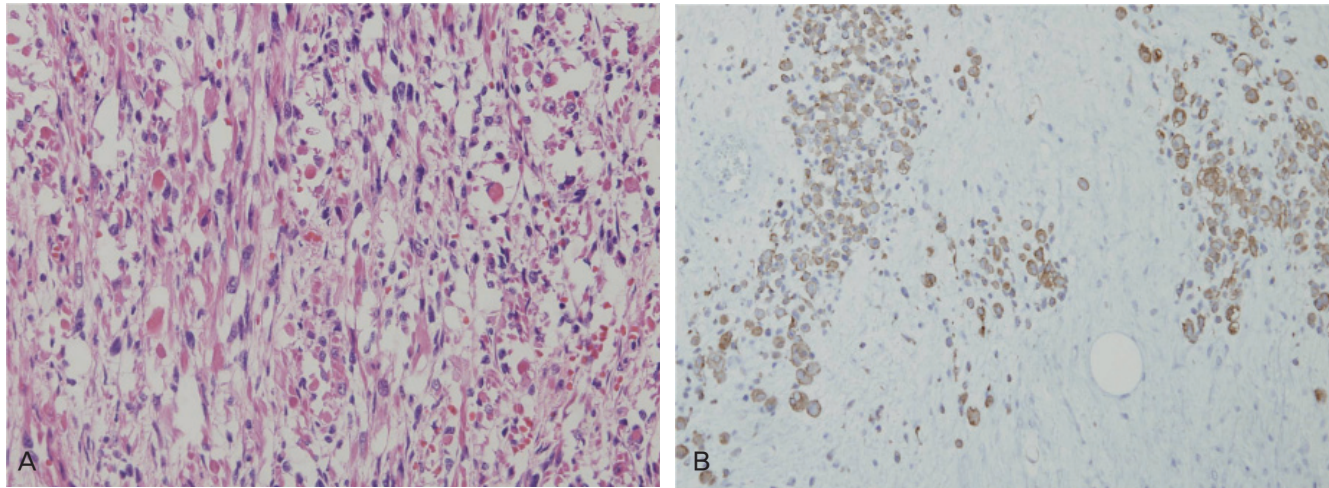


Fig. 4. (A) The tumor consists of oval to spindle shaped malignant embryonal rhabdomyoblasts with eosinophilic cytoplasm (H&E, ×200). (B) The desmin immunoreactivity confirms the myoblastic differentiation (×100).

Table 1. Treatment data and clinical outcomes of the 10 patients aged >40 years with primary cervical rhabdomyosarcoma (From Baiocchi G et al. J Obstet Gynaecol Res 2011;37:940-6, with permission from Journal of Obstetrics and Gynaecology Research.) [8]

Author	Age (yr)	Staging (IRS group)	Surgery	Radiotherapy	Chemotherapy	Outcome	Follow-up (month)	Comment
Ober, 1971	75	I	Polypectomy (3x) + vulvoperineal resection	14.4 Gy Vaginal apex	-	DOD	96	Extension to uterine corpus
Brand, 1987	48	I	TAH + BSO	54 Gy whole pelvis + 35 Gy vaginal apex (radium)	CPM (3 month)	NED	266	Radiotherapy after vaginal recurrence
Vlahos, 1999	45	I	TAH + BSO + PPLND	-	-	DOD	4	Metastasis to abdomen
Miyamoto, 2004	46	I	Polypectomy (x2) + TAH + BSO	-	VCN + ACD	NED	45	-
Ferguson, 2007	56	I	TAH + PPLND	-	VCN + ACD	NED	37	-
Ferguson, 2007	51	I	TAH + BSO + PPLND	-	VCN + ACD + CPM + ADR	NED	7	-
Ferguson, 2007	52	I	TAH + BSO + PPLND	Whole pelvis	-	DOD	17	Recurred after 9 months
Ferguson, 2007	58	I	TAH + PPLND	25 Gy vaginal apex	-	NED	27	Previous BSO
Ferguson, 2007	46	I	TAH + BSO + PPLND	-	-	DOD	12	Recurred after 7 months
Sanders, 2008	47	I	RH + BSO + PLND	-	VCN + ACD + CPM	NED	14	-
Baiocchi, 2010	47	I	RH + BSO + PLND	-	VCN + ACD + CPM	NED	12	-
Current case	52	IIc	RH + BSO + PLND + RV	-	-	-	19	-

IRS, Intergroup Rhabdomyosarcoma Study; DOD, died of the disease; TAH, total abdominal hysterectomy; BSO, bilateral salpingo-oophorectomy; CPM, cyclophosphamide; NED, no evidence of disease; PPLND, pelvic and para-aortic lymph node dissection; VCN, vincristine; ACD, actinomycin D; ADR, adriamycin; RH, radical hysterectomy; RV, radical vaginectomy; PLND, pelvic lymph node dissection.

Discussion

The genitourinary tract is the second most common site of RMS.

The peak incidence age of cervical RMS is the second decade and uterine corpus RMS is most common at menopause phase.

The corresponding tumor in the vagina usually occurs in infants and young children and establishing the diagnosis may be difficult

in an older age group without clinical suspicion of malignancy and in whom cervical polyps are common. The initial clue to the diagnosis is often the gross appearance, in that the polyp may be unusually large and myxoid or there may be multiple polyps. Clinical manifestations of RMS are serosanguinous vaginal discharge and polyp-like mass protruding through vagina. In advanced disease, pain, bladder irritation, and tenesmus were noted. In terminal status, hydronephrosis, anemia, and cachexia were developed.

Survival rate of RMS depends on the site of origin, size, extent of disease, residual disease after treatment, histologic subtype and anaplasia. In vaginal lesions, a better prognosis has been documented than in cervical lesions [2].

The most widely used classification for RMS is that of Horn and Enterline which consists of four histologic subtypes: embryonal, botryoid subtype of embryonal, alveolar, and pleomorphic RMS [3]. There are distinct histological subtypes of RMS that differ in their clinical presentation and behavior. Overall, the embryonal subtype is more common in young children accounting for up to 49% of all RMS [4] and has a better prognosis. The alveolar RMS is secondary most common and more common in adolescents that incidence is about 25% and that is frequently metastatic at diagnosis, and has a worse prognosis. The pleomorphic subtype predominates in adults, among whom RMS is relatively infrequent [5].

According to Ghavimi et al. [5] all patients were treated according to protocols consisting of surgery, radiotherapy, and multiple drug chemotherapy, overall survival rate was 63%. Early stage embryonal RMS of the cervix has been found to have an excellent prognosis. Zeisler et al. [6] demonstrated that the survival rate was 92% in early stage RMS after local excision with chemotherapy. The estimated 5-year survival rates for patients with and without anaplasia were 68% [7].

Only eleven cases of uterine cervix RMS in women over age of 40 years have been reported (Table 1) [8]. We present here the 12th case. As the patient had extensive disease we opted to perform a radical hysterectomy with pelvic lymph node dissection although its role has no consistent data in literature. Our case is the third patient over 40 years of age who was treated with radical hysterectomy followed by the Intergroup Rhabdomyosarcoma Study group IV adjuvant chemotherapy schema [9].

Recently in Korea, Jang et al. [10] reported a RMS in 2001 but it was a case in a 31 year-old woman. In the present case, the patient was a middle-age woman who underwent radical surgery without adjuvant treatment. This is an unusual case of RMS and

the prognosis of the patient needs to be carefully monitored on follow-up.

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중년여성에서 발생한 자궁경부 배아성 횡문근육종 1예

연세대학교 의과대학 ¹산부인과학교실, ²병리학교실

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자궁경부 배아성 횡문근육종은 매우 드물며 사춘기 후반과 20대 초반에서 가장 많이 발생한다. 현재까지 국내외적으로 40세 이상의 중년 여성에서 발생한 자궁경부 배아성 횡문근육종의 보고는 11예에 불과하며 한국에서의 보고는 아직 없는 상황이다. 본 저자는 52세의 중년 여성에서 발생한 자궁경부 배아성 횡문근육종을 경험하였기에 간단한 문헌 고찰과 함께 보고하는 바이다.

중심단어: 배아성 횡문근육종, 자궁경부