Case Report

A Rare Case of Cervical Rhabdomyosarcoma in an Adolescent- A Case Report

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Abstract

We present a rare case of cervical embryonic rhabdomyosarcoma (ERMS) in adolescents, managed with fertility-sparing surgery and chemotherapy. This type of malignant tumour is rare, and the management is not codified. A 15-year-old girl presented with a tongue-like mass per vagina and abnormal discharge. She underwent vaginoscopy under anaesthesia, which revealed a polyp from the endocervix. Hysteroscopic resection was performed, and histopathology showed ERMS. Postoperative imaging suggests the presence of residual local disease without evidence of distant metastasis. A multidisciplinary team (MDT) meeting was conducted, and the family opted for fertility-sparing management. The patient underwent repeat hysteroscopy-guided local excision, followed by eight cycles of chemotherapy, with close surveillance. In conclusion, polyps are rather odd in adolescents; malignancy should always be suspected. Fertility-sparing surgery with chemotherapy is a plausible option in well-selected cases.

Keywords: Adolescent polyp; case report; cervical embryonal rhabdomyosarcoma; fertility sparing surgery; paediatrics and adolescent gynaecology

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Introduction

Cervical Embryonal Rhabdomyosarcoma (ERMS) is extremely rare, and treatment modality is mainly based on the clinician's experience and can range from conservative to radical surgery with chemotherapy (1,2). As it tends to occur in younger age groups, this makes radical surgery a devastating option (1,2). Due to its rarity, there is a lack of evidence, and the currently available literature primarily consists of individual case reports and case series. The management is traditionally extrapolated from that of primary genitourinary ERMS, which is radical surgery followed by chemotherapy with or without radiotherapy (1,3).

The disease stage, tumour subtype, and patient's preferences will be the primary considerations in decision-making. Fertility-sparing surgery followed by chemotherapy is a possible choice in well-selected cases, with radiotherapy reserved for recurrence or advanced disease. Primary cervical ERMS generally has a better prognosis compared to other types of sarcomas of genital tracts (1,2,4). We presented a rare case of cervical ERMS in an adolescent, managed by both a Gynae-oncologist and Paediatrics and Adolescent Gynaecology (PAG) specialist using fertility-sparing surgery via a minimally invasive surgery (MIS) approach followed by chemotherapy with good outcomes.

Case Report

A 15-year-old girl with no prior medical problem, presented with a tongue-like mass per vaginal for 5 months, which is associated with blood-stained, foul-smelling vaginal discharge. She attained menarche at the age of 11, with regular menstruation. No family history of malignancy. She was from a rural area and was brought to medical attention in a small district hospital with one gynaecologist. At that time, a polypoidal mass protruded from the introitus measuring 6 x 3 cm. The surface looks unhealthy and infected, covered with slough, and has a serous discharge (Fig. 1A). She was then referred to a PAG specialist in a tertiary hospital. Due to logistic constraints, the patient lives more than 130 km away from the tertiary hospital with poor road conditions, she could only be seen by a PAG specialist after 3 days. The mass was no longer visible at the perineum during the assessment by the PAG specialist. A computed tomography (CT) scan was performed, showing an ill-defined peripherally enhancing hypodense solid mass of 3.5 x 3.8 x 7.2 cm in the vaginal (Fig. 1B). An examination under anaesthesia was done. Vaginoscopy revealed an irregular mass of 6x3cm arising from the endocervix. The vaginal wall, fornixes, and the other part of the cervix were normal. Hysteroscopy polypectomy was done using a resectoscope whereby the entire polyp base was resected. There was minimal bleeding, and the patient recovered well post-surgery and was discharged the next day.

Due to its rarity, histopathological examination was deemed challenging. The local general pathologist initially reported it as an endocervical polyp with atypical features. A senior pathologist specialising in gynaecology pathology based in the capital of Malaysia was consulted, and it was reported as Adenosarcoma. Subsequently, the slides were discussed with a renowned Professor of Gynaecology pathology in the United Kingdom, with the conclusion of cervical ERMS of botryoid subtype (Fig. 1C).

The pelvic magnetic resonance imaging (MRI) and CT scan after primary resection of the tumour showed a suspicion of residual local disease with no distant metastasis. A multidisciplinary team (MDT) meeting was held. Parents and the child opted for fertility-sparing management after an in-depth discussion, and they were committed to frequent follow-ups and understood the risks of recurrence.

The second combined surgery was carried out by the PAG specialist with the Gynae-oncologist, whereby a diagnostic vaginoscopy and hysteroscopy were performed to identify the suspicious area previously mapped out by the radiologist. The previous surgical site was scarred with no gross suspicious lesion seen on both endo- and ectocervix. The uterine cavity and vaginal were normal. The PAG specialist performed a targeted transcervical resection of the endocervix, which appeared suspicious on MRI using a resectoscope, followed by a cone biopsy by the gynae-oncologist. Fortunately, histopathology examination (HPE) showed no residual malignancy on both specimens. She then received eight cycles of chemotherapy (VAC regime-Vincristine, Actinomycin D and cyclophosphamide). She was assessed after 3rd and 6th cycles of chemotherapy via pelvic ultrasound, hysteroscopy, colposcopy and MRI. Upon completion of her chemotherapy, she had surveillance hysteroscopy and colposcopy every 3-4 months. At present, she was 18 months post-completion of chemotherapy, and she remained well and free from disease recurrence (Fig. 1D).

Discussion

Rhabdomyosarcoma (RMS) is a highly malignant skeletal muscle tumour arising from embryonal mesenchyma (1,3). It is the most common soft tissue sarcoma in children and young adults (4). Based on the World Health Organisation (WHO) classification, RMS is classified into four main groups, which are the embryonal (botryoid, anaplastic), alveolar (solid, anaplastic), spindle cell/sclerosing (including newer variant), and pleomorphic (5) comprised 4-6% of all cancers in this age group and is commonly found in the head and neck, with the genitourinary tract being the second most common site. (1,3,4). A total of 20% of it occurs in the vagina, and only 0.5% of primary RMS is found in the cervix (1,3,4). Due to such rarity, there is inadequate experience to suggest a superior management standard (1,3,4).

Botyroid subtype accounts for 10% of ERMS cases (3). The majority of cervical ERMS cases present with exophytic polypoid growth and abnormal per vaginal bleeding, prompting patients to seek medical attention at an early stage of the disease. This typically results in a better prognosis, as seen in our patient (1,4).

Recent literature suggests that sarcoma of the cervix is less aggressive than sarcoma of the vagina or uterus, hence the evolvement of a more conservative approach in early disease (1). Furthermore, embryonal RMS of the botryoid variant is associated with a much more favourable outcome than the other subtypes, such as alveolar or undifferentiated (1,3,4).

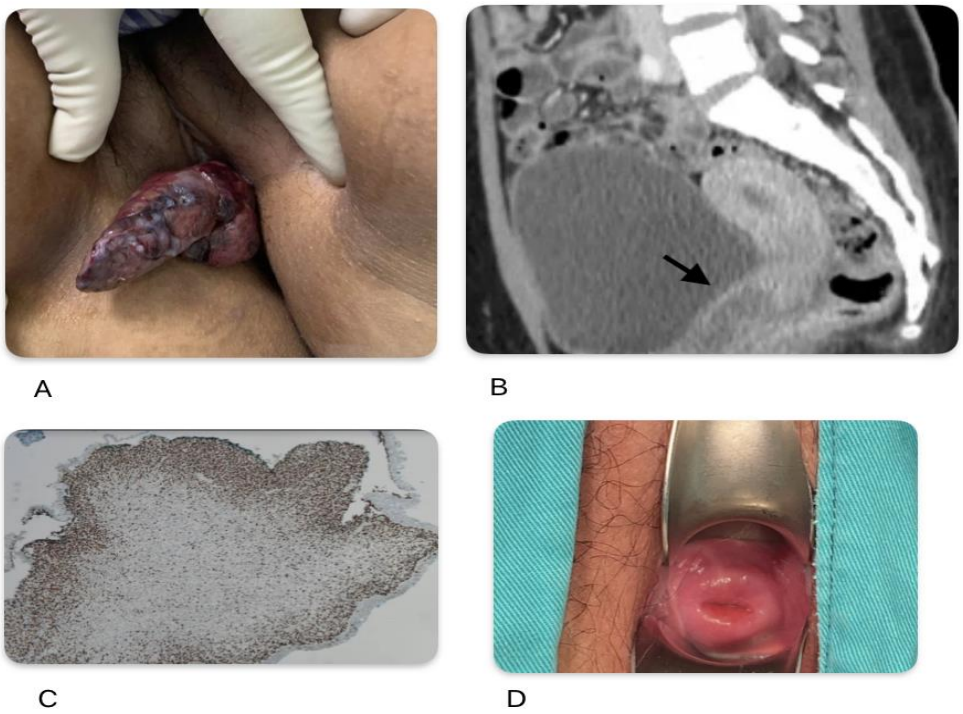

FIGURE 1: (A) showed a polypoidal mass protruding from the introitus; (B) showed a CT image of an ill-defined peripherally enhancing hypodense solid mass 3.5 x3.8x7.2 cm in the vaginal; (C) ki67 immunostain that shows markedly elevated proliferation index in subepithelial layer and lesser in the center which confirms presence of cambium layer which is classically seen in Botryoid subtype; (D) image of the cervix appearance during the recent follow-up (12 months post completion of treatment)

Inter-Group Rhabdomyosarcoma Study Group (IRSG) has classified the ERMS patients into three main clinical groups (1,6). Clinical Group III has an approximately 70% 5-year survival rate for patients with gross residual disease after initial surgery; Clinical Group II (microscopic residual tumour after surgery) has an 80% 5-year survival rate; and Clinical Group I (no residual tumour after surgery) has a more than 90% 5-year survival rate (1,6). From this classification, it can be concluded that even with gross residual disease (clinical group III), the prognosis is relatively promising compared to other malignancies.

The histopathology examination for our patient was deemed challenging due to its rarity, hence unfamiliar among local experts. International collaboration was sought to aid with the final diagnosis. This effort has greatly influenced the direction of decision-making on whether to offer the option of fertility-sparing or a more aggressive radical surgery. With the help of an experienced gynaecology pathologist, the final report of cervical ERMS of the botryoid subtype has opened a new window of opportunity towards fertility-sparing surgery for our patient rather than an aggressive radical surgery recommended in cases of sarcoma, whereby prognosis is deemed poorer (1,3,7). However, this diagnosis dilemma has delayed the timeline to start chemotherapy by 2 months.

Our patient lies in clinical Group I; hence, a local excision of the primary tumour, followed by adjuvant chemotherapy to target any possible microscopic disease, is reasonable. However, due to a lack of evidence to support long-term outcomes, the patient will need close surveillance and eventually a completion surgery after her child-bearing desires. This patient also benefitted greatly from MDT input, whereby the PAG specialist, Gynae-oncologist, Pathologist and Radiologist worked closely to plan her management. She received the MIS approach during both surgeries, which were done by our PAG specialist, who is well-trained in this field. This made her post-operative recovery pleasant and speedy.

We analysed five publications involving cervical ERMS among adolescents from different parts of the world, including first-world countries and resource-underdeveloped countries, to better represent the

TABLE 1: Table showed the summary of 6 publications including current case report and the analysis

Author	Country/ Year	Number of subjects	Subtype	Disease	Treatment	Outcome
Kriseman et al.	USA/ 2012	Total 11 6 patients; age<19; 5patients age>19	8(70%)- Botryoide 2(18%) non-Botryoide 1(9%)-undifferentiated	T1a-2a. no distance/nodal metastasis	Multimodal tx; surgery + CM[a]/RT[b] or both	23 mth[c] f/up[d] -3 local recurrence (all >19yo) -6 no disease (55%) **-1 died of disease (age<19) (9%)** ****refused CM on recurrence**
Jayi et al.	Morocco/ 2014	1 (age :16)	-Botryoide	Local disease	Polypectomy followed by CM(VAC), lacked of response hence hysterectomy +CM	Well
Buruiana et al.	UK/ 2020	3(age :15,16, 19)	-Botryoide (first 2 cases) -High grade sarcoma involving internal os	Local disease	-CM(VIA) followed by trachelectomy then CM - 3rd case: modified radical hysterectomy +PLND + 9x CM-high grade sarcoma	well
Bell et al.	Ghana/ 2021	2 cases (age 17, 16)	Botryoide	Local disease	Polypectomy >6 cycles CM(VAC)	well
Dondi et al.	Systemic review/ 2021	12 studies, 35 subjects (age 2-37	Not mentioned	Local disease	Local resection + CM	Well 5 recurrences 0 death
Leek Mei et al.	Malaysia/ 2023	1 case (Age 15)	Botryoide	Local disease	Polypectomy + CM	No recurrence at time of publication (18mth completion CM)
Summary		**53 patients**		**Local disease**		**8 recurrence (15.1%); 1 death (1.88%) ****

CM[a]: chemotherapy; RT[b]: Radiotherapy; mth[c]: months; f/up[d]: follow up; VAC: Vincristine Adriamycin, cyclophosphamide; VIA : Vincristine, Ifosfamide, Actinomycin; PLND : pelvic lymph node dissection

** death due to recurrence and refusal of further treatment

disease and its management and have a more non-biased discussion (1-5).

The analysis included one case report, 3 case series, and one systemic review as the adolescent to young adult group's patient cohort was similar to ours (Table 1). There were a total of 53 patients, including our case. Of those with local disease, only one patient responded poorly to chemotherapy and hence needed a radical hysterectomy; 8 had local recurrences (15.1%); there was only one reported death (1.88%), but it was a case of refusal of treatment during recurrence, no subtype of the tumour mentioned (Table 1). Therefore, our team was reassured that fertility-sparing treatment followed by chemotherapy and close surveillance is feasible in managing adolescents with primary cervical ERMS.

Conclusion

Fertility-sparing surgery via MIS technique is a feasible option in managing adolescents with early-stage primary cervical ERMS of botryoid subtype as the prognosis is relatively good. MDT discussion and international collaboration are paramount when managing rare cancers in centres with limited resources. Guardians and patients need to understand that there is no standard treatment of date, and opting for fertility-preserving management requires high commitments concerning close surveillance.

Conflict of interest

The authors declared that no conflict of interest could prejudice the impartiality of the research reported.

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Patient consent

The patient and her guardian (patient's mother) consented to this case report and the use of her clinical pictures for learning purposes and publication.

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