

## Case Report

# Sarcoma Botryoides of Uterine Cervix in a 16-Year-Old Girl

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### Abstract

*Sarcoma botryoides of uterine cervix is a malignant neoplasm of cervix which accounts 0.2% of all the malignant tumours of the uterus and its peak incidence is between 14–18 years. Here we report a case of a 16-year-old newly married girl who presented with the complaint of vaginal bleeding for one and half months. Histopathology from cervical tissue revealed sarcoma botryoides grade 2, score 5. She was treated by neo-adjuvant chemotherapy and surgery. She was alive for three years from diagnosis and died due to lung metastasis.*

**Key words:** *Sarcoma botryoides; Embryonal rhabdomyosarcoma; Cervix*

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### Introduction

‘Sarcoma botryoides’, ‘Botryoid sarcoma’ or ‘Botryoid rhabdomyosarcoma’ is a subtype of embryonal rhabdomyosarcoma found in the walls of hollow mucosa-lined structures such as the nasopharynx, common bile duct, urinary bladder of infants and young children or the vagina in females, typically younger than age eight.<sup>1</sup> The name comes from the gross appearance of ‘grape bunches’. The sarcoma botryoides of the cervix uteri occurs very rarely in fertile age<sup>2</sup> since it accounts 0.2% of all the malignant tumours of the uterus.<sup>3</sup> The childhood variant (mean age 23.5 months) of this tumour most frequently affects vagina. In postmenopausal and fertile age (mean age 14–18 years) the cervix uteri is the most common site.<sup>4</sup> The tumour rarely involves the uterine cervix. It arises from the cervical mucosal epithelium or the middle tunica and usually appears as a cervical polyp protruding out from the external orifice. The tumour is rapidly-growing in nature and it metastasizes mainly to lungs and liver by blood circulation.<sup>5</sup>

### Case report

A 16-year-old girl presented at Bangabandhu Sheikh Mujib Medical University with vaginal bleeding for

one and half months. Her previous gynaecological history was uneventful. She had a staging examination under anaesthesia. On speculum examination, multiple finger-like growths were found in exocervix. These growths also extended to the left side of cervix. Cystoscopy and proctosigmoidoscopy showed no abnormalities.

Her full blood count, blood chemistry and liver function tests were within normal range. Chest radiograph was normal and MRI of abdomen showed cervical lesion with involvement of parametrium, anterior vaginal fornix and possible infiltration anteriorly to bladder and posteriorly to rectal wall. Skeletal survey demonstrated bony metastasis at left sacroiliac joint and local invasion into adjacent hip bone. She was classified as stage IV according to the criteria of the Intergroup Rhabdomyosarcoma Study. Microscopic examination revealed sarcoma botryoides, grade 2, score 5. Immunohistochemistry report showed a sarcoma composed of oval and spindle cells arranged irregularly and in fascicles with mitoses and extensive necrosis.

Neo-adjuvant chemotherapy and surgery were planned for the patient. She received neo-adjuvant chemotherapy with inj. vincristine 1.5 mg/m<sup>2</sup> on days 1, 8 and 15, inj. dactinomycin 1.35 mg/m<sup>2</sup> on

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day 1 and inj. cyclophosphamide 2200 mg/m<sup>2</sup> on day 1. Vincristine and dactinomycin were given as bolus and cyclophosphamide was administered as a prolonged infusion. The treatment was repeated every three weeks for three consecutive courses. After completion of three cycles of chemotherapy, follow-up abdominal and transvaginal ultrasound showed small residual disease. Then radical hysterectomy was planned, but the patient refused.

After third cycle of chemotherapy, the patient became pregnant and refused to terminate pregnancy. She was clinically well throughout the pregnancy period and delivered a male baby by caesarian section. Hysterectomy was done just after delivery of the baby. There was residual sarcoma in the operated specimen. Then adjuvant chemotherapy was planned. She again complained of vaginal bleeding, passage of fleshy mass per vagina and respiratory distress. Marked deterioration of general condition made her unfit for chemotherapy. Palliative treatment was given. Subsequently she died three years after diagnosis due to extensive lung metastasis.

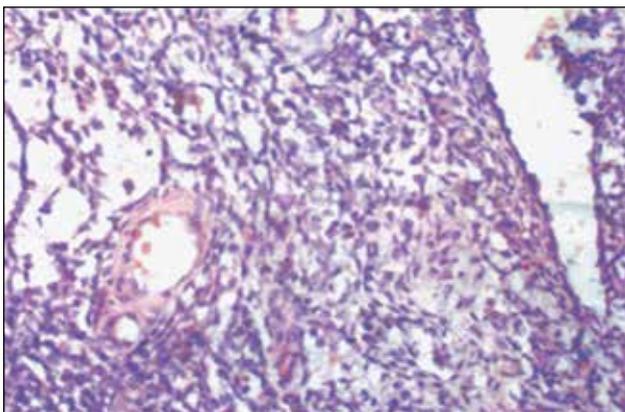


Fig 1. Histopathology of sarcoma botryoides

## Discussion

Rhabdomyosarcoma is the most common malignancy of the lower genital tract in young girls.<sup>2</sup> Daya et al<sup>5</sup> studied on thirteen patients aged 12 to 26 years and found that cervical sarcoma botryoides are preferentially located in the uterine cervix in fertile women. The proper treatment of the cervical sarcoma botryoides is still not well-assessed and it is constantly an object of investigation. Before 1970, the radical surgery including pelvic lymphadenectomy was considered as the first choice

although no significant improvement of survival was proved yet.

The disease is uniformly fatal with a 5-year survival rate between 10–35%.<sup>6</sup> So radical surgery is the choice of treatment. New multidrug chemotherapy regimens with or without radiation therapy are now used in combination with less radical surgery with good results although outcome data are not yet available.<sup>7</sup> We achieved a good outcome in short term survival with chemotherapy, but ultimate result was not satisfactory due to interruption of treatment by pregnancy.

Sarcoma botryoides is usually reported as a vaginal tumour in the female reproductive tract of infants.<sup>8</sup> However, it also occurs rarely in the cervix or uterine fundus.<sup>9</sup> Survival is higher and prognosis is better in vaginal lesions. The survival rates of vaginal and cervical lesions have been reported 96% and 60% respectively.<sup>10</sup>

The prognosis of sarcoma botryoides of the cervix is more satisfactory than other rhabdomyosarcomas of the genital system. The prognosis becomes better especially when it arises as a single polypoid lesion and is completely removed during surgery.<sup>11</sup> But this patient had multiple polyps and aggressive local disease. Sarcoma botryoides of cervix in teen age girl is a rare malignancy, but may occur in this age. Early diagnosis and treatment are essential for better prognosis.

Clinicians must take care about examining any lesions and polyps in cervix at any age and any lesion should be biopsied. The benefits of chemotherapy regimen are not widely described.<sup>12</sup> Therefore, the patient should be under close supervision.

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